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Case Report

Intramuscular Hemangioma of Lateral Rectus Muscle with Rare Presentation as an Epibulbar Mass: A Case Report and Review of Literature

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Keywords
Intramuscular hemangioma · Extraocular muscle · Differential diagnosis · Histopathology · Case report

Abstract
Introduction: Intramuscular hemangiomas of extraocular muscles are extremely rare tumors that usually present as retro-orbital masses causing proptosis. We describe a previously unreported presentation, in the form of an epibulbar mass; this easily accessible location allows direct imaging, complete surgical resection, and histopathological confirmation, providing a unique perspective. ***Case Presentation:*** A 69-year-old woman presented with a painless dark red mass in the lateral part of the right eye, which had been slowly enlarging over the last 18 months. Clinical features and imaging were suggestive of a benign vascular tumor of the conjunctiva. During surgical resection, the mass was observed to be enmeshed within the fibers of the lateral rectus muscle. Careful dissection from muscle fibers was needed for complete excision. Histopathology revealed the diagnosis of an intramuscular hemangioma of extraocular muscle. ***Conclusion:*** In this report, we describe the atypical anterior epibulbar presentation of intramuscular hemangioma of the lateral rectus muscle. We discuss the differential diagnoses and management of this rare tumor along with a review of existing literature. Careful surgical resection achieved complete resolution in this case without recurrence.

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Introduction

Intramuscular hemangiomas (IMHs) are benign vascular tumors that comprise less than 1% of all hemangiomas [1]. Of these, only 15% occur in the head and neck muscles, the commonest muscle involved being the masseter [1, 2]. Extraocular muscle (EOM) involvement is especially rare with only a few cases reported in literature [1–10]. Patients with IMH of EOM commonly present with proptosis due to intra-orbital tumor location, and diagnosis is mainly based on orbital MRI findings with histopathological confirmation. Herein, we describe a case of an isolated IMH of the lateral rectus muscle with a unique presentation in the form of an anterior epibulbar mass.

Case Presentation

A 69-year-old lady presented with a painless mass in her right eye that had slowly enlarged over the past 1.5 years. Her past medical history was significant for uterine cancer, treated surgically 3 years ago, without any recurrence since. The best corrected visual acuity was 20/25 bilaterally, and the intraocular pressures were normal. Slit lamp examination revealed a dark red lobulated epibulbar lesion, on the temporal side of the right eye, measuring 7 mm × 5 mm (Fig. 1a). It was not compressible, pulsatile, or reducible and did not increase in size with Valsalva maneuvers. While the conjunctiva over the lesion was freely mobile, the mass itself had limited mobility, especially along the horizontal meridian. There was no proptosis, eyes were orthophoric, and ocular movements were full in all directions. The rest of the anterior and posterior segments were normal in both eyes. Clinical findings were consistent with the provisional diagnosis of a benign vascular tumor. Anterior segment OCT (AS-OCT) over the lesion revealed multiple intralesional hypo-reflective spaces filled with moderate to high reflective material, causing back-shadowing, corresponding to blood-filled vascular lumens (Fig. 1b, c). Due to concerns about growth and cosmesis, an excisional biopsy was undertaken. Intraoperatively, the mass was found to be enclosed within the fibers of the lateral rectus muscle. Bridle suture was passed under the muscle to stretch it, and complete excision of the tumor was done after careful dissection from the muscle fibers (Fig. 2). Histopathology revealed a well-circumscribed proliferation of dilated, blood-filled venous spaces lined by flattened endothelial cells and separated by fibroconnective tissue of varying thickness, confirming the diagnosis of a cavernous IMH of an EOM (Fig. 3). Post-operative recovery was uneventful and without any recurrence over 2 years of follow-up.

Discussion

Isolated IMHs of EOMs are extremely rare, benign vascular tumors. They are congenital, slow-growing masses that often remain undetected until later decades when they become symptomatic [2]. To date, only eleven cases of hemangiomas involving EOMs have been reported in literature [1–10]. The commonest clinical presentation reported was painless, progressive proptosis (seen in 10 cases); other symptoms included eyelid swelling (5 cases), ocular movement restriction (3 cases), and diplopia (1 case) [1–10]. The location of the IMH can lead to further complications. Visual acuity may be decreased in cases where the tumor encases the optic nerve, causing mechanical compression [3, 5, 8]. In one reported case, the tumor even caused bony erosion of the orbital roof with intracranial extension [10].

