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Anterior orbital leiomyoma originating from the supraorbital neurovascular bundle

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Abstract

Purpose: To present a young female patient with left anterior orbital leiomyoma that originates from the supraorbital neurovascular bundle.

Case presentation: A 41-year-old female patient was admitted to our clinic with a complaint of swelling of the left upper eyelid. Based on the ophthalmological and imaging assessments, the excisional biopsy with the preliminary diagnosis of dermoid cyst was planned. The histopathological and immunohistochemical examinations of excised sample revealed surprisingly that the tumour was a leiomyoma. No recurrence was detected in the patient's follow-up.

Conclusion: Although it is rare, orbital leiomyoma should be considered in the differential diagnosis of patients with orbital tumour.

Keywords

Orbital leiomyoma, orbital tumour

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Introduction

Leiomyoma is a rare and benign smooth muscle tumour of the orbit. Although the origin of leiomyoma in orbit has been a matter of speculation, it is most probably developed from smooth muscle cells and pericytes in vascular wall. Additionally, however, the capsulopalpebral muscle of Hessar and Muller muscle may be its origin.[1–3] Herein, we present a case of anterior orbital leiomyoma. As the inferomedial part of the tumour is bonded to supraorbital neurovascular bundle in our subject, we suggest that the tumour is caused by the smooth muscle cells in the supraorbital neurovascular bundle. The study was conducted in accordance with the tenets of the Declaration of Helsinki by obtaining a written consent from the patient.

Case report

A 41-year-old female patient was admitted to our clinic with a complaint of swelling of the left upper eyelid for 6 months. Visual acuity was 10/10 in the right eye. Best corrected visual acuity was 7/10 with the refractive errors of +3.25D spherical and −3.0D astigmatism in the left eye with Snellen card. Intraocular pressure was 14 mm Hg bilaterally. On palpation, a firm and immobile mass of approximately 10×10 mm^2 was detected in the midline of her left upper eyelid under eyebrow. Hertel exophthalmometry readings detected no proptosis. Bilateral ocular motilities were not limited in all directions of gaze. The slit lamp biomicroscopic and dilated fundus examinations were unremarkable in her both eyes. The patient's history and general physical examination were normal. Magnetic resonance imaging (MRI) revealed a well-circumscribed − 9×9 mm^2 in size – solid mass keeping intensive contrast in the left frontobasal, subgaleal area (Figure 1(a) and (b)). The tumour was located extraconally in the anterior orbit. Although there was a connection between cortical frontal osseous and solid mass, there was no bone destruction in MRI. In the light of these

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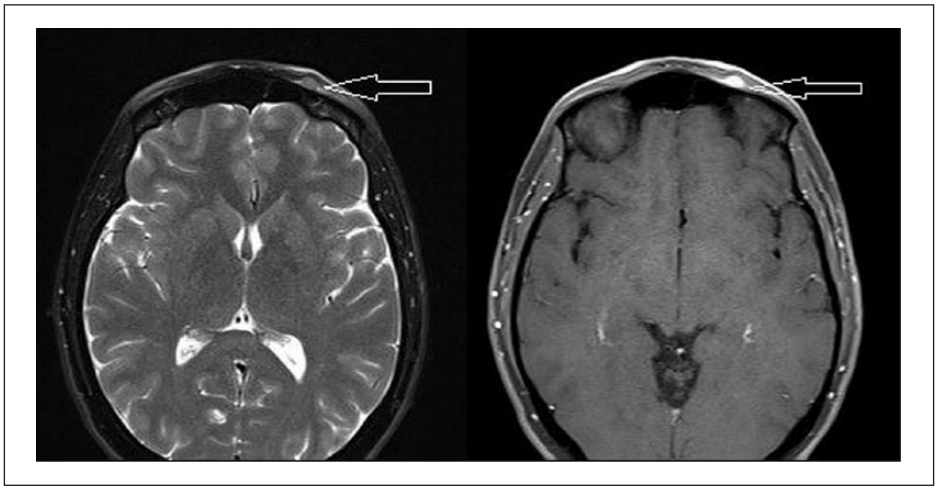

Figure 1. Preoperative MRI revealed solid mass keeping intensive contrast in the nasal part of superior orbital rim (arrows): (a) axial MRI without contrast and (b) axial MRI with contrast.

