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AJNR Am J Neuroradiol 1993, 14 (2) 473-474
http://www.ajnr.org/content/14/2/473

This information is current as of October 26, 2023.
MR Appearance of an Orbital Leiomyoma

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Summary: The authors describe the MR appearance of an intraconal orbital vascular leiomyoma that probably arose from smooth muscle in the wall of a vein. Cavernous hemangiomas, schwannomas, neurofibromas, and other well-encapsulated masses can have a similar appearance.

Index terms: Orbits, neoplasms; Orbits, magnetic resonance

Leiomyoma is a rare neoplasm of the orbit with only 16 reported cases in the world literature (1–8); none have been reported in the imaging literature to our knowledge. This report describes the magnetic resonance (MR) appearance of an intraconal leiomyoma, an entity that should be included in the differential diagnosis of well-defined orbital masses.

Case Report

A 57-year-old white man presented with the gradual onset of painless left proptosis. He denied any decreased vision or associated diplopia. On physical examination, the proptosis was apparent, but there was no tenderness or palpable mass. Vision was 20/20 OD with normal fields by Goldman perimetry. The extraocular muscles were intact and the fundus was normal. MR revealed a well-circumscribed, round 12-mm intraconal mass displacing the optic nerve superiorly and causing mild proptosis. It was isointense to gray matter on T1-weighted and became hyperintense on both proton density- and T2-weighted images (Figs. 1A–1C). At surgery, a firm, encapsulated intraconal mass was found that was adherent to the optic nerve above and was tethered by a fibrous band below. Histologic examination demonstrated smooth mus-

Received March 10, 1992; revision requested May 3; revision received June 4 and accepted July 21.

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AJNR 14:473–474, Mar/Apr 1993 0195-6108/93/1402-0473 © American Society of Neuroradiology

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cle cells and numerous interspersed thick-walled vessels and vascular channels surrounded by an inflammatory pseudocapsule (Fig. 1D). The findings were compatible with a vascular leiomyoma (angiomyoma) that probably arose from smooth muscle cells residing in the wall of a vein. The patient had an uneventful postoperative course with resolution of the proptosis.

Discussion

Leiomyoma is a very rare neoplasm in the orbit. Of cases reported since 1963, the vast majority occur in patients under the age of 40. There is no sex predilection. Patients usually present with gradual onset of proptosis and/or diplopia. Leiomyomas may occur either intra- or extraconally, with a slight preference for the former.

The tumor consists of bundles of spindle-shaped smooth muscle cells interspersed in abundant endothelial sinusoids or dilated capillaries (4). There is actually a spectrum of histologies based on the relative proportion of vascular and smooth muscle elements. If the tumor is primarily vascular with smooth muscle cells interspersed in the interstitial spaces, it may be called an angiomyloma or hemangioleiomyoma (5, 6). Alternatively, a predominantly solid smooth muscle tumor without conspicuous vascular components is similar to leiomyomas found elsewhere in the body (7). The latter is unusual with the vast majority of tumors being very vascular. Proceeding even further on the vascular end of the scale, the tumor becomes a cavernous hemangioma. All of these tumors have a very well-defined fibrous capsule. Occasionally there are satellite lobulations (8) and, therefore, a wide excision is required to insure complete removal.

The cell of origin is presumably the vascular smooth muscle cell (especially those related to small veins) or possibly the pericyte (4). Other possibilities for the source of smooth muscle cells in the orbit include: 1) rests within the connective tissue trabeculae of the orbital fat; 2) vestigial smooth muscle spanning the inferior orbital fissure; or (3) the muscles of Muller associated with tarsi and lids (9). However, the conspicuous vascular pattern strongly supports the notion that most of these tumors arise from vascular smooth muscle (2, 8).

The MR signal characteristics in this case are like those of neoplasm in general; that is, isointense to low intensity on short TR images, and high intensity on long. The appearance is identical to the more common cavernous hemangioma (10). Although gadolinium was not given in this case, a vascular leiomyoma would be expected to enhance intensely, similar to cavernous hemangiomas. A tumor with densely packed smooth muscle cells and relatively sparse vasculature would probably display signal characteristics similar to leiomyomas found in the uterus (11), ie, lower signal on T2 imaging. It should be noted that these assumptions about enhancement and signal characteristics are speculative and await further research.

In conclusion, leiomyoma is a well-circumscribed, benign orbital mass that is usually intraconal and probably arises from vascular smooth muscle. The MR appearance is similar to other well-encapsulated masses such as cavernous hemangiomas, neurofibroma, schwannoma, fibrous histiocytoma, and hemangioleiomyoma. Although rare, leiomyoma should be considered in this differential.

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