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Short-Term Radiographic Evaluation of a Tri-Tapered Femoral Stem in Direct Anterior Total Hip Arthroplasty

William G. Messamore, M.D., Ph.D., Matthew L.G. Vopat, M.D., Elizabeth A. Helsper, M.D., Andrew J. Bachinskas, M.D., Michelle J. Nentwig, M.D., and Tarun Bhargava, M.D.

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ABSTRACT

Introduction. Direct anterior approach (DAA) total hip arthroplasty (THA) has become increasingly popular, largely due to utilization of a true internervous and intermuscular plane. However, recent literature has demonstrated an increased rate of femoral implant subsidence with this approach. Hence, different femoral implants, such as the tri-tapered femoral stem, have been developed to facilitate proper component insertion and positioning to prevent this femoral subsidence. The purpose of this study was to evaluate the subsidence rate of a tri-tapered femoral stem implanted utilizing a DAA, and to determine if the proximal femoral bone quality affects the rate of subsidence.

Methods. A retrospective analysis of 155 consecutive primary THAs performed by a single surgeon was conducted. Age, gender, primary diagnosis, and radiographic measurements of each subject were recorded. Radiological evaluations, such as bone quality, femoral canal fill, and implant subsidence, were measured on standardized anteroposterior (AP) and frog-leg lateral radiographs of the hip at 6-week and 6-month postoperative follow-up evaluations.

Results. The average subsidence of femoral stems was 1.18 ± 0.8 mm. There was no statistical difference in the amount of subsidence based on diagnosis or proximal femoral quality. The tri-tapered stem design consistently filled the proximal canal with an average of 91.9 ± 4.9% fill. Subsidence was not significantly associated with age, canal flare index (CFI), or experience of the surgeon.


INTRODUCTION

Cementless total hip arthroplasty (THA) has emerged as an alternative to cemented systems to improve osseointegration of components and to increase long-term survival of implants. This is particularly important as there has been a recent shift towards younger and higher demand patients undergoing THA. One of the main challenges with cementless stems is achieving solid primary fixation. Stable primary fixation promotes bone ingrowth and facilitates implant stability postoperatively. Additional considerations in cementless systems are preservation of bone stock and reduction of stress-shielding. The primary stability of a cementless femoral stem is affected, not only by the implant design, but also by other factors such as the mechanical quality of the host bone, the presence of gaps around the bone–implant interface, the body weight of the patient, and the size of the implant.

Several surgical approaches can be utilized during THA with direct anterior approach, direct posterior approach, and lateral approach being the most popular. Currently, the direct posterior is the most commonly used approach that allows the surgeon excellent visibility of the joint and precise placement of implants. There is decreasing popularity of the lateral approach. Both posterior and lateral approaches necessitate cutting the gluteus muscle and short external rotators, which may result in muscle weakness, postoperative limp, and increased rehabilitation time.

The direct anterior approach (DAA) has become increasingly more popular among arthroplasty surgeons, in large part due to its utilization of a true internervous and intermuscular plane. This surgical technique protects the hip abductors, the posterior capsule, and the short external rotators. Therefore, with proper component placement, the need for postoperative hip precautions is reduced. Though some studies have demonstrated favorable early results using the DAA compared to more traditional approaches, recent literature has raised concern for increased risk of femoral stem subsidence. This concern has led to the more widespread use of bone-preserving stems. Certain stem design features, such as a tri-taper design, have evolved to improve proximal load transfer, reduce the risk of stress-shielding, and decrease implant loosening. This tri-tapered, wedge-shaped stem incorporates mediolateral and anteroposterior longitudinal tapers to transfer compressive load to the bone interface and a lateral-to-medial taper to enhance load transfer to the medial cortex of the proximal femur. Due to their shorter length and curved distal bullet tip, these implants are more easily inserted and positioned through the DAA since they require less proximal surgical exposure than is required for insertion of a longer stem. Early results support the principle of using metaphyseal-anchoring, calcar-guided short stems.

The purpose of this study was to evaluate the rate of subsidence of a tri-tapered femoral stem implanted utilizing a DAA and to determine if the proximal femoral bone quality affects the rate of subsidence.

METHODS

A retrospective analysis of 221 consecutive primary THAs performed through an anterior approach by a fellowship-trained orthopaedic surgeon (TB) between July 2014 and January 2016 was undertaken. The study was approved by our institutional review board. Three patients were excluded from the dataset for having implants other than a tri-tapered femoral stem. Five were excluded for having no follow-up imaging, 57 for missing 6-week or 6-month follow-up images, and one patient who developed an early infection postoperatively following IV drug use, leaving a cohort of 155 patients with adequate postoperative radiographs for evaluation (Figure 1).
The study group included all patients who were suitable for primary THA ranging in age from 18 to 91 years and who had surgery performed by the lead surgeon through a DAA with implantation of a tri-tapered femoral stem (DJO TaperFill Hip System). All patients received appropriate perioperative antibiotics and antithrombotic agents. All patients were encouraged to ambulate on postoperative day 1. Patients were also encouraged to be weight-bearing and to discontinue the use of walker or cane as quickly as tolerated.

