

Transcatheter arterial embolization of an orbital lymphoma

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Abstract:

A 69-year-old male patient presented with painless protrusion of his left eye for 6 months. A large, anterior orbital vascular mass was seen in the superior conjunctival fornix. Neuroimaging and Doppler studies revealed a highly vascular circumscribed, enhancing mass lesion in the superior orbit. Carotid angiogram showed a feeder vessel which was embolized followed by complete excision of the lesion. Further investigations confirmed primary extra-marginal zone B-cell non-Hodgkin's lymphoma of the orbit. To our knowledge, this is the first case report of a primary orbital lymphoma treated by Transcatheter arterial embolization and excision.

Keywords:

Embolization, lymphoma, orbit

INTRODUCTION

The most common primary malignant tumor of the orbit is lymphoma.^[1] Marginal zone B-cell lymphoma of mucosa-associated lymphoid-tissue (MALT) account for 59% of all orbital lymphomas.^[1] Orbital lymphomas may rarely involve extraocular muscles, but has never been reported as presenting as a vascular mass lesion involving the same. We report a patient presenting with a primary MALT lymphoma of the orbit involving the superior extra-ocular muscle complex with rapid inflow from the left middle meningeal artery which was successfully managed by Transcatheter Arterial Embolization (TAE) followed by surgical excision.

CASE REPORT

A 69-year-old male presented with complains of painless progressive swelling in the left eye for 6 months [Figure 1a]. There was no history of trauma. He had undergone imaging elsewhere and was diagnosed to have cavernous hemangioma. He was not on any systemic medications. His best corrected visual acuity (BCVA) was 6/6; N6 (Snellen's) in both eyes.

On examination, reddish, mulberry like mass lesion was seen in superior fornix with overlying

dilated tortuous blood vessels, blanching on digital pressure, but showing no variation with the Valsalva maneuver [Figure 1b]. Hertel's examination showed 3 mm of proptosis with 4 mm inferior displacement of the left globe. Retrobulbar resistance was increased on the left. There was no thrill or bruit. Rest of the ophthalmic examination in both eyes was normal.

Magnetic Resonance Imaging (MRI) showed a well-circumscribed soft tissue lesion along left superior rectus/ Levator (SR-LPS) muscle complex, which could not be identified separately from the mass. On T1 weighted images, the mass was isointense whereas on T2 weighted images a central hyperintense area with peripheral hypointensity was seen [Figure 2a-c]. Color Doppler ultrasonography (USG) demonstrated a highly vascular well-circumscribed lobulated mass lesion of the orbit with prominent arterial flow. Imaging and clinical picture being consistent with an orbital vascular tumor, the patient underwent angiography. Carotid angiogram showed a tumor blush in the left supra-orbital region fed by the anterior branch of left middle meningeal artery. This was embolized using 500 micron polyvinyl alcohol (PVA) particles following selective catheterization under general anesthesia. Post embolization angiogram showed complete obliteration of tumor blush [Figure 3a and b]. Patient subsequently underwent complete excision of the lesion. There was

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minimal intraoperative bleeding. The SR-LPS complex could not be separated from the mass and was excised partially along with the lesion [Figure 4a]. Histopathological examination was suggestive of diffuse non-Hodgkin's lymphoma [Figure 4b]. Immunohistochemistry stains showed positivity for CD45, CD20, CD79a and bcl-2 with negativity for CD3, CD10, bcl-6, CyclinD1. These features confirmed the diagnosis of a low-grade non-Hodgkin's B cell lymphoma of marginal zone sub-type. Systemic evaluation (Whole body PET Scan) failed to show any systemic involvement and the patient was referred to radiation oncologist for further management. At follow up visit after six weeks, no residual or recurrent lesion was noted. The patient had complete ptosis of the left upper eyelid for which he was advised surgical correction [Figure 5]. His BCVA was noted to be 6/6; N6 and he has been advised to follow up on a periodic basis both with his oncologist and ophthalmologist.

DISCUSSION

Marginal zone B-cell lymphoma of mucosa associated lymphoid tissue was first recognized by Isaacson and Wright in 1983, as a distinct sub-type of low-grade B-cell lymphoma.^[2] Ocular adnexal lymphomas are an inhomogenous group of tumors. They account for 1-2% of non-Hodgkin's lymphomas (NHL) and 8-10% extranodal lymphomas.^[3] Majority of non-Hodgkin's lymphomas of the orbit are extranodal marginal-zone B-cell type. This tumor is seen more commonly in the 5th-7th decades of life with a slight female predominance. The patients usually present with a palpable, firm or rubbery mass, which maybe visible as pink subconjunctival 'salmon-patch'. Other symptoms are

progressive proptosis, decreased visual acuity, motility disturbances and diplopia.^[4]

