KANSAS JOURNAL of MEDICINE

Case Report

Lower Abdominal Juvenile Xanthogranuloma in an Infant: A Rare Site of Presentation

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INTRODUCTION

Juvenile xanthogranuloma (JXG) is a non-Langerhans cell histiocytosis most commonly seen in infancy or early childhood.[1] The first documented case, described by Adamson in 1905, was termed "congenital xanthoma multiplex."[2] JXG is characterized by solitary or multiple yellow to red papules or nodules that are benign and typically self-limiting.[3] These lesions most often appear on the head, neck, or upper trunk, but they may occur anywhere on the body and, in rare cases, involve the eyes, deep soft tissues, or internal organs.[4-6] Atypical presentations, particularly in uncommon locations such as the abdomen, rarely are reported and can pose diagnostic and therapeutic challenges. Here, we present the case of a 10-month-old girl with a large JXG localized to the lower abdominal skin.

CASE REPORT

A 10-month-old female presented with an abdominal skin lesion. Her mother reported that the lesion had been present for two months, was progressively enlarging, and was accompanied by a new, smaller adjacent growth. She also noted that the lesion bled after being accidentally scratched and did not improve with application of Neosporin.

Physical examination revealed a 1.2 cm pink-yellow nodule with central induration on the lower abdomen (Figure 1), along with a smaller yellow umbilicated papule just lateral to the primary lesion. A shave biopsy of the primary lesion was performed to the level of the dermis using a Dermablade, removing the bulk of the lesion.

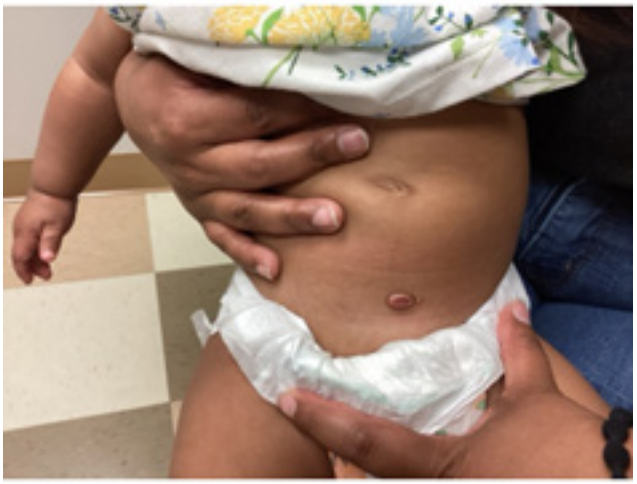

Figure 1. 1.2-centimeter yellowish pink plaque with induration present centrally.

Histopathologic evaluation demonstrated a diffuse dermal infiltrate composed predominantly of histiocytes, including Touton-type giant cells, with scattered lymphocytes and eosinophils (Figures 2 and 3). Immunohistochemical staining showed that the histiocytes were positive for CD68 (Figure 4) and negative for S100, supporting the diagnosis of juvenile xanthogranuloma and helping to exclude other histiocytic disorders. At one-month follow-up, there was no evidence of lesion recurrence.

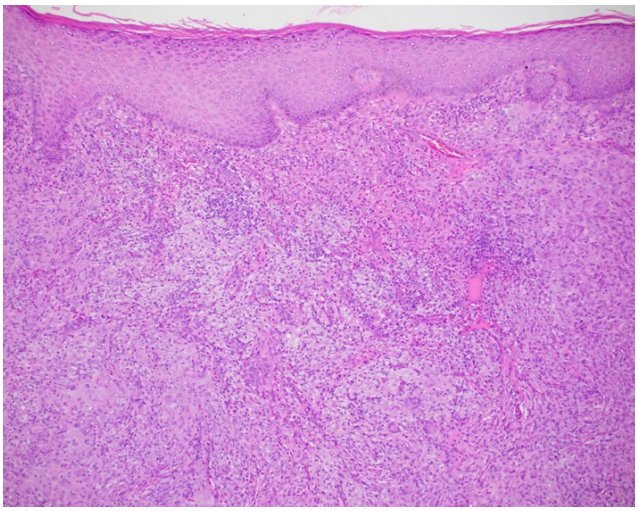

Figure 2. Histological sections revealed foamy histiocytes with scattered Touton giant cells and eosinophils (100x).

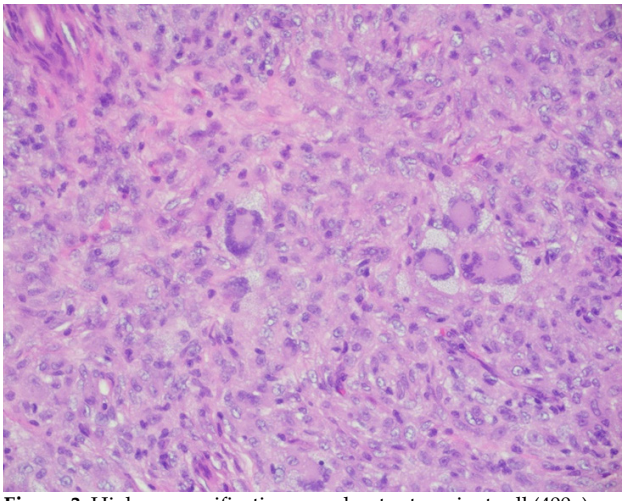

Figure 3. Higher magnification reveals a touton giant cell (400x).