Femoral Canal Fill. Four locations on the femur identified on the anteroposterior (AP) radiograph were used to measure canal fill: (A) at the neck cut, (B) 10 mm above the LT, (C) at the LT, and (D) 60 mm below the LT (Figure 3A), as previously described by Grant et al.18 The canal width and implant width were recorded at each location, with percent fill at each location calculated as implant width divided by canal width. Total average fill was calculated as the average percent fill at all four locations. A line was extended from the lateral cortex to eliminate the flare of the canal at the two proximal locations.

Implant Subsidence. The distance (in millimeters) of the distal femoral stem relative to the lesser trochanter (T-T Distance) was measured on AP radiographs at 6-week and 6-month clinical follow-up (Figure 3B). The implant subsidence was calculated as the difference between these measurements at 6 weeks and 6 months after surgery. Subsidence of 3 mm was defined as significant as previously described by Albers et al.14

Data Analysis. Descriptive statistics included summarizing all variables based on the type of data collected with a measure of central tendency and a measure of variability. Categorical data were summarized with frequencies and percentages. Continuous data were tested for normality using histograms and the Kolmogorov-Smirnov exact test. When data passed the assumption of normality, they were summarized with means and standard deviations. Bivariate associations were assessed with the Chi-square, Mann-Whitney, and Kruskal-Wallis tests. Paired data were evaluated using the paired t-test, or nonparametric Wilcoxon signed-ranks exact test and Friedman test. Because some data were not normally distributed, nonparametric Spearman’s rho was used to assess correlations. All tests were conducted with IBM SPSS Statistics, version 23.
RESULTS

Of the patients in our cohort, 55% (85/155) were female and 45% (70/155) were male. The mean age was 63.7 ± 12 years (range 18 - 91). The majority of our cohort (95%) was non-Hispanic Caucasian. Eighty-three percent (128/155) of patients had a diagnosis of primary osteoarthritis, 11% (17 /155) had osteonecrosis, 2% (93/155) had fracture, 3% (5/155) had dysplasia, and 1% (2/155) had post-Perthes deformity. CFI analysis revealed only one patient had Dorr A bone, 76 had Dorr B bone, and 78 had Dorr C (Table 1).

The mean subsidence of the study cohort was 1.18 mm ± 0.83 (0.0 - 3.1) with only 26% (26/155) femoral stems having subsided more than 2 mm. The mean subsidence for patients treated for primary osteoarthritis was 1.18 ± 0.84 mm, 1.12 ± 0.70 mm for osteonecrosis, 1.27 ± 1.08 mm for fracture, 1.34 ± 0.59 mm for dysplasia, and 1.35 ± 1.05 mm for post-Perthes deformity (Table 2).

Table 1. Demographics data for patients included in the study.

<table>
<thead>
<tr>
<th>Race</th>
<th>No. (%)</th>
<th>Diagnosis</th>
<th>No. (%)</th>
</tr>
</thead>
<tbody>
<tr>
<td>White</td>
<td>148 (95.48)</td>
<td>Primary</td>
<td>128 (82.58)</td>
</tr>
<tr>
<td>Black</td>
<td>5 (3.22)</td>
<td>Osteonecrosis</td>
<td>17 (10.96)</td>
</tr>
<tr>
<td>American Indian</td>
<td>1 (0.64)</td>
<td>Fracture</td>
<td>3 (1.93)</td>
</tr>
<tr>
<td>Hispanic</td>
<td>1 (0.64)</td>
<td>Dysplasia</td>
<td>5 (3.22)</td>
</tr>
<tr>
<td></td>
<td></td>
<td>Perthes</td>
<td>2 (1.29)</td>
</tr>
</tbody>
</table>

Age

<table>
<thead>
<tr>
<th>Proximal Femora Classification</th>
<th>Mean ± SD (yr.)</th>
<th>No. (%)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Mean ± SD (yr.)</td>
<td>63.69 ± 12 (18 - 91)</td>
<td>No. (%)</td>
</tr>
<tr>
<td>55 - 64 years</td>
<td>51 (32.90)</td>
<td>78 (50.32)</td>
</tr>
<tr>
<td>65 - 74 years</td>
<td>40 (25.80)</td>
<td>Mean CFI ± SD = 3.01 ± 0.57 (1.61 - 5.02)</td>
</tr>
<tr>
<td>≥ 75 years</td>
<td>32 (20.64)</td>
<td>2 (1.29)</td>
</tr>
</tbody>
</table>

Sex

<table>
<thead>
<tr>
<th></th>
<th>Male</th>
<th>Female</th>
</tr>
</thead>
<tbody>
<tr>
<td>70 (45.16)</td>
<td>85 (54.83)</td>
<td></td>
</tr>
</tbody>
</table>

Since the Kolmogorov-Smirnov exact test showed that subsidence was not normally distributed (k = 0.116, p = 0.029), the data set was evaluated using the nonparametric Wilcoxon signed-ranks exact test. When the latter statistical test was applied, no statistically significant differences were found between subsidence and other variables, including gender (p = 0.526), race (p = 0.246), Dorr grade (p = 0.580), or etiology (p = 0.861).