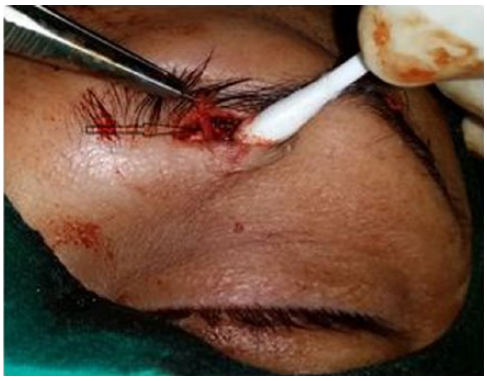

Figure 2. After tumour was removed completely, supraorbital nerve is seen (arrow).

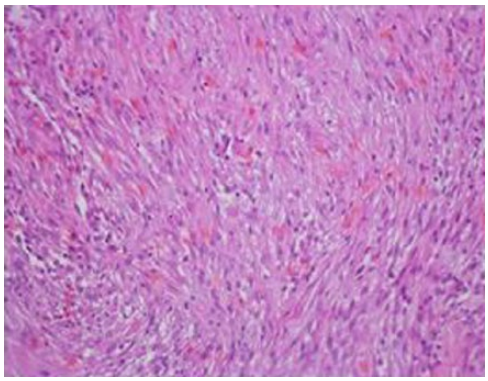

Figure 3. Haematoxylin and eosin staining revealed that bundle of uniform, spindle-shaped cells were arranged in whorls.

findings, our preliminary diagnosis was dermoid cyst. An excisional biopsy was performed via the left transcutaneous anterior orbitotomy. Under eyebrow, horizontal skin incision was made. The mass was dissected from the surrounding tissue by blunt dissection. Because the tumour was attached to surrounding tissue, it was broken into pieces and removed completely. As the bottom portion of the mass adhered to the periosteum, it was excised together with its periosteum. Additionally, because the inferomedial part of the mass abutted on the supraorbital neurovascular bundle, it was excised together with it and caused bleeding (Figure 2). But it could be controlled by cautery. Excised tumour was lobular form with yellow-grey colour, and it was set for histopathological examination and immunohistochemical investigation. Histopathological examination revealed diffuse infiltration of uniform, spindle-shaped cells arranged in whorls. The nuclei of the tumour cells were elongated cigar-shaped with blunted ends. There were no mitotic figures, nuclear pleomorphism and nuclear atypia (Figure 3).[2,4] Immunohistochemical staining was positive for smooth muscle actin (SMA) and h-caldesmon, but negative for desmin and S100 protein. CD31 and CD34 immunostaining showed sparse accompanying thin-walled vascular structures. Ki67 index was 1% (Figure 4). According to histopathological and immunohistochemical findings, our diagnosis was orbital leiomyoma. In the postoperative examination, there was anaesthesia on the frontal area of our patient, due to excision of supraorbital neurovascular bundle preoperatively. No complication was found except anaesthesia, and no recurrence was detected in 18 months of follow-up.

Discussion

Leiomyoma is a benign, slow-growing tumour arising from smooth muscle cells. Although they are most commonly seen in uterus, soft tissues and gastrointestinal tracts, they may also develop from each tissue with the smooth muscle cells. Leiomyoma occurs most frequently in the ciliary body and iris of eye compared with the orbit.[5]

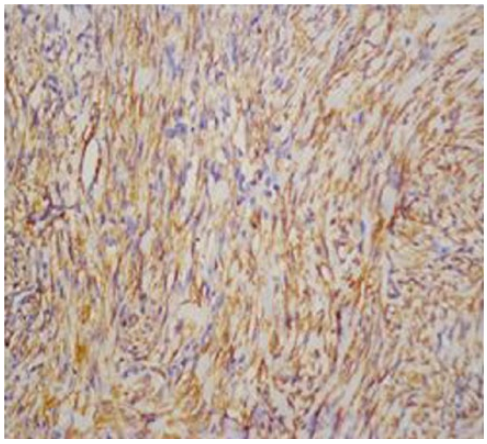

Figure 4. Immunohistochemical staining showed the diffuse staining with SMA.