The diagnosis of an IMH of the EOM is challenging in the absence of a visible mass; it is based mainly on radiological features with histopathological confirmation where

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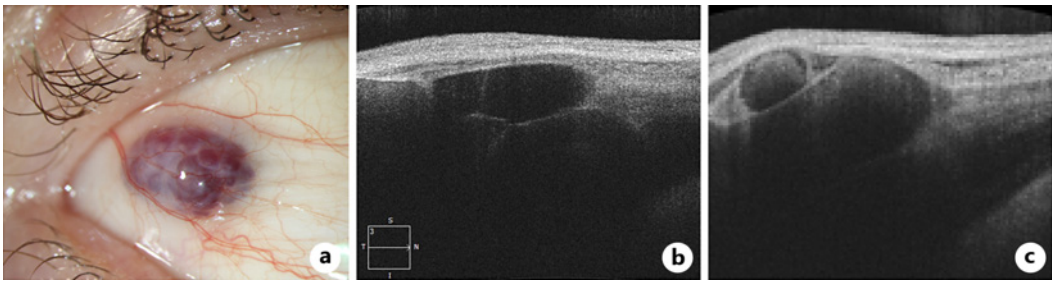

Fig. 1. a Clinical examination of the right eye revealed a purple lobulated subconjunctival mass located temporally. **b** AS-OCT over the lesion demonstrated hypo-reflective intralesional spaces within the mass. **c** These spaces were filled with hyper-reflective material causing back-shadowing.

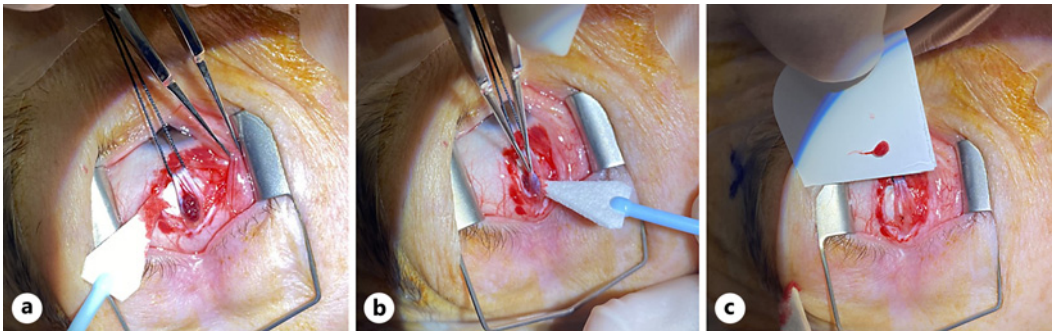

Fig. 2. a Surgical resection of the lesion was done by passing a bridle suture underneath the LR muscle to stretch it, (**b**) followed by careful dissection of the mass from the muscle fibers. **c** A complete muscle-sparing tumor excision was achieved.

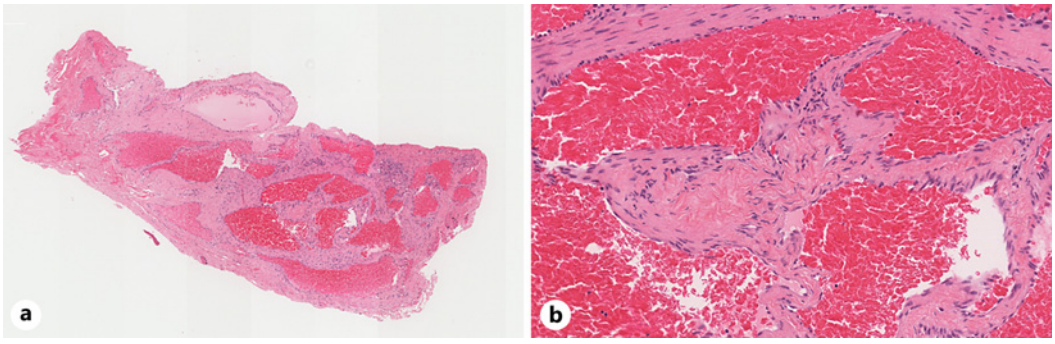

Fig. 3. a Histopathological examination of the specimen revealed a well-circumscribed proliferation of dilated, blood vessels (H&E stain, magnification ×10). **b** The blood-filled venous spaces were lined by flat endothelial cells and separated by fibroconnective tissue (H&E stain, magnification ×40).