As shown in Table 3, the tri-tapered stem design filled most of the femoral canal in all four regions of the AP radiographs. In the proximal canal, averaging the three most proximal measurements, the tri-tapered stem percent fill was approximately 92 ± 5% at 6 weeks and 91 ± 5% at 6 months. The most distal measurement of 60 mm below the lesser trochanter averaged 87 ± 8% at 6 weeks and 87 ± 8% at 6 months.

When analysis was performed with CFI, age, and surgeon experience as covariates, no correlation was found with increasing subsidence (Figure 4). Correlation comparison was executed by Spearman’s rho ranking test (Table 4). Subsidence was not significantly associated with age (ρ = 0.029), CFI index (ρ = 0.015), or experience of the surgeon (ρ = 0.078). No significant association was found between increasing age and decreasing CFI (ρ = -0.155).

Table 2. Average subsidence of femoral stem from 6-week to 6-month follow-up.

<table>
<thead>
<tr>
<th>Subsidence</th>
<th>Average Subsidence Based on Diagnosis</th>
</tr>
</thead>
<tbody>
<tr>
<td>Mean ± SD</td>
<td>1.18 ± 0.83 (0.0 - 3.1)</td>
</tr>
<tr>
<td>No. (%)</td>
<td>Osteonecrosis ± SD = 1.12 ± 0.70 (0.1 - 2.9)</td>
</tr>
<tr>
<td>≤ 1 mm</td>
<td>82 (52.90)</td>
</tr>
<tr>
<td>1 - 2 mm</td>
<td>47 (30.32)</td>
</tr>
<tr>
<td>2 - 3 mm</td>
<td>24 (15.48)</td>
</tr>
<tr>
<td>&gt; 3 mm</td>
<td>2 (1.29)</td>
</tr>
</tbody>
</table>

Average Subsidence Based on Sex

<table>
<thead>
<tr>
<th>Sex</th>
<th>Average Subsidence Based on Sex</th>
</tr>
</thead>
<tbody>
<tr>
<td>Male</td>
<td>1.13 ± 0.82 (0.0 - 3.1)</td>
</tr>
<tr>
<td>Female</td>
<td>1.21 ± 0.83 (0.0 - 3.0)</td>
</tr>
</tbody>
</table>

Table 3. Calculated percentage fill of femoral stems from 6-week to 6-month follow-up.

<table>
<thead>
<tr>
<th>Percent Fill at 6-Week Post-Operative</th>
<th>Percent Fill at 6-month Post-Operative</th>
</tr>
</thead>
<tbody>
<tr>
<td>Neck Cut ± SD</td>
<td>91.6% ± 5.6% (73.3% - 100.0%)</td>
</tr>
<tr>
<td>LT + 10 mm ± SD</td>
<td>92.3% ± 4.4% (76.7% - 100.0%)</td>
</tr>
<tr>
<td>LT ± SD</td>
<td>91.73% ± 4.75% (71.25% - 100.0%)</td>
</tr>
<tr>
<td>LT - 60 mm ± SD</td>
<td>86.7% ± 8.63% (55.7% - 100.0%)</td>
</tr>
</tbody>
</table>

Figure 4. Scatter plots with canal flair index, age, and surgeon experience as covariates.
The DAA has become more popular among surgeons and patients due to its true internervous and intermuscular surgical approach. However, as the DAA has become more widely adopted, some authors have suggested that the approach itself is a risk factor for early femoral stem failure of cementless THA. In their series of revision THA, Meneghini et al. observed the need for revision of a loose femoral stem was 0.29 times more likely with a direct anterior approach than with a posterior approach when controlling for other independent variables. This increase in aseptic failures was attributed to surgeon inexperience, intraoperative femur fractures, and procedure difficulty.

In our study of 155 consecutive patients receiving THA through a DAA, there were no cases of clinically significant femoral stem subsidence with an average subsidence of 1.18 ± 0.83 mm. There was no statistically significant subsidence based on gender, age, or diagnosis. A strength of our study was that we analyzed a consecutive series of patients thereby eliminating any selection bias. Also, the hip reconstruction was standardized among our cohort, as all patients had the same DAA and surgical implants.

The variability in proximal femora anatomy and mechanical quality of host bone is difficult to quantify. CFI was used as a way to measure and categorize our study population objectively according to the Dorr classification. Whereas other publications looking at femoral stem subsidence have excluded patients with Dorr C bone, a strength of our study was inclusion of these patients. There was no correlation between subsidence and Dorr classification ($p$-value = 0.580) or between subsidence and CFI ($r$ value = 0.015). Biomechanically, we would expect to see an increase in subsidence with an increase in CFI (suggesting more osteoporotic bone), but our study did not detect that association.

