Primary Resection and Immediate Autologous Reconstruction of Fronto-orbital Infantile Myofibromatoses

Jennifer L. Lavie, MD*
Camille L. Rogers, PhD*
Mark W. Stalder, MD*†
Hugo St. Hilaire, MD, DDS*†

Summary: Infantile myofibromatosis is an unusual and rare lesion of the bone and soft tissue, which can be seen in the craniofacial skeleton. These complex tumors present a challenge to craniofacial surgeons regarding diagnosis, management, and safe and effective surgical treatment, frequently requiring complex reconstruction. We present the case of a 7-month-old girl with multicentric infantile myofibromatosis of the right parietal and fronto-orbital region, the associated clinical presentation, histopathologic findings, and surgical management, along with a review of the relevant literature. (Plast Reconstr Surg Glob Open 2021;9:e3261; doi: 10.1097/GOX.0000000000003261; Published online 22 January 2021.)

CASE REPORT

A developmentally normal 5-month-old girl presented to pediatric neurosurgery for evaluation of 2 bone nodules of her right fronto-orbital and parietal regions that had been present since birth, increasing in size over the previous 4 months. Magnetic resonance image (MRI) showed an enhancing right fronto-orbital lesion (2 × 1.8 × 1.5 cm), and an enhancing right parietal lesion (1.5 × 1.0 × 0.5 cm), both concerning for histiocytosis. An excisional biopsy of the more accessible parietal lesion was performed to obtain a tissue diagnosis.

Initially, we closely observed the fronto-orbital lesion, as regression commonly occurs.² However, repeat computed topography (CT) imaging at 6 months of age demonstrated significant interval growth (Fig. 1A). Because of the lesion’s intimate relation to the orbit and the

infantile multicentric myofibromatosis involving the craniofacial skeleton in a pediatric patient. We discuss histopathology unique to the condition, the surgical resection, and the subsequent reconstruction of the bone defect.

From the *Division of Plastic and Reconstructive Surgery, Louisiana State University Health Sciences Center, New Orleans, La.; and †Division of Plastic and Reconstructive Surgery, Children’s Hospital, New Orleans, La.

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possibility of its continued growth damaging the eye, surgical management was indicated.

The patient underwent en bloc resection of the lesion with a margin of healthy bone that included a lateral segment of the fronto-orbital rim and a significant portion of the orbital roof (Fig. 1B). The specimen was sent to pathology and was consistent with the parietal lesion. Immediate reconstruction was performed with a segment of full-thickness calvarial bone graft harvested from the right temporo-parietal region and shaped to fit the defect of the fronto-orbital rim and roof. Bioabsorbable polylactic acid plates (Stryker, Kalamazoo, Mich.) were used to reconstruct the parietal donor site defect. Bony contour of the resected structures was reestablished, and the contralateral side was aesthetically matched when the soft tissue was redraped (Fig. 2). Six months follow up demonstrated complete anatomic restoration of the affected segments, with excellent contour and no recurrence.

**DISCUSSION**

The clinical presentation of infantile myofibromatosis, mostly presenting during the first year of life, includes nodules that are painless, well-encapsulated, rubbery to hard in texture, and freely mobile to fixed. The orbit is a common location for solitary lesions; however, there are few reported cases of multicentric lesions involving the craniofacial skeleton without visceral involvement. Mynatt et al. reviewed 24 cases from the English literature, and only 1 had multicentric disease with orbital involvement, similar to our patient. Morbidity is related to the locally infiltrative nature of the lesions, and multicentric lesions of the head and neck
can cause significant problems due to extrinsic compression of nearby structures. Some of the reported complications include stridor in cases of lesions of the larynx, proptosis, eyelid ptosis, displacement of the globe, and development of astigmatism with orbital lesions, and nasal obstruction with lesions of the nasal bones.2,5–7

Infantile myofibromatosis can have a benign radiographic appearance while ultimately being locally destructive, as in our patient. In craniofacial and calvarial involvement, the lesions are often radiolucent with a sclerotic rim. The differential diagnoses for these findings include both Histiocytosis X and fibrous dysplasia, all having similar clinical presentations in the craniofacial skeleton.5,8 Thus, the histopathologic analysis is critical for definitive diagnosis.1 Defining features of these lesions are their nodularity and myofibroblastic appearance.8 They possess a combination of spindled, fibroblast-like cells and fusiform, smooth muscle-like cells with elongated fascicular nuclei and a central hemangiopericytoma pattern. There are, on average, 3–4 mitotic figures per high power fields with mild or no atypia.1,5,8

Currently there exists no definitive treatment algorithm for multicentric infantile myofibromatosis. Fortunately, spontaneous regression is common; so patients are frequently managed with observation. Nonsurgical methods of treatment are available, including corticosteroids, chemotherapeutic agents, or radiotherapy, but these can introduce a significant risk of adverse outcomes in infants and children.2,10 Thus, clinical judgment is essential when there exists a risk of significant morbidity, which necessitates operative intervention.5,6,7,10 Although reported recurrence rates are relatively low (7%), complete local excision with immediate reconstruction is recommended in surgical cases.2,6,8

CONCLUSIONS

Infantile myofibromatosis is a unique clinical and pathologic condition that may be treated with observation and nonsurgical measures, as many cases spontaneously regress. However, surgical resection is recommended if there is an apparent or impending mass effect with the potential to result in detrimental functional or aesthetically morbid outcomes. In our patient, resection and immediate autologous reconstruction was indicated to prevent impending orbital compression and visual impairment, which was performed with a very satisfactory reconstructive result without complications.

Mark W. Stalder, MD
Division of Plastic and Reconstructive Surgery
Louisiana State University
1542 Tulane Avenue New Orleans, La 70112
Division of Plastic and Reconstructive Surgery
Children’s Hospital New Orleans
200 Henry Clay
New Orleans, LA 70118
E-mail: mstald@lsuhsc.edu

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