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Firm, hyperpigmented subcutaneous nodule in the inguinal fold of an infant

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Abstract

Subcutaneous juvenile xanthogranuloma (JXG) of the inguinal fold, an unusual location, was diagnosed in an infant. Subcutaneous JXG should be included in the differential diagnosis of subcutaneous nodules of the lower body, despite the absence of the characteristic yellowish hue usually associated with JXG.

Keywords: juvenile xanthogranuloma, location, morphology, variant, subcutaneous, hyperpigmented, deep

Introduction

Juvenile xanthogranuloma (JXG) is a benign non-Langerhans cell histiocytosis (LCH) that commonly presents as a yellowish papule or nodule on the upper body of infants and young children. Owing to its location in the deep dermis and subcutis, subcutaneous variants may lack the characteristic yellow hue. This, in combination with an unusual location, may complicate the clinical diagnosis. In such cases, biopsy is often necessary for definitive diagnosis.

Case Synopsis

A healthy, seven-month-old girl presented with an asymptomatic nodule of the left inguinal fold that appeared at one month of age. Per her mother, the lesion had intermittently developed overlying erosions. Physical examination showed an approximately 2 cm firm, well-demarcated, hyperpigmented subcutaneous nodule. The most convex areas of the subcutaneous nodule were associated with overlying pink-brown color. (**Figure 1).** No ulcerations, sinus tracts, or erosions were noted on exam. There was no history of trauma to the area and no family history of sick contacts.

The differential diagnosis included pilomatricoma, mastocytoma, Langerhans cell histiocytosis, or hypertrophic scar. Infectious etiologies, such as

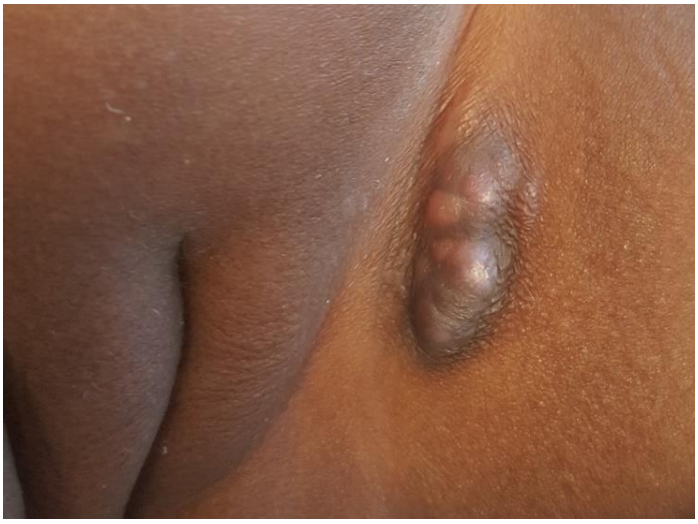

Figure 1. *Well-demarcated, hyperpigmented dermal nodule with overlying pink-brown papules in the left inguinal fold.*

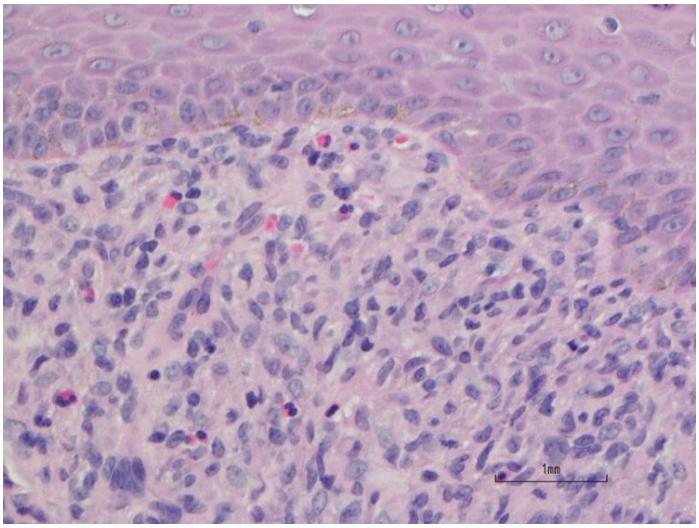

Figure 2. *Dense dermal histiocytic infiltrate with prominent superficial eosinophils (H&E, 40×).*

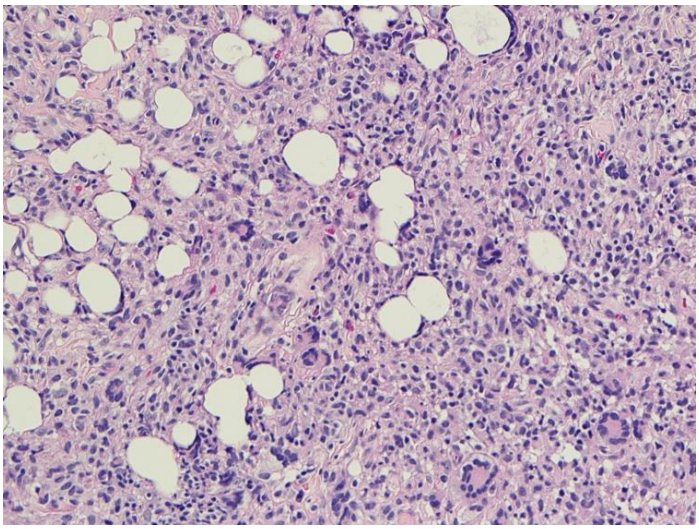

Figure 3. *Eosinophils extending to the subcutis and deep Touton giant cells (H&E, 20×).*

scrofuloderma, were also considered owing to history of intermittent overlying erosions.