Surgeon inexperience has been implicated as a cause for early failure of THA performed through a DAA. While the primary surgeon in our study had performed over 1,000 anterior total hip arthroplasties, our series included patients immediately following a transition to a newer implant. We attempted to detect a change in outcomes based on familiarity with the new implant by analyzing the cases consecutively. There was no correlation between experience with the new implant and risk of subsidence. The implant used had the advantage of being designed with the DAA in mind and was shorter than other implants on the market and incorporated a distal flare of the stem to aid in implantation through a DAA affecting our distal canal fill percentages. The stem filled the proximal femoral canal reliably with 91.88 ± 4.94% fill. In smaller femurs, the measurements of distal fill occasionally would include the flare and give artificially lower percentage of distal fill when using the previously described method of Grant et al.

There were no cases of femoral periprosthetic fractures in our series.

Limitations of this study included the retrospective nature of our cohort. Fifty-one patients were excluded from our study because their follow-up radiographs were not available to us for review. Patients referred to our practice by primary providers from outlying areas and insured by various healthcare plans frequently had postoperative radiographs taken at outside facilities to which we had limited access. Nevertheless, we were able to evaluate 70% of our patients in this consecutive series. We were also limited in our access to immediate postoperative imaging and could have missed very early subsidence in the first six weeks, despite no patient having clinically significant subsidence during this period.

Though using a new implant, the primary surgeon in this study was experienced in the technical technique and beyond the “learning curve” that has been well described to correlate with early complications. Therefore, the results of this study may not be representative of that for more inexperienced surgeons. The results of Meneghini et al. likely included many surgeons early in their “learning curve” as that study included 69 different referring surgeons.

In conclusion, the results from this study provide evidence that a THA utilizing the DAA and a tri-tapered femoral stem can achieve consistent and reliable fit regardless of proximal femoral bone quality.

REFERENCES


Keywords: hip arthroplasty, hip prosthesis, diagnostic imaging, orthopedic surgery
INTRODUCTION

Pulmonary edema is a common cause of dyspnea and a leading reason for inpatient admission. Pulmonary edema results from accumulation of fluid or proteins in the alveoli, resulting in a ventilation perfusion mismatch, hence resulting in shortness of breath. The principal mechanisms underlying formation of pulmonary edema are transudation of protein poor fluid into the pulmonary interstitial and alveolar spaces, as happens in cardiogenic pulmonary edema; and increased permeability of the pulmonary endothelium as happens in non-cardiogenic causes such as acute respiratory distress syndrome (ARDS).

Pulmonary edema etiology can be divided into cardiogenic and non-cardiogenic classes. By far, heart diseases are the leading cause of pulmonary edema in inpatient services. Non-cardiogenic pulmonary edema commonly is associated with acute respiratory distress syndrome and pulmonary embolism. Distant organ dysfunction such as renal failure is emerging as a cause of pulmonary edema, especially with widespread access to hemodialysis services and enhanced survival. Noncompliance with this lifesaving procedure, as is common with many medical prescriptions, is the leading reason for this association. This case report detailed a dramatic presentation of pulmonary edema due to noncompliance with hemodialysis.

CASE REPORT

A 50-year-old, African American male with a past medical history of end stage renal disease (ERSD) on hemodialysis presented to the emergency department with shortness of breath, exertional dyspnea (New York Heart Association Class IV), weight gain, bilateral leg edema, and new-onset hemoptysis. He reported to have missed a hemodialysis session, making his last session about seven days prior to presentation. He denied fever and recent flu-like illness. He had no previous history of heart disease or congestive heart failure. On presentation, he was sickly, but with a normal temperature and heart rate. He had tachypnea with a respiratory rate of 22 breaths per minute and elevated blood pressure of 197/93 mmHg. He had pulse oximetry reading of 64% on room air. He was placed on 15 L of oxygen on a non-rebreather mask. Physical examination revealed edema, jugular venous distention, and an S3 gallop on cardiac auscultation. Pulmonary auscultation revealed crackles in the lower lung fields bilaterally, decreased aeration in the upper lung fields bilaterally, and diffuse expiratory wheezes. The patient had anasarca and bilateral lower extremity pitting edema. Significant blood was seen on examination of the patient’s sputum.

Noteworthy laboratory findings included a white blood cell count of 17.2x10^3/L, hemoglobin 14.6 g/dL, blood urea nitrogen 61 mg/dL, serum creatinine 10.9 mg/dL, procalcitonin 5.26 ng/mL, brain natriuretic peptide (BNP) 480 pg/mL, and lactic acid 4.3 mmol/L. The admission chest x-ray (Figure 1) and a computerized tomography (CT) angiogram (Figure 2) ruled out pulmonary emboli but revealed multi-lobar airspace opacities. A 2-D transthoracic echocardiogram revealed an ejection fraction of 65%, grade 2 diastolic dysfunction, and pulmonary artery pressure was estimated at 90 mmHg. There were no valve regurgitations or stenosis noted on Doppler echo.

Figure 1. Admission chest x-ray showed cardiomegaly and bilateral pulmonary alveolar infiltrates.

