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Primary Intraosseous Hemangioma of the Orbit: CT and MR Appearance

Christopher Sweet, Richard Silbergleit, and Bharat Mehta

Summary: An 8-year-old girl had acute onset of blurred vision, proptosis, and lateral and vertical gaze palsies on the right. CT and MR examinations showed an expansile mass involving the greater wing of the right sphenoid bone. Pathologic analysis of the surgical specimen revealed a capillary hemangioma. The lesion was inhomogeneous but predominantly isointense with gray matter on T1-weighted images. On T2-weighted images the lesion was inhomogeneous with areas of both high and low signal intensity. The rim enhanced uniformly, and there was inhomogeneous enhancement of the bulk of the lesion.

Index terms: Hemangioma; Orbits, neoplasms

Primary intraosseous hemangioma is an uncommon tumor of bone. These benign neoplasms tend to involve the vertebrae and skull. Orbital lesions occur infrequently. We present a case of a cellular capillary hemangioma arising from the greater wing of the right sphenoid bone, emphasizing the computed tomographic (CT) and magnetic resonance (MR) imaging findings.

Case Report

A previously healthy 8-year-old girl had blurred vision and excessive tearing on the right side. An initial diagnosis of acute conjunctivitis was made and the patient was sent home on antibiotics. The next day, the symptoms worsened and included diplopia, frontal headache, and pain over the lateral aspect of the right orbit. Examination by an ophthalmologist disclosed lateral and upward gaze palsies on the right, slight proptosis of the right globe, extrinsic compression of the posterolateral aspect of the right globe, and normal visual acuity. A CT examination of the orbits showed a well-defined lesion centered in the greater wing of the sphenoid on the right with extension into the lateral extraconal space of the right orbit and anteroinferior aspect of the right middle cranial fossa. The lesion extended laterally, displacing the temporalis muscle. The rim enhanced intensely. The central portion of the lesion en-

hanced mildly and inhomogeneously. The underlying brain parenchyma did not appear to be involved (Fig 1A and B).

An MR examination of the brain and orbits was performed for further characterization of the lesion. Axial T1-weighted (483/17/2 [repetition time/echo time/excitations]), axial T2-weighted (2100/90/1), and coronal contrast-enhanced T1-weighted (567/17/2) images were obtained. The study confirmed a well-defined expansile mass within the greater wing of the sphenoid bone on the right (Fig 1D-F). On T1-weighted images the lesion was inhomogeneous. The bulk of the lesion was isointense with gray matter, with foci of both increased and decreased signal intensity scattered throughout the lesion. On T2-weighted images there was markedly inhomogeneous signal intensity suggestive of blood flow and/or blood degradation products. The lesion enhanced inhomogeneously after contrast administration, with the rim enhancing intensely. The mass extended into the right posterolateral extraconal space causing a mild proptosis and deformity of the posterolateral portion of the globe with medial displacement of the optic nerve. The mass extended laterally into the temporal fossa and posteriorly into the anteroinferior aspect of the right middle cranial fossa.

A definitive surgical resection was performed. A bicoronal incision allowed dissection of the lesion from the undersurface of the right temporalis muscle. The orbital and middle cranial fossa portion of the tumor was dissected. The tumor was completely extradural and a portion of the dura was resected to ensure clear margins. There was no invasion of the underlying right temporal lobe. Pathologic analysis showed a vascular neoplasm characterized by medium to large vascular spaces intermixed with areas of cellular granulation tissue, typical of capillary hemangioma (Fig 1G). The vascular nature of the neoplasm was confirmed by immunoreactivity of the cells with CD34, factor 8, and vimentin. The patient made a full recovery with complete resolution of the gaze palsies. Follow-up examination performed 8 months after discharge revealed no recurrence of the tumor, and she has remained symptom free.