In this patient, amulberry-like reddish lesion in superior orbit which on imaging showed a heterogenous contrast enhancement suggested a vascular lesion. This was further supported by Doppler studies and confirmed on carotid angiogram showing a feeder vessel with a rapid flow. Considering the age, location of the mass, clinical and radiological picture, a hemangiopericytoma of the orbit was a more probable diagnosis. Since hemangiopericytomas are notorious for intraoperative hemorrhage, the use of pre-operative embolization has been described in a quite a few reports.^[5-8] Presentation of an orbital lymphoma as a rapid flow vascular tumor has not been described previously in literature. Transcatheter Arterial Embolization has been employed successfully for preventing gastrointestinal bleeding secondary to gastrointestinal lymphoma.^[9] We excised the mass lesion as our clinical diagnosis was hemangiopericytoma, as the definitive treatment of hemangiopericytoma is total resection, to reduce the likelihood of recurrence, malignant transformation, or need for reoperation.^[10,11] Usually, incisional biopsy suffices for histopathological diagnosis of lymphoma; and after immunohistochemistry, they are further managed with chemotherapy or radiotherapy. Due to lack of characteristic clinical features of an orbital Marginal Zone Lymphoma, an en bloc resection of the tumor mass was done.

To conclude, though orbital lymphomas are known to have myriad clinical manifestations, this report is the first of an

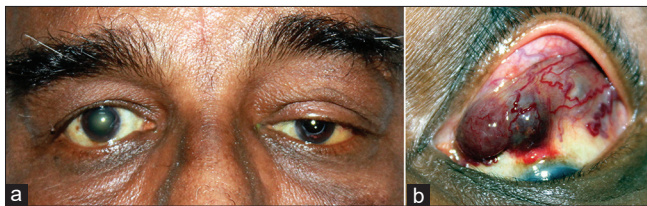


Figure 1: (a) Pre-operative clinical photograph showing left eye ptosis. (b) Mulberry-like mass lesion in the left superior fornix with overlying dilated and tortuous blood vessels

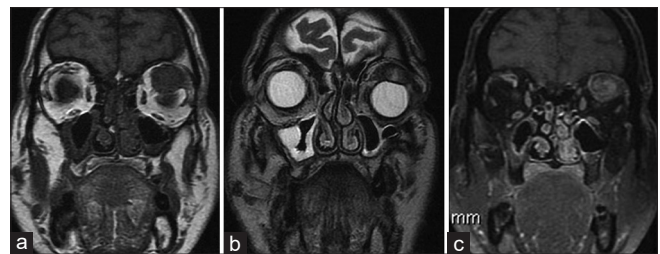


Figure 2: (a) T1 weighted Magnetic Resonance Imaging (MRI) showing a well circumscribed isointense lesion in left superior orbit. SR-LPS complex cannot be identified separately. (b) T2 weighted images showing heterogenous centrally hyperintense but peripherally hypointense lesion. (c) Post Contrast MRI showing moderate heterogenous enhancement

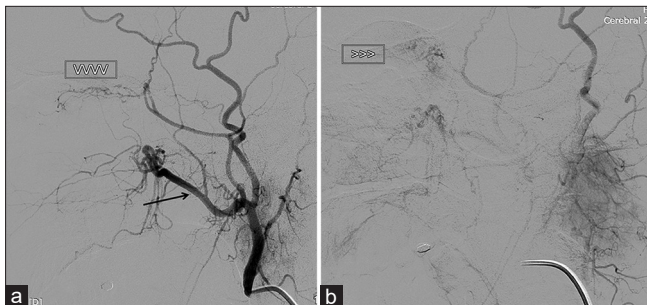


Figure 3: (a) Carotid angiogram showing small tumor blush in left supraorbital region (fed by anterior branch of middle-meningeal artery (Arrow)). (b) Post embolization angiogram showing complete obliteration of tumor blush

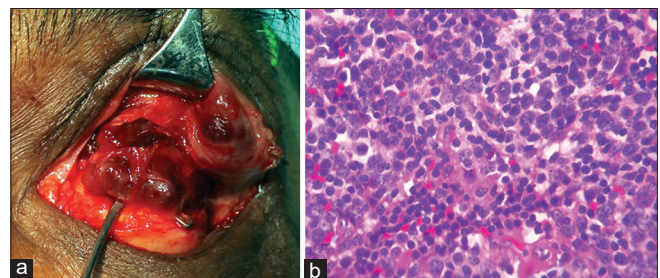


Figure 4: (a) Intraoperative appearance of the lesion inseparable from sr-LPS complex (SR hooked by squint hook). (b) Photomicrograph showing large lymphoid cells arranged around blood vessels (Hematoxylin & Eosin Stain; 100)



Figure 5: Postoperative clinical photograph of the patient after excision of the mass showing left eye complete ptosis

orbital lymphoma mimicking a vascular tumor with a feeder vessel, managed by excision after embolization.

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Conflicts of interest

There are no conflicts of interest.

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