Figure 2. Admission CT angiogram showed diffuse multi-lobar airspace opacities.

Figure 3. Day 3 chest x-ray showed stable cardiomegaly and improvement of pulmonary infiltrates after two days of hemodialysis.
The patient was started on antibiotics for possible healthcare-associated pneumonia and received hemodialysis for three consecutive days. His hemoptysis resolved after the first session of hemodialysis and, after two days of dialysis treatment, his physical exam was improved with resolved edema, jugular venous distension, and crackles on lung auscultation. A repeat chest x-ray was done on Day 3 of admission (Figure 3), which showed significant improvement in the pulmonary infiltrates.

**DISCUSSION**

Pulmonary edema results from fluid or protein accumulation in the alveoli, largely from processes that alter the Starling forces, changing the net flow of liquids across a membrane.\(^1\)\(^2\)\(^3\) The distinction between cardiogenic and non-cardiogenic pulmonary edema often is difficult. History and examination are often similar and measurement of pulmonary artery pressure of less than 25 mmHg on echocardiography and right heart catheterization measurement of the pulmonary capillary wedge pressure level less than 18 mmHg predicts non-cardiogenic causes.

Non-cardiogenic pulmonary edema is often secondary to acute respiratory distress syndrome, high altitude, opioid overdose, pulmonary embolism, and in our case, renal failure.\(^4\)\(^5\)\(^6\) Accumulation of fluid in the lungs among renal failure patients results from down regulation of the epithelial salt-water transporters such as ENaC, sodium-potassium ATPase and aquaporin-5 in the lung. These transporters are responsible for sodium absorption from the alveolar cavity into the alveolar epithelium cells, with water following passively.\(^7\) Additionally, accumulation of inflammatory cytokines (such as interleukins IL-6, IL-8, IL-1β), tumor necrosis factor α, macrophage inflammatory protein 2, nuclear factor-κB, chemokines, and activated innate immune cells in acute and chronic kidney injury have been theorized to provoke and initiate pathological cascades that leads to acute lung injury and ARDS.\(^8\)

Patients with pulmonary edema generally present with dyspnea, bilateral lower-extremity edema, and chest radiographs showing bilateral alveolar filling patterns.\(^9\) Hemoptysis, as in the above case, is an uncommon presenting symptom. The management of pulmonary edema involves treatment of the underlying disease and supportive measures, such as mechanical ventilation, maintenance of adequate nutrition, and lowering the pulmonary artery wedge pressure with diuretics, ultrafiltration (during hemodialysis), and fluid restriction.

Since our patient had severe hypertension and normal ejection fraction, it is possible diastolic dysfunction contributed at least in part to the pulmonary edema. Moreover, our patient had no mitral stenosis or regurgitation. However, the dramatic improvement in his symptoms following hemodialysis implied, at least to some degree, that the pulmonary edema was volume dependent in the setting of hemodialysis dependent end-stage renal failure.

**REFERENCES**


**Keywords**: pulmonary edema, hemoptysis, renal failure
INTRODUCTION

In 1959, Hirayama et al. first described a benign, self-limiting cervical myelopathy, now known as Hirayama disease, monomelic amyotrophy, or juvenile spinal muscular atrophy of the distal upper extremity. Its usual presentation is unilateral progressive muscular weakness and wasting in the distal upper limb musculature with spontaneous arrest within several years. Often mistaken for a degenerative upper motor neuron disease, Hirayama disease is the result of asymmetric microcirculatory changes in the anterior horn cells of the distal cervical spinal cord resulting from repetitive neck flexion. Over time, the circulatory changes lead to cord atrophy, which manifests as upper extremity weakness. Although both flexor and extensor muscles of the hand are involved, the finger extensors and wrist flexors primarily are affected. The majority of patients develop unilateral amyotrophy, although some present with asymmetric or bilateral atrophy. Sensation and muscle stretch reflexes generally are preserved. The right upper limb is affected more frequently regardless of handedness. Muscle fatigue and contraction fasciculation are the most common presenting symptoms, followed by cold paresis, atrophy, and tremor. The disease predominantly presents in young males, in the teenage years or early twenties. There is no known genetic predisposition of the disease, and it rarely has familial occurrence. Most of the reported cases are from Japan and Asian countries, with little awareness of the disease in North America.

In this case report, an incidence of Hirayama disease that was diagnosed in North America is presented, its pathoetiology explored, and appropriate diagnostic techniques in everyday medical practice are discussed.

