Pediatric giant juvenile xanthogranuloma in the parotid gland

Abstract
A full-term, asymptomatic 5-month old infant presented with a nontender parotid mass from birth. Imaging as well as expectant management with steroids was undertaken. After the lesion continued to persist, a superficial parotidectomy was ultimately performed, demonstrating histiocytes, Touton type giant cells, and eosinophils. S-100 and CD1a stains were negative, ruling out LCH and confirming juvenile xanthogranuloma (JXG). JXG is an uncommon benign fibrohistiocytic proliferative disorder, and is classified within the non-Langerhans cell group of histiocytoses. Distinguishing JXG from the more ominous Langerhans cell histiocytosis (LCH) requires histopathology and specialized immunohistochemistry (IHC) staining. Definitive diagnosis requires surgical biopsy, with conservative management recommended unless critical structures such as the facial nerve are involved.

Introduction
Juvenile xanthogranuloma (JXG) is an uncommon benign fibrohistiocytic proliferative disorder, classified within the non-Langerhans cell group of histiocytoses. It is characterized as a yellow-orange cutaneous nodule that tends to regress, resolving completely or leaving a residual atrophic scar. Deeper soft tissues as well as internal organs can be affected by JXG, but those lesions are usually accompanied by a cutaneous lesion. It is extremely rare to have solitary non-cutaneous lesions. Distinguishing JXG from the more ominous Langerhans cell histiocytosis (LCH) requires histopathology as well as specialized IHC staining techniques. A rare case of giant JXG arising in the parotid and occluding the ear canal is described.

Case Presentation
A full-term 5-month old infant presented with an indurated parotid mass from birth that did not transilluminate or display overlying skin changes. The patient had left facial swelling and an occluded left external auditory canal. An MRI revealed a 5.0 x 4.0 x 3.9 cm mass within the parotid, involving the mandibular condyle and extending through the middle ear overlying the otic capsule (Figure 1a, 2a). vascular flow voids and scattered hemosiderin deposits were noted with gadolinium enhancement, suggestive of atypical hemangioma (Figure 2b).

Prednisolone (2mg/kg BID) for 1 month with ranitidine failed to reduce the size of the lesion. A biopsy was recommended for histologic diagnosis. At age 10 months, the patient underwent a superficial parotidectomy, with biopsy revealing sheets of histiocytes admixed with eosinophils, Touton type giant cells, and lymphocytes. IHC stains for S-100/CD1a were negative (Figure 3), ruling out LCH and confirming JXG. Postoperatively he healed well without recurrence.

Conclusions
Given its similarity to other conditions, its self-limiting nature, and implications for treatment, JXG warrants careful consideration in the context of pediatric parotid masses. Definitive diagnosis requires biopsy, with surgical excision recommended if critical structures such as the facial nerve are involved.