Orbital leiomyoma is an extremely rare neoplasm. It was first described in 1896 by Lodato.[6] The majority of patients with orbital leiomyoma are young adults with male preponderance.[7] Orbital leiomyoma can occur as an extraconal or intraconal tumour. It would prefer anterior orbit rather than posterior orbit.[1,2,4,8] Our patient was a 41-year-old female with anterior orbital leiomyoma located extraconally. Although orbital leiomyomas commonly induce painless and slowly progressive proptosis and ocular motility disability, visual acuity and visual field are usually unaffected.[5,8] The cause of low vision in left eye of our patient was amblyopia.

As the leiomyomas rarely settle in orbita and they share similar histopathological features with the other spindle cell tumours of the orbit, including neurogenic tumours such as schwannoma, neurofibroma, fibrous histiocytoma and solitary fibrous tumour, the diagnosis of orbital leiomyoma is difficult histopathologically. As in our case, the fact that the nuclei of the tumour cells in leiomyoma have blunted ends is an important feature of the differential diagnosis of leiomyoma from the other spindle-cell tumours of orbit.[1] Additionally, the absence of mitotic figure and cellular atypia of the nuclei contribute to distinguish it from the malign spindle cell tumours.[4] Transmission electron microscopy and/or immunohistochemistry should be used for definitive diagnosis. As the immunohistochemistry is cheap, easy and faster, it is more preferred than transmission electron microscopy.[1,5] Although orbital leiomyoma typically shows positive immunoreactivity with SMA, desmin and vimentin, SMA is the most specific to leiomyoma. In our case, immunohistochemistry was performed to confirm the diagnosis, and the tumour showed positive immunoreactivity with SMA (Figure 4).

The definitive treatment for orbital leiomyoma is a complete surgical extraction. If you do incomplete surgical removal, recurrence may be seen rarely.[3,8]

The origin of orbital leiomyoma is a confusing issue. Although Nath and Shukla[9] suggested that the most likely source of histopathogenesis of orbital leiomyoma is the capsulopalpebral fascia of Hessar, it is believed more today that the origin of orbital leiomyoma is of a vascular source. Leiomyomas can be grouped according to the vascular component of the tumour.[10–13] Angioleiomyoma, angiomyoma and angiomyofibroma are the vascular variants of leiomyoma. And this supports the idea that the origin of leiomyoma is smooth muscle cells on vascular walls or pericyte.[2] Additionally, it is believed that the origin of leiomyoma is pericyte because it has a large number of evolutionary stages ranging from poorly differentiated mesenchymal cells of capillary walls to more highly differentiated smooth muscle cells of venular walls.[2] As the inferomedial part of the tumour is bonded to supraorbital neurovascular bundle in our subject, we suggest that the tumour is caused by the smooth muscle cells in the supraorbital neurovascular bundle. Until today, few cases of orbital leiomyoma have been reported in the literature.[14] Additionally, as far as we know, there is no case report related to orbital leiomyoma originating from the supraorbital neurovascular bundle. This is the point that makes our case interesting.

Conclusion

Although it is rare, orbital leiomyoma should be considered in the differential diagnosis of patients with orbital tumour. The localization and neighbourhoods of leiomyoma in orbit may be important for the course of the surgical procedure.

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Ethical approval

Our institution does not require ethical approval for reporting individual cases or case series.

Informed consent

Written informed consent was obtained from the patient(s) for their anonymized information to be published in this article. Written informed consent was obtained from the patient.

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