CASE REPORT

A 20-year-old male of Bhutanese descent, born in Nepal, presented to neurology clinic for evaluation of left-arm weakness, tremors, and atrophy. Onset of symptoms were three years prior, with gradual progression leading to functional impairment. Interestingly, the patient’s older brother also suffered from progressive weakness in his left arm, which was significantly and functionally impaired. Of note, the patient denied any loss of sensation, numbness, tingling, bulbar symptoms, and weakness of other extremities, including his right arm. On physical examination, the patient had notable loss of muscle bulk in his left forearm, both proximally and distally (forearm circumference: left proximal, 22.25 cm, left distal, 15.5 cm, right proximal, 24.10 cm, right distal, 17.0 cm). Mild tremor and dysmetria in finger-nose–finger testing of the left hand was noted. Strength testing of upper extremities revealed diminished strength in biceps, triceps, brachioradialis, wrist extensors and flexors, finger extensors and flexors, and finger abductors of the left arm, when compared to the right arm. Sensation of touch, temperature, and vibration remained intact in bilateral upper extremities. There was no loss of tone, clonus, fasciculation or pronator drift noted in either arm. Deep tendon reflexes in all extremities and cranial nerve functions were intact bilaterally.

Electromyography (EMG) of the patient’s extremities revealed reduced recruitments, as well as increased amplitude and duration of motor unit action potentials in all muscles of the left upper extremity, except the deltoid. The EMG findings were suggestive of a chronic disorder involving the motor neurons, axons, or both. Magnetic resonance images (MRI) of the brain and spinal cord (cervical, thoracic, and lumbar) were ordered. Imaging of the brain, thoracic, and lumbar spine were unremarkable. MRI of the cervical spine demonstrated asymmetric caliber of the cervical cord, which was smaller on the left. The asymmetry was best visualized on axial imaging and thought to relate to underlying cord atrophy. Mild disc bulges in the cervical spine (C3 through C7) with overall mild narrowing of the underlying thecal sac, but no significant spinal stenosis or mass effect upon the cord, were noted. Given the lack of radiological evidence for radiculopathy, the absence of generalized motor neuron disease, and the positive family history for asymmetric distal upper extremity weakness, there was high suspicion for monomelic amyotrophy or Hirayama disease.

A confirmation MRI of the cervical spine in neck flexion was obtained. Flexion MRI showed diffuse narrowing of the thecal sac accentuated by neck flexion due to tightening of the dura along the dorsal aspect of the thecal sac. There was also re-demonstration of the asymmetric atrophy of the left half of the cervical spinal cord (Figure 1). Additionally, the enhancing dorsal venous plexus, from the level of C3 to the upper thoracic spinal canal, was noted to have prominent and diffuse engorgement on neck flexion, a finding consistent with Hirayama disease.

DISCUSSION

Although first described in 1959, due to its benign course, pathologic study of Hirayama disease was not available until 1982, when the first autopsy was conducted on a 38-year-old male who succumbed to lung cancer. Pathological study revealed an anterior-posterior flattening of the cervical cord, along with asymmetric left sided anterior horn atrophy and central necrosis, all of which were consistent with ischemic changes. Initial conventional neuro-radiologic investigations, such as x-rays and CT myelograms, prior to 1970s failed to reveal significant radiologic abnormalities due to their inability to provide dynamic imaging. However, advancement of imaging techniques in the 1980s, particularly magnetic resonance imaging, combined with knowledge of the pathophysiology of the disease, allowed investigators to use radiological imaging to observe dynamic changes in the spinal cord. Particularly, the MRI of cervical...
Hirayama disease is a progressive myelopathy primarily affecting males in their teens or twenties. It starts with a mild weakness in the arms or hands, followed by atrophy of the shoulder muscles. The disease is more common in Asian populations, particularly in Japan. The clinical features include asymmetric lower cervical spinal cord atrophy, asymmetric cord flattening, and loss of attachment between the posterior dura mater and subjacent lamina. Among the diagnostic features of Hirayama disease, the non-flexion, neutral position MR imaging of cervical spine is the most likely to be ordered by North American physicians evaluating unilateral cervical myelopathy, as they are less likely to be aware of Hirayama disease and the utility of the cervical flexion MRI in its diagnosis.

Chen et al. conducted a retrospective review of the MRIs of 46 patients with confirmed Hirayama disease to identify the sensitivity and specificity of neutral position image findings in disease diagnosis. They noted the following findings to be of particular usefulness in diagnosing Hirayama disease: abnormal cervical curvature, localized lower cervical cord atrophy, asymmetric cord flattening, loss of attachment between the posterior dural sac and subjacent lamina, and intramedullary high-signal intensity on T2-weighted MR images. According to their findings, loss of attachment between the posterior dural sac and the subjacent lamina has the highest combined sensitivity and specificity, at 93.5% and 98% respectively. Localized lower cervical cord atrophy, asymmetric cord flattening, and loss of attachment all had an accuracy of over 80% in detecting Hirayama disease.

Of interest, the possibility existed of a genetic or familial predisposition to Hirayama disease. Our patient had an older brother who had a significantly progressed right-handed weakness and atrophy, which had similar characteristics to Hirayama disease. Given that majority of the cases reported are in individuals of Asian or Japanese origin, there may be inherent susceptibility to the disease in certain genotypes. It also could be possible that the disease, having been discovered in Japan and researched extensively by investigators in Asia and Japan, is simply under diagnosed or misdiagnosed in North America due to lack of suspicion or knowledge.