possible. MRI of the orbit usually demonstrates EOM enlargement due to a mass lesion with contrast enhancement. In most cases, a single EOM is involved, but rarely, multiple muscles may be involved [3, 5]. The common differential diagnoses of a retroocular IMH include thyroid eye disease, and inflammatory or neoplastic processes, while other rare ones include dermoid, neurofibromatosis, amyloidosis, cysticercosis, and hematoma [2, 8].

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However, in our case, the presentation was unique with a discrete dark red subconjunctival mass lesion, and differential diagnoses considered were conjunctival lymphangioma, hemangioma, and arteriovenous malformations. Clinically, one can distinguish IMH from conjunctival lymphangioma using mobility; IMHs are adherent to the muscle (move along with the EOM during ocular movements), and their overlying conjunctiva is mobile; whereas, lymphangiomas are adherent to the conjunctiva. Due to the anterior location without any orbital component, we could easily image the mass using AS-OCT to establish its vascular nature.

Histologically, IMHs are classified into capillary, cavernous, and mixed types, comprising 50%, 29%, and 21% of all cases, respectively, with the capillary type being commonest in the head and neck region [1, 2]. On reviewing the existing literature on IMHs of EOMs, histopathological classification was available in ten of the eleven reported cases [1, 3–10]. Including this case, capillary, cavernous and mixed types were seen in 27.3%, 45.5%, and 27.3% of cases, respectively. Thus, unlike the rest of the head and neck, the cavernous type of IMH is the commonest in EOMs.

IMHs elsewhere in the body have been managed with various approaches, including surgical excision, radiotherapy, cryotherapy, corticosteroids, and intralesional sclerosing agent injection. In IMH of the EOMs, although surgical resection is the treatment of choice, it is very challenging due to the retrobulbar tumor location with a high risk of complications such as intraoperative massive hemorrhage, injury to crucial structures in the orbital apex, cerebrospinal fluid leakage, optic nerve transection, and post-operative diplopia [2, 3, 8]. Recurrence due to incomplete excision can occur in 9–28% and has been treated in 1 patient with stereotactic radiotherapy [1, 3, 8, 9]. Radical surgery in the form of exenteration or orbital decompression has also been reported due to incessant growth [3, 8]. Response to steroids is poor, and intralesional injection is avoided as it can precipitate retrobulbar hemorrhage [1, 2, 8, 10]. Because of the above limitations in the various treatment options, these tumors are often monitored conservatively [2, 10]. In our case, complete muscle-sparing surgical excision was successfully carried out without any intraoperative complications.

To summarize, IMHs of the EOM are extremely rare tumors that usually present as orbital masses causing proptosis. The atypical anterior epibulbar type seen in this case has not been reported previously. The subconjunctival location, adherence to the underlying muscle, and AS-OCT findings of a multilobulated lesion can distinguish it from the more common conjunctival lymphangioma. Careful surgical dissection with sparing of the muscle fibers is advocated in such cases.

Statement of Ethics

Ethical approval is not required for this study in accordance with local or national guidelines. Written informed consent was obtained from the patient for publication of this case report and any accompanying images.

Conflict of Interest Statement

The authors have no conflicts of interest to declare.

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Author Contributions

H.K. and Z.S.K. were responsible for conception, critical revisions, and data interpretation. A.N. was responsible for design, data acquisition, and drafting the work. S.A.-M. was responsible for acquisition and analysis of data for the work. All authors have approved the final version and agreed to be accountable for all aspects of the work.

Data Availability Statement

All data generated or analyzed during this study are included in this article. Further inquiries can be directed to the corresponding author.

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