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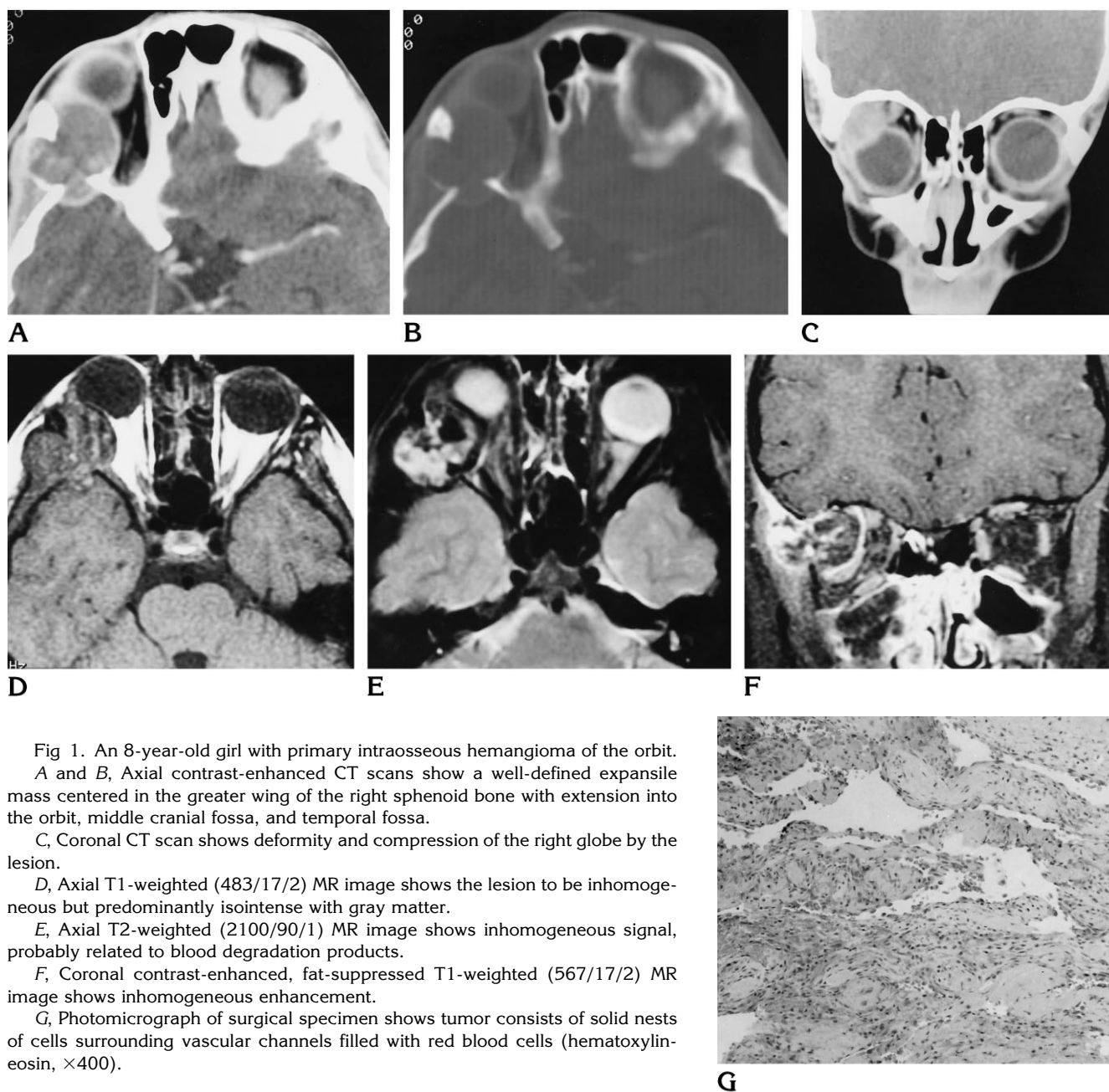


Fig 1. An 8-year-old girl with primary intraosseous hemangioma of the orbit.

A and B, Axial contrast-enhanced CT scans show a well-defined expansile mass centered in the greater wing of the right sphenoid bone with extension into the orbit, middle cranial fossa, and temporal fossa.

C, Coronal CT scan shows deformity and compression of the right globe by the lesion.

D, Axial T1-weighted (483/17/2) MR image shows the lesion to be inhomogeneous but predominantly isointense with gray matter.

E, Axial T2-weighted (2100/90/1) MR image shows inhomogeneous signal, probably related to blood degradation products.

F, Coronal contrast-enhanced, fat-suppressed T1-weighted (567/17/2) MR image shows inhomogeneous enhancement.

G, Photomicrograph of surgical specimen shows tumor consists of solid nests of cells surrounding vascular channels filled with red blood cells (hematoxylin-eosin, $\times 400$).

Discussion

Primary intraosseous hemangiomas are rare, benign, slow-growing neoplasms. More than 50% are found in the vertebra or skull (1). When they arise within the calvaria, they are normally confined to the frontal or parietal bones. These lesions are usually solitary and occur more frequently in females than males, in a ratio of three to one (2). They are typically found in adults, although persons of any age may be affected (3). Osseous hemangiomas are of two histo-

logic types: cavernous or capillary. The cavernous type usually involves the skull (4). The pathogenesis of these neoplasms is unknown; however, the cause is believed to be either congenital or related to previous trauma (5).

The characteristic calvarial radiographic pattern of a sunburst of radiating trabecula with or without a thin peripheral sclerotic rim was not present in our case. This pattern has been described in a pathologically proved case of orbital cavernous hemangioma (6). Moreover, the

characteristic high signal intensity on T1-weighted images seen in hemangiomas of the vertebral bodies was absent in our case. This signal intensity in vertebral hemangiomas reflects the fatty content of these lesions (7); thus, the lack of high signal intensity in our case may reflect a relatively low fat content of the lesion. The characteristic high signal intensity seen on T2-weighted images was present in large portions of the tumor in our case. The exact cause of this high signal intensity is uncertain (7). Cavernous and capillary hemangiomas have similar imaging features and are differentiated by their histopathologic appearance (1, 8). Angiography was not performed in this case, but it has been reported to show increased vascularity (9). The clinical presentation of orbital hemangiomas is nonspecific. Patients may present with an asymptomatic lump, whose expansile nature may lead to proptosis, diplopia, optic atrophy, and ptosis (2). The preferred treatment is complete surgical resection. Radiation therapy is reserved for subtotal resection or for an unresectable lesion in symptomatic patients.

A diverse spectrum of benign and malignant neoplasms affect the orbit in children. The differential diagnosis of an expansile lesion in the pediatric osseous orbit includes aneurysmal bone cyst, giant cell tumor, fibrous dysplasia, Langerhans cell histiocytosis, sarcoma (particularly rhabdomyosarcoma), metastatic disease, hematic cyst, or a dermoid (10). In a review of

the literature, we found 21 reports describing intraosseous hemangioma involving the orbit (11). Our case has some unusual salient features, such as the rapid onset of symptoms, sphenoidal location, age at presentation, capillary type of hemangioma, and atypical imaging features of a skull hemangioma. Intraosseous hemangioma should be included in the differential diagnosis of expansile osseous orbital lesions.

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