Increased greatly from the 49% sensitivity of the neutral position MRI. Apart from the obvious finding of anterior displacement of the posterior dura, there are several other flexion MRI findings characteristic of Hirayama disease. These include stretching or tightening of the dural sac, asymmetric flattening of the anterior lower cervical cord, subarachnoid space effacement, posterior epidural space enlargement forming a crescent shaped mass, epidural venous congestion manifesting as epidural flow voids, and enhancement of the epidural space on post-contrast studies.

In our patient, flexion MRI showed distinct tightening of the dural sac and contrast enhancement of the venous plexus, both well-documented Hirayama disease sequelae. The etiology for engorgement of the posterior venous plexus in cervical flexion is multifactorial and a prominent imaging finding in Hirayama disease. Cervical flexion causes anterior shift of the dural canal creating a negative pressure in the posterior spinal canal, compression of the anterior internal vertebral venous plexus diverting blood flow to the posterior venous plexus, and decrease in the drainage of the internal vertebral plexus into the jugular veins, all of which result in dilation of the posterior vertebral venous plexus.

The non-flexion, neutral position MR imaging of cervical spine is the study most likely to be ordered by North American physicians evaluating unilateral cervical myelopathy; as they are less likely to be aware of Hirayama disease and the utility of the cervical flexion MRI in its diagnosis. Although generally considered less sensitive, there is evidence suggesting that neutral position MRI can be a very useful tool in diagnosis.

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CONCLUSION

The intent of this article was to familiarize North American clinicians and radiologists with Hirayama disease, its pathoetiology, and diagnostic neuroradiological techniques, using a case diagnosed in the region. Knowing the typical presentation and demographic prevalence of the disease should help interpret imaging studies better and suggest additional imaging techniques to arrive at the correct diagnosis. In the case of Hirayama disease, a dynamic MRI with neutral and flexion views can assist both the radiologist and clinician.

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Keywords: Hirayama disease, monomelic amyotrophy, spinal muscular atrophy, distal, with upper limb predominance
Nonbacterial Thrombotic Endocarditis

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INTRODUCTION

Nonbacterial thrombotic endocarditis (NBTE) is an uncommon condition that describes aseptic lesions of the heart valves. Historically, NBTE has been synonymous with marantic endocarditis, Libman-Sacks endocarditis, and verrucous endocarditis. NBTE differs from culture-negative endocarditis, which describes infectious origins based on clinical history and symptomatology that have not been readily identified or difficult to culture. It is most commonly a postmortem finding (autopsy series 0.9 - 1.6%) and seen in advanced malignancy.

Based on autopsy reports, patients with an underlying malignancy are six times more likely to develop NBTE compared to the general population (1.25% vs. 0.2%). Solid tumor cancers are associated more commonly with NBTE, with mucin-secreting adenocarcinomas having the highest observed rates. The pathogenesis of NBTE is unclear, but endothelial injury plus hypercoagulability are thought to work synergistically. Interestingly, heart valve vegetations in NBTE have been discovered to be distinctly different from infectious vegetations. Due to the lack of an inflammatory reaction at the site of deposition, NBTE valvular vegetations are dislodged easily, explaining their higher rates of embolization and end-organ infarction. This report presents a rare case of NBTE in a patient with newly diagnosed cancer.

CASE REPORT

A 29-year-old female with a history of alcohol abuse and recurrent pancreatitis presented to her primary care physician complaining of severe dyspnea on exertion, fatigue, and pallor. The patient was referred to the emergency department for further evaluation after outpatient laboratory work revealed significant pancytopenia. She reported subjective fever, chills, sore throat, and productive cough. She denied any symptoms of bleeding at the time. The patient never used tobacco products and quit drinking alcohol several years prior to hospital admission.

Family history was remarkable for a maternal grandfather with history of leukemia and Hodgkin's lymphoma. Her vital signs on admission were remarkable for tachycardia. Physical examination was significant for skin pallor and dry mucosal. Complete Blood Count on presentation showed hemoglobin of 4 g/dl, platelets of 56x10^9/L, and white blood cells of 14x10^9/L, with 40% peripheral blasts, but the Disseminated Intravascular Coagulopathy panel was negative. Peripheral blood flow cytometry was concordant with a diagnosis of acute myelomonocytic leukemia (AMML). She was admitted to the hospital for transfusions and further workup.

A bone marrow biopsy confirmed the diagnosis of AMML, with complex karyotype t (16;16) as well as negative NPM1 and CEBPA mutations. An initial echocardiogram revealed an ejection fraction of 55% with no valvular abnormalities. She was started on broad spectrum antibiotics and blood cultures were collected on admission. Induction chemotherapy was initiated with cytarabine and daunorubicin. Subsequent to chemotherapy treatment, her respiratory status rapidly deteriorated, eventually requiring intubation with worsening bilateral infiltrates on chest x-ray. Bronchoscopy done shortly after intubation was consistent with diffuse alveolar hemorrhage (DAH).