Two 4-millimeter punch biopsies of the lesion were performed for histopathology and culture. Histologic examination showed a dense dermal histiocytic infiltrate with prominent superficial eosinophils extending deep into the subcutis, as well as deep Touton giant cells **(Figures 2, 3)**. Strong diffuse staining for CD163, fascin, and factor XIIIa was seen. Staining was positive for S100 and negative for CD1a, CD207 (Langerin), and CD117 **(Figures 4, 5)**. Tissue fungal and mycobacterial cultures were negative. A diagnosis of subcutaneous juvenile xanthogranuloma

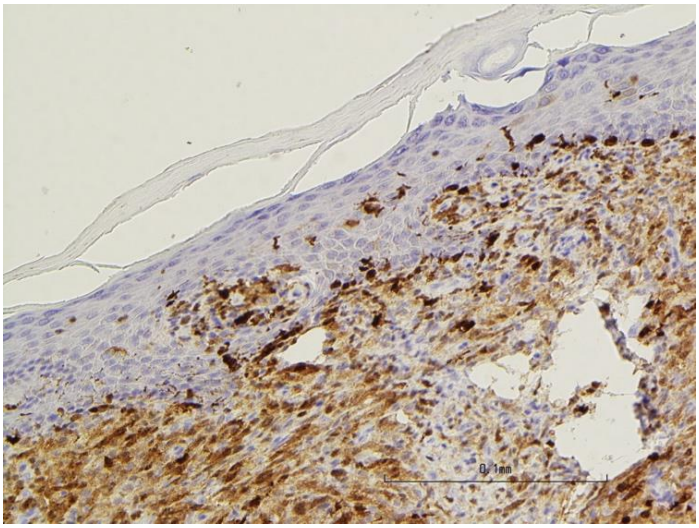

Figure 4. *S100 positive lesional cells as well as positive internal control for dendritic epidermal Langerhans cells (original magnification 20×).*

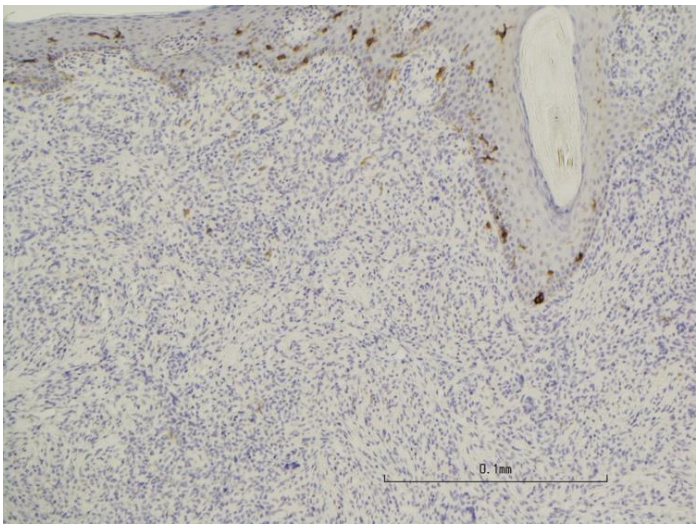

Figure 5. *CD1a negative lesional cells as well as positive internal control for Langerhans cells (original magnification 10×).*

(JXG) was made based on histopathology and negative tissue cultures.

Given the likelihood of spontaneous regression, the lesion was managed expectantly.

Case Discussion

Juvenile xanthogranuloma (JXG) is a common, self-healing, non-Langerhans cell histiocytosis (LCH), usually located on the upper body of infants and young children [1]. JXG most commonly presents as yellowish papules or nodules, although upon initial presentation they may appear red-brown before developing their more recognizable yellow color [1].

Less common cutaneous variants have been described, including keratotic, lichenoid, pedunculated, subcutaneous, clustered, plaque-like, giant, and disseminated forms [2]. Extracutaneous involvement of JXG may involve the eye, lung, liver, spleen, testis, pericardium, gastrointestinal tract, kidney, and deeper soft tissues [2]. Subcutaneous JXG has infrequently been reported in the literature, with almost all cases located on the upper body, including the scalp, forehead, cheek, elbow, retroauricular area, and eyelids [2-4]. Our case was notable for its unusual location on the lower body, specifically the left inguinal fold. The history of intermittent erosions was likely secondary to frictional trauma from the diaper.

Histologically, JXG is characterized by a dense mixed infiltrate of histiocytes, lymphocytes, plasma cells, eosinophils, as well as Touton giant cells. In older lesions, histiocytes are lipid-laden and foamy. Histiocytes stain positively for CD68 and factor XIIIa and usually negatively for S100 and CD1a [1, 2]. Subcutaneous JXG is notable for the mixed histiocytic infiltrate primarily located in the deep dermis and subcutis [2]. Because the upper dermis is less involved, the characteristic yellow color is often absent on exam, making clinical diagnosis difficult. Therefore, in cases of subcutaneous JXG, biopsy is often necessary for definitive diagnosis.

Conclusion

We report a healthy infant with subcutaneous JXG, unusually located on the lower half of the body. We present this case so that clinicians may consider subcutaneous JXG in the differential diagnosis of subcutaneous lesions on the lower half of the body, even in the absence of yellow color.

References

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