

Orbital leiomyoma¹

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Abstract

Orbital leiomyoma is a rare, benign smooth muscle tumor of the orbit occurring intraconally or extraconally. The tumor affects males twice as frequently as females with a mean age of 30 years. Clinically, patients with orbital leiomyomas located deep in the orbit present with painless proptosis or displacement of the globe progressing slowly over several months or years. Leiomyomas located in the anterior orbit and eyelids may present with progressive painless swelling of the eyelids. On MRI, orbital leiomyoma is isointense with respect to the extraocular muscle and cerebral gray matter on T1-weighted images and hyperintense on T2-weighted images. Diagnosis is based on histopathologic findings and pattern of immunoreactivity. The tumor is composed of spindle cells arranged in a fibrous stroma rich in dilated sinusoidal capillaries. The nuclei of tumor cells are oval with blunted ends and there are no mitotic figures. Immunohistochemically, the tumor stains positive with smooth muscle actin, desmin, and vimentin. Total tumor excision is the best treatment option. Recurrence after incomplete excision is possible but malignant transformation has not been reported in the absence of high dose external beam radiotherapy.

Keywords

Orbital leiomyoma, spindle cell tumors, proptosis; immunohistochemistry; histopathology, smooth muscle actin, tumor excision

Disease name and synonyms

- Orbital leiomyoma
- Mesectodermal leiomyoma

Definition

Orbital leiomyoma is a rare, benign smooth muscle tumor of the orbit occurring intraconally or extraconally

Differential diagnosis

Orbital leiomyoma is indistinguishable from other orbital tumors on orbital computed tomography (CT) and magnetic resonance imaging (MRI).

The pathologic differential diagnosis of orbital leiomyoma includes other benign spindle cell tumors of the orbit, including schwannoma, neurofibroma, fibrous histiocytoma, and solitary fibrous tumor of the orbit, as well as malignant spindle cell tumors, such as leiomyosarcoma, [fibrosarcoma](#), and amelanotic spindle cell malignant melanoma.

The differentiation of leiomyoma from malignant spindle cell tumors is made based on the absence of mitotic figures and cellular morphology (Folberg *et al*, 1983; Jakobiec *et al*, 1975; Wojno *et al*, 1983).

Positive immunoreactivity with smooth muscle actin (SMA), a marker expressed only in smooth muscle tumors, and negative immunoreactivity for myoglobin, neuron specific enolase (NSE), Leu-7, CD-34, S-100, and HMB-45 rules out other spindle cell tumors.

Pathogenesis

Orbital leiomyoma is believed to arise from the smooth muscle of orbital blood vessels, pericytes, Müller's muscle, or capsulopalpebral muscle of Hesser (Betharia *et al*, 1991; Jakobiec *et al*, 1973; Nath and Shukla, 1963). Tumors located in the anterior orbit close to the eyelids may develop from the capsulopalpebral muscle of Hesser (Nath and Shukla, 1963). The pericyte is a possible cell of origin for orbital leiomyoma. Transmission electron microscopic studies have shown that the pericyte undergoes several evolutionary stages, ranging from poorly differentiated mesenchymal cells of capillary walls to more highly differentiated smooth muscle cells of venular walls (Jakobiec *et al*, 1973).

Clinical description

Clinically, patients with orbital leiomyomas located deep in the orbit present with painless proptosis or displacement of the globe progressing slowly over several months or years. Leiomyomas located in the anterior orbit and eyelids may present with progressive painless swelling of the eyelids (Betharia *et al*, 1991, Jakobiec *et al*, 1975, Jolly *et al*, 1995; Neetens and Smet, 1984).

The case of a 10-year-old girl presenting with a 1-year history of progressive proptosis in the left eye has been recently reported (Gündüz *et al*, 2004). The patient had 7 mm proptosis in the left eye, which was pushed medially and inferiorly. The motility of the left eye was restricted in all positions of gaze, especially superiorly and temporally. Visual acuity in the left eye was 20/20. Slit-lamp biomicroscopy revealed normal

findings. On funduscopy, an elevated choroidal mass in the superotemporal quadrant arising from compression of the globe by the orbital tumor was noted. The right eye was normal.

Computed tomography (CT) and MRI usually demonstrate a well-circumscribed orbital tumor. On MRI, orbital leiomyoma is isointense with respect to the extraocular muscle and cerebral gray matter on T1-weighted images and hyperintense on T2-weighted images. Orbital leiomyoma shows moderate contrast enhancement.

Orbital leiomyoma has been reported to occur in the anterior orbit infiltrating the eyelids, in the intraconal orbit, and in the apical orbit. Orbital leiomyoma with orbital apex involvement may demonstrate intracranial involvement (Jakobiec *et al*, 1975, Kulkarni *et al*, 2000). In the case recently reported by Gündüz *et al* (2004), the tumor occurred in a deep extraconal position in the orbit, unlike the previous extraconal leiomyomas that have been reported to occur in the anterior orbit (Betharia *et al*, 1991, Henderson and Harrison, 1970, Jakobiec *et al*, 1975, Jolly *et al*, 1995, Neetens and Smet, 1984; Sanborn *et al*, 1979).

