**Solitary Intraosseous Infantile Myofibroma of the Orbital Roof**

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**Background:** There are several rare tumors that can cause proptosis in an infant, including infantile myofibroma.

**Methods:** A 3-month-old infant developed a painless, bone-destructive superomedial orbital mass, raising concern for orbital malignant neoplasms. Computed tomography disclosed a bone-destructive mass of the sphenoid wing. On magnetic resonance imaging, the intraosseous mass was well-circumscribed, surrounded by cortical bone, and showed prominent enhancement.

**Results:** Superomedial orbitotomy and biopsy revealed a lesion composed of spindled to stellate cells, without mitotic activity, set in a fibromyxoid stroma. Immunohistochemical stains were positive for vimentin and actin. Ultrastructurally, there were actinlike thin filaments, mitochondria, and rough endoplasmic reticulum, confirming a myofibroblastic proliferation and supporting the diagnosis of congenital infantile myofibroma.

**Conclusion:** Infantile myofibroma is a benign tumor that occurs rarely in the ocular region but can cause prominent bone destruction, misleading the clinician to suspect a malignant neoplasm.


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**REPORT OF A CASE**

A 1-month-old white girl was noted by her mother to have painless left proptosis that did not change on straining or crying and persisted for 2 months. There was no birth trauma or neurofibromatosis. On examination, the child seemed systemically healthy. The visual acuity was fix and follow in both eyes. The ocular findings revealed 5 mm of left proptosis (Figure 1), with resistance to retropulsion and upper eyelid retraction.

Orbital computed tomography demonstrated an osteolytic lesion with a soft tissue component involving the left sphenoid bone (Figure 2). Magnetic resonance imaging was employed to further characterize the mass and disclosed a 2.3 × 2.0 × 2.0-cm, discretely margined, lytic lesion with homogeneous enhancement. On T1-weighted images the mass appeared nearly isointense with brain, and on T2-weighted images it was mildly hyperintense compared with brain (Figure 3 and Figure 4). The margins of the expansile mass were delimited by cortical bone.

A transcutaneous superomedial extraperiosteal biopsy was performed. At surgery the orbital periosteum was intact, with the mass entirely confined to the orbital roof. The gray gelatinous mass was almost totally removed by piecemeal excision. A thin layer of tissue lining the cortical bone adjacent to brain was left to prevent inadvertent dural leak.

Histopathologically, the lesion consisted of spindled to stellate cells with zones of sparse growth and prominent fibromyxomatous stroma (Figure 5). Mitotic activity was absent. Immunoperoxidase stains were positive for vimentin (a mesenchymal cell intermediate filament) and actin (a contractile protein found in many cell types). Smooth muscle stains (muscle-specific actin and desmin) were negative. Immunoperoxidase stains for factor
VIII, low-molecular-weight keratin, S100 protein, and leukocyte common antigen were also negative. Ultrastructural examination of glutaraldehyde-fixed tissue revealed mitochondria, rough endoplasmic reticulum, and actin-like thin filaments within tumor cells (Figure 6). Smooth muscle structures were absent. These characteristic findings of myofibroblasts supported the diagnosis of congenital myofibroma.

**COMMENT**

In an extensive review of 340 orbital tumors in children, Kodsi et al found only 1 case (incidence, 0.3%) of orbital myofibroma. In a series of 645 orbital tumors, Shields et al reported no case of orbital myofibroma. Fibrous tissue proliferations in the orbit, including myofibroma, are rare and are best understood based on their age of onset and associated systemic findings.
Fibrous proliferations of childhood can be classified into 2 groups based on clinical and histopathologic features. The first group includes those tumors that also occur in adults, such as nodular fasciitis, palmar and plantar fibromatosis, desmoid tumor, abdominal and extra-abdominal fibromatosis, and juvenile hyaline fibromatosis. The second group includes tumors found exclusively in children with no adult counterpart, such as fibrous hamartoma of infancy, infantile digital fibromatosis, fibromatosis coli, infantile (desmoid) fibromatosis, and infantile myofibromatosis. Tumors in this latter group are often initially suspected clinically to be malignant based on their rapid growth and histopathologic features.

Infantile myofibromatosis was initially recognized by Stout in 1954. The disease has been referred to under several synonyms including “congenital multiple fibromatosis,” “multiple mesenchymal hamartomas,” “multiple vascular leiomyomas of newborn,” and others. Most tumors are recognized at birth or before age 2 years. The tumor is unifocal in 74% and multifocal in 26% of cases. Multifocal tumors can show autosomal dominant transmission. Tumor recurrence is uncommon and death from myofibromatosis occurs in 7% of patients, usually those with multicentric visceral tumors causing cardiac or gastrointestinal tract compromise.

Myofibromatosis of infancy occurs in the head and neck region in 36%, trunk in 33%, upper extremity in 13%, and lower extremity in 18% of cases. Ocular involvement is rare, generally limited to the eyelids and orbit. Massive proptosis in a newborn from a solitary myofibroma involving the sinuses, brain, and medial orbit has been reported. The nonresected mass remained stable and became less apparent as the facial features grew at their normal rate. Others have described less advanced infantile myofibromas involving the eyelid and lacrimal apparatus that were managed with resection and left little cosmetic deformity. Multifocal systemic myofibromatosis with orbital involvement has been recognized.

Our patient had an intraosseous myofibroma. Isolated orbital bone involvement with myofibroma has been recognized in the zygomatic bone. In our case, computed tomography showed prominent bone destruction and the margins of the intraosseous mass were indistinct, prompting suspicion for lesions such as fibrous dysplasia, Langerhans cell histiocytosis, rhabdomyosarcoma, metastatic neuroblastoma, and primary bone tumors. The magnetic resonance features of this tumor were quite striking. The tumor showed crisp margination within bone, bowed the cortical rim of bone outward, and demonstrated marked enhancement with gadolinium.

Pathologically, both visceral and intraosseous myofibroma display similar features. The firm, light pink to gray tissue exhibits vague nodularity produced by areas containing relatively abundant extracellular matrix. The cells are typically spindled and may show a fascicular growth pattern resembling smooth muscle. Immunohistochemical staining is usually positive for vimentin and actin. Less typical lesions may demonstrate highly cellular growth, with occasional necrosis and prominent vascularity, mimicking sarcoma.

The differential diagnosis of these lesions includes juvenile hyaline fibromatosis, a lesion with similar microscopic findings. However, juvenile hyaline fibromatosis typically occurs after age 2 years and is locally aggressive. Infantile myofibromatosis, in contrast, occurs at a younger age and is generally localized. Other lesions of bone that may be considered in the differential diagnosis include fibrous dysplasia, nonossifying fibroma, fibrosarcoma, neurofibroma, and nodular fascitis. However, each is distinguished from myofibroma based on histological findings. Fibrous dysplasia has characteristic bone formation. Nonossifying fibroma displays a storiform growth pattern admixed with xanthoma cells and giant cells. Fibrosarcoma has a herringbone growth pattern, often with nuclear atypia. Neurofibroma has spindled cells with wavy nuclei arranged in short fascicles and are positive for S100 protein. Nodular fasciitis contains plump fibroblasts in a loose stroma with scattered inflammatory cells.

The management of infantile myofibroma involves conservative resection to excise the mass without affecting vital structures. In some instances, the mass will regress spontaneously. Recurrence can develop but has not yet been found with orbital myofibroma. Systemic evaluation for multicentric myofibromatosis is advised. Infants with the solitary unifocal form have an excellent prognosis, whereas those with the multifocal form should be followed up for visceral compromise.

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