The patient developed neutropenic fever requiring her antibiotics to be expanded to vancomycin, meropenem, acyclovir, and voriconazole. Multiple sets of blood cultures were negative even though she continued to be persistently febrile with no source of infection.

A week after being intubated, she suddenly developed signs of neurological damage including not withdrawing to pain and absent brainstem reflexes; a head CT could not be done due respiratory instability. She quickly developed multi-organ failure and required continuous renal replacement therapy (CRRT). The patient also developed left leg ischemia caused by an arterial clot in the deep femoral artery.

Family discussion led to comfort care. Autopsy revealed a massive non-hemorrhagic infarct involving left frontal, left parietal, and left occipital lobes of the brain resulting from an embolus originating from fibrin vegetation on the aortic valve (Figure 1A). There was pulmonary hemorrhage involving primarily the right and left lower lung lobes (Figure 1B). In addition, fibrin vegetations on the tricuspid valve were noted (Figure 1C). Multiple infarcts in the left and right kidney, as well as the spleen, were noted (Figure 1D). Fibrin thrombi within capillaries of both kidneys and lungs was compatible with disseminated intravascular congestion.

Figure 1. Postmortem evidence of nonbacterial thrombotic endocarditis symptomatology. A) Whole brain exam showing massive non-hemorrhagic infarct affecting multiple territories. B) Pulmonary hemorrhage. C) Fibrin vegetation on the aortic valve. D) Infarcts in bilateral kidneys.
DISCUSSION

NBTE is a rare condition describing sterile thrombi deposition on heart valves in the absence of bacterial growth in blood cultures.\(^1\) NBTE is clinically significant as it increases the risk of systemic embolic events including stroke (a rare event).\(^2\) This case is unusual in that the embolic phenomenon was fatal, as most cases are discovered incidentally post-mortem and are not well described in the literature. Microscopically, NBTE results from agglutinated platelets mixed with strands of fibrin, which is unusual in this case of severe thrombocytopenia.\(^3\) NBTE is most commonly associated with advanced malignancy (80% of cases), systemic lupus erythematosus, and antiphospholipid syndrome. The most common symptom of NBTE is embolic phenomena in up to 50% of patients rather than valvular dysfunction.\(^4\) In this case, NBTE led to significant morbidity and mortality from kidney failure requiring CRRT and massive stroke. Even though NBTE has a low incidence, when cultures yield negative results and significantly thrombogenic patients are not responsive to empirical antibiotic treatment, the possibility of NBTE should be considered.\(^5\)

CONCLUSION

This report described a rare case of NBTE in a young patient with a new diagnosis of Acute Myeloid Leukemia (AML). NBTE is a rare condition associated with high mortality and morbidity rates. It is uncommonly reported in patients with AML and is often a postmortem diagnosis but can present with organ failure from infarction. Diagnosis of NBTE requires a high degree of clinical suspicion especially in patients with malignancy who have clinical features suggestive of endocarditis with a negative workup for an infectious etiology.

REFERENCES


Keywords: nonbacterial thrombotic endocarditis, thrombosis, acute myeloid leukemia, malignancy, rare diseases
Retained Mediastinal Contrast
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Figure 1. Barium swallow study from 2006 demonstrating non-communicating extraluminal contrast within the mediastinum.

Figure 2. Axial computed tomography image from 2019 demonstrating non-communicating extraluminal contrast within the mediastinum.

Figure 3. Sagittal computed tomography image from 2019 demonstrating non-communicating extraluminal contrast within the mediastinum.

CASE DESCRIPTION
A 79-year-old female is presented with history of total laryngectomy, placement of tracheoesophageal puncture for phonation, and radiation therapy secondary to laryngeal cancer in the year 2000. Over the course of 2004, she developed progressive dysphagia due to esophageal strictures requiring a number of dilation procedures with the complication of esophageal perforation in late 2005. The perforation was treated conservatively, however, her esophageal strictures became refractory to dilation. In 2006, the patient underwent a barium swallow study for diagnostic purposes and subsequently had multiple x-rays and computed tomography (CT) scans demonstrating pre-existing non-communicating extraluminal contrast, likely from her known esophageal perforation in late 2005 (Figure 1). Following those scans, she underwent reconstruction of the pharyngoesophageal stricture with tubed left radial forearm free flap. In 2019, she presented for aortic valve replacement work-up and underwent a CT scan with findings initially puzzling to the medical team. The scan demonstrated the retained contrast 13 years later (Figures 2 and 3). The patient did not exhibit any systemic signs of infection or dysphagia.

DISCUSSION
This unprecedented finding has not been reported in the literature previously. Although the patient has not had any clinical sequelae secondary to the retained barium contrast, long-term effects remain unknown. This finding demonstrated the importance of utilizing an absorbable water-soluble contrast as a first step for the diagnosis of perforated viscus given the serious implication of non-absorbable barium contrast and resultant inflammatory reaction. However, in a contained rupture, as in this case, barium contrast did not elicit mediastinitis and the patient remained asymptomatic from it.
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Keywords: contrast media, laryngeal cancer, esophageal diseases
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