Morphological features

Grossly, the tumor has a solid lobulated red-tan appearance. The tumor consists of bundles of spindle-shaped cells with oval nuclei arranged in a fibrous stroma with dilated sinusoidal capillaries (Gündüz *et al*, 2004). Sometimes the leiomyoma can present as a predominantly solid smooth muscle tumor (Nath and Shukla, 1963) that does not feature a conspicuous vascular component. On the other hand, leiomyoma can present as a predominantly vascular tumor with smooth muscle cells interspersed in the interstitial spaces (Henderson and Harrison, 1970; Wolter, 1965). Such tumors are described as hemangioliomyomas.

The nuclei of tumor cells in leiomyoma have blunted ends (Gündüz *et al*, 2004). This is an important feature of leiomyoma that is not usually seen with other benign spindle cell tumors (Betharia *et al*, 1991; Sanborn *et al*, 1979). There are no or few mitotic figures. Tumor cells display intense fuchsinophilia on Masson trichrome stain.

On transmission electron microscopy, leiomyoma has characteristic features, such as intracytoplasmic bundles of parallel filaments, cytoplasmic fusiform densities, and a rod-shaped nucleus (Jakobiec *et al*, 1973, Jakobiec *et al*, 1975, Jolly *et al*, 1995, Sanborn *et al*, 1979; Shields *et al*, 1994). These features are useful in differentiating leiomyoma from other spindle cell tumors. Immunohistochemically, orbital leiomyoma has been reported to demonstrate positive immunoreactivity with

desmin, vimentin, muscle specific actin, and SMA (Badoza *et al*, 1999; Jolly *et al*, 1995). Negative immunoreactivity for myoglobin, NSE, Leu-7, CD-34, S-100, and HMB-45 equivocally rules out other spindle cell tumors that are considered in the differential diagnosis, as discussed above.

Diagnostic methods

Diagnosis is based on histopathologic findings and pattern of immunoreactivity.

Epidemiology

Orbital leiomyoma is an uncommon tumor. Sixteen well-documented cases of orbital leiomyoma have been published over the past 40 years (Badoza *et al*, 1999; Betharia *et al*, 1991; Carrier *et al*, 1993; Gündüz *et al*, 2004; Henderson and Harrison, 1970; Jakobiec *et al*, 1973; Jakobiec *et al*, 1975; Jolly *et al*, 1995; Kulkarni *et al*, 2000; Nath and Shukla, 1963; Neetens and Smet, 1984, Saga *et al*, 1982; Sanborn *et al*, 1979; Vigstrup and Glenthoj, 1982; Wolter, 1965). The age of the patients ranged from 9 years to 57 years, the mean age being 30 years. Of the 16 patients with orbital leiomyoma, 11 were males, 4 were females, and the sex of the patient was not reported in one case. Orbital leiomyoma is more common in males. The reason for this male preponderance is not clear.

Treatment

The best treatment of orbital leiomyoma is complete excision of the tumor. Jakobiec and associates pointed out the presence of satellite nodules around the tumor, which should be removed to prevent recurrence (Jakobiec *et al*, 1975). If the tumor is fragmented during removal as in our case, all the fragments should be excised. Whenever complete excision is not possible especially for tumors near the orbital apex, the surgeon should consider leaving a small residual behind to avoid damage to the vital structures. Recurrence after incomplete excision has been observed (Jakobiec *et al*, 1973; Sanborn *et al*, 1979) but spontaneous malignant transformation does not occur in a recurrent tumor. In the case reported by Jakobiec, it is possible that the high dose external beam radiotherapy (114 Gy) administered might have contributed to the development of sarcomatous transformation of the recurrent leiomyoma (Jakobiec *et al*, 1975). This is the only case with a recorded death causally related to orbital leiomyoma. Usually, the life prognosis is excellent for completely excised and recurrent orbital leiomyomas. The same is also true for orbital leiomyoma with intracranial extension (Kulkarni *et al*, 2000).

Orbital leiomyoma is not radiosensitive (Jakobiec *et al*, 1973; Jakobiec *et al*, 1975). External beam radiotherapy has no role in the treatment of the residual tumor after incomplete excision. If a subtotal tumor excision is made, the patient should be followed with serial neuroimaging studies for signs of tumor recurrence.

In the case reported by Saga and associates, incomplete excision of the orbital leiomyoma was followed by idiopathic orbital inflammation, which responded to a course of oral steroids (Saga *et al*, 1982). The reason for the occurrence of idiopathic orbital inflammation after incomplete excision of the orbital leiomyoma is not clear. In other cases that were incompletely excised at initial surgery, (Jakobiec *et al*, 1973, Jakobiec *et al*, 1975 and Sanborn *et al*, 1979) orbital inflammation did not develop.

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