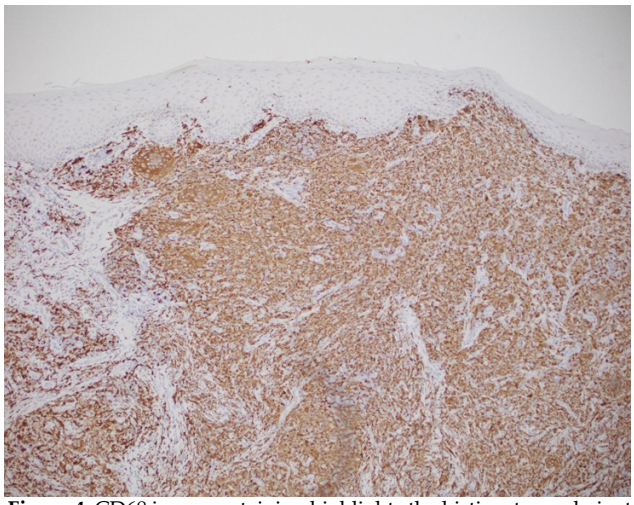

Figure 4. CD68 immunostaining highlights the histiocytes and giant cells (100X).

DISCUSSION

This case is typical in terms of patient age and histopathologic findings but is notable for the size and location of the lesion. The lower abdominal location, rather than the more common head and neck distribution, represents an uncommon presentation of JXG. JXG typically presents as a solitary cutaneous lesion in infancy or early childhood, with a strong predilection for the first two years of life.

Cutaneous lesions usually regress spontaneously within 3-6 years and rarely require treatment. However, atypical presentations may raise concern for extracutaneous involvement and warrant careful evaluation.[1] Characteristically, JXG lesions appear as red or yellow papules or nodules measuring 0.5-2 cm.[4] Although most found on the head, neck, or upper trunk, lesions can occur anywhere on the body. Abdominal involvement has been reported but remains rare; to our knowledge, lower abdominal cutaneous JXG has not been previously described, highlighting the uniqueness of this case.

Extracutaneous involvement, while uncommon, has been reported in the central nervous system, lungs, liver, spleen, kidneys, bone marrow, and gastrointestinal tract.[5] Ocular involvement may occur, particularly in children younger than two years or in those with multiple lesions, and can lead to serious complications, including vision loss.[1]

JXG is thought to originate from macrophages derived from dermal dendrocytes.[7] The clinical differential diagnosis includes Spitz nevus, mastocytoma, dermatofibroma, molluscum contagiosum, and malignancy.[8] Biopsy often is necessary to confirm the diagnosis and exclude alternative conditions. Immunohistochemical staining provides additional support: lesional histiocytes are typically positive for CD68, CD163, and CD4, and negative for CD1a, CD207 (langerin), and S100, which helps distinguish JXG from Langerhans cell histiocytosis.[4,9]

In rare cases (<1%), JXG is associated with juvenile myelomonocytic leukemia (JMML) and neurofibromatosis type 1 (NF1).[10] In patients with NF1, the presence of JXG is considered a warning sign for JMML, with a 20- to 30-fold increased risk compared to those with NF1 alone.[10] Although systemic involvement is uncommon, it can result in significant morbidity and, rarely, mortality. Treatment generally is not required, and systemic therapy is reserved for rare cases with visceral involvement affecting vital organ function.[10]

CONCLUSIONS

This case illustrates a rare histiocytic skin disorder with an unusual lower abdominal presentation and possible early multifocality, which prompted diagnostic biopsy. In infants presenting with an enlarging papule or nodule, JXG should be included in the differential diagnosis, and histologic confirmation should be pursued when the presentation is atypical, or the diagnosis is uncertain. Although most JXG lesions regress spontaneously, shave biopsy can serve both diagnostic and therapeutic purposes. In this case, there was no evidence of ocular or systemic involvement consistent with the typically benign course of the disease. Caregivers should be counseled regarding the potential for ocular and systemic involvement, particularly in patients with multiple lesions, and the importance of close follow-up should be emphasized.

ARTICLE INFORMATION

Received Nov. 6, 2025; Accepted for publication March 2, 2026; Published online Apr. 16, 2026, Kans J Med 2026 Mar-Apr; 19:40-42. https://doi.org/10.17161/kjm.vol19.24768.

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Funding Sources: No funding was utilized in the preparation of this manuscript.

Conflict of Interest Disclosure: Brandon R Litzner is Board member of Rural Access Dermatology Society.

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Keywords: *juvenile xanthogranuloma; lower abdominal lesion; cutaneous nodule; pediatric dermatology; case report*