

Case Report

Lateral Orbitotomy for Cavernous Malformation: A Case Report with Technical Consideration

Haroon K¹, Taher T², Alamgir A³, Huq N⁴, Haq MRU⁵, Hossain SS⁶

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Abstract:

Cavernoma is benign tumour of the orbit. It results in proptosis, visual disturbance, diplopia and ptosis of the eye. It is amenable to surgery and this results in complete after surgical removal without recurrence. We report a case of right orbital cavernoma in a female, who presented with non-pulsatile painless proptosis, slight ptosis and double vision while looking at the right side. Her MRI revealed that she had a cavernoma of the right orbit. She was operated by the lateral orbitotomy. This easily removed the tumour, cosmetically acceptable and she became symptom free.

Key Words: Cavernous malformation, lateral orbitotomy, proptosis

Abbreviations: NF-Neurofibromatosis

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Introduction:

Orbital cavernomas are the most frequent intraconal, intraorbital primary tumor in adults followed by capillary hemangiomas, lymphangiomas, and hemangiopericytomas. They represent 4% of all orbital tumors and 9–13% of all intracranial cavernomas¹.

Kennedy had reported of 820 cases of orbital tumours in 1984. Of these 72(8.8%) were classified as vascular lesions. On further breakdown, only 14 cases were cavernous haemangioma².

Although not true neoplasms, cavernous malformations are the most common benign orbital mass in adults. Patients with cavernous malformations are usually middle-aged adults (mean age, 43–48 years), and there

is a female predominance among this patient population. Because these lesions are slow growing, progressive painless proptosis is the most common clinical sign at patient presentation³. The typical clinical presentation is of mostly painless proptosis (mean 5–6 mm), pain, lid swelling, diplopia, lump, and recurrent obstructed vision. Middle-aged women are the most commonly affected group, and the average duration from symptom onset to presentation is 4 years⁴. Easy to remember are the “Six P’s” of orbital lesions: proptosis, pain, progression, pulsation, palpation, and periorbital changes⁵.

These are low-flow circumscribed lesions usually situated behind the globe, most commonly within the

1. Dr. Kaisar Haroon, Assistant Professor, Department of Neurosurgery, NINSH
2. Dr. Tania Taher, Assistant Professor, HFRC Medical College Hospital
3. Dr. Abdullah Alamgir, Associate Professor, Department of Neurosurgery, NINSH
4. Dr. Naila Huq, Associate Professor, Department of Neuropathology, NINSH
5. Dr. Md. Rakib-UI Haq, Medical Officer, Department of Neurosurgery, NINSH
6. Prof. Sk. Sader Hossain, Professor and Head, Department of Neurosurgery, NINSH

Address of Correspondence: Dr. Kaisar Haroon, Assistant Professor, Department of Neurosurgery, NINS. Email:kaisar298@gmail.com, Phone +01711196577, ORCID: 0000-0002-3065-7877

muscle cone. They do not usually interfere with visual acuity, except when the tumor lies in the orbital apex, where it may affect the optic nerve. Cavernous hemangiomas are readily amenable to surgical treatment and do not recur⁶.

In the differential diagnosis of orbital tumors, thyroid related orbitopathies, inflammatory processes, and pseudotumors can be misdiagnosed⁷.

CT scan and MRI are the main investigation modality. CT shows homogeneous soft tissue density, and may show small calcifications or phleboliths. MR shows isointense T1 signal, bright T2 signal, dark internal septations, and a dark circumferential rim that represents a fibrous pseudocapsule⁴.

Histopathologically cavernous angiomas are vascular anomalies, consisting of endothelium-lined caverns filled with blood at various stages of thrombosis and organization, and separated by a collagenous stroma devoid of mature vessel wall elements¹.

There are four primary routes by which transorbital lesions can be reached: 1) the anterior orbitotomy without osteotomy; 2) lateral orbitotomy; 3) medial orbitotomy; and 4) a combination of the lateral and medial orbitotomies. The lateral approach was first proposed by Kronlein¹³ in 1889, and it was later modified by Berke⁸. Intraconal orbital tumors with well-demarcated margins and lateral localization may be removed with lateral orbitotomy⁹.

Case report:

A fifty years old non-diabetic, non-hypertensive woman came to us with history of progressive proptosis of the right eye a little ptosis and double vision of the right eye for about two years. She stated that her right eye gradually became prominent and it came out. There was no pain or pulsation. Her right upper eye lid also came down and she could not elevate the eye lid as before. Her diplopia was occasional, more during trying to look to the right side. This was slowly progressive. She has no history of trauma, fever or blurring of vision. On examination of the eyes, the right eye was pushed forward and medially, there was slight ptosis. Her visual acuity and color vision was normal. Other neurological examinations were normal.

Her MRI showed a tumour at the lateral and superior part of anterior orbit just above the eye ball. It was isointense in T1 and hyperintense in T2 with peripheral contrast enhancement.

The patient was operated upon through a right lateral orbitotomy. This begins with an S-shaped incision in the outer third of the eyebrow, which is directed downward and outward to the outer canthal level and continues posteriorly about 2 cm. the skin and subcutaneous tissue were retracted. The lateral orbital rim was exposed. It was removed along with the lateral wall of the orbit with the help of drill and saw. The tumour was purple in color, well circumscribed and solid. It was about 2cm in diameter. It was totally excised. The orbital rim and bone was placed and fixed with miniplate and screws. The wound was closed in layers. Her stitches were removed on 6th POD and discharged on with advice for followup.

After surgery her movement of the right eye was normal, her diplopia had improved but her ptosis had not improved. On her followup after one month following surgery, her ptosis had improved.



Fig.-1: Cavernoma coming out after removal of the lateral orbital rim and lateral orbital wall

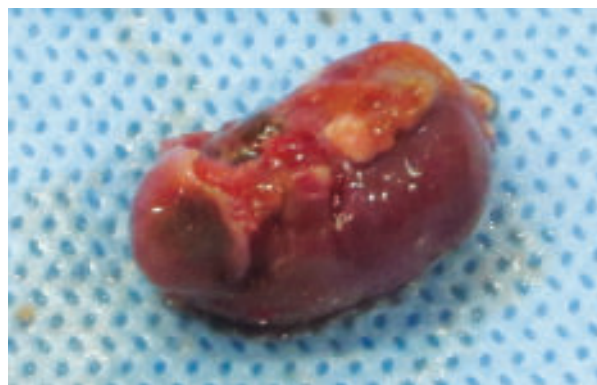


Fig.-2: Cavernoma after excision

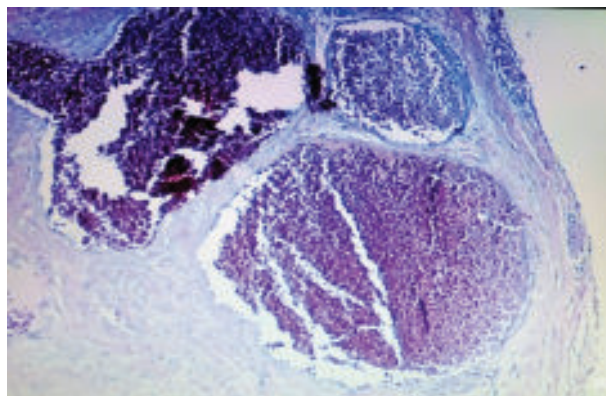


Fig.-3 : *Histopathological photomicrograph. (haematoxylin and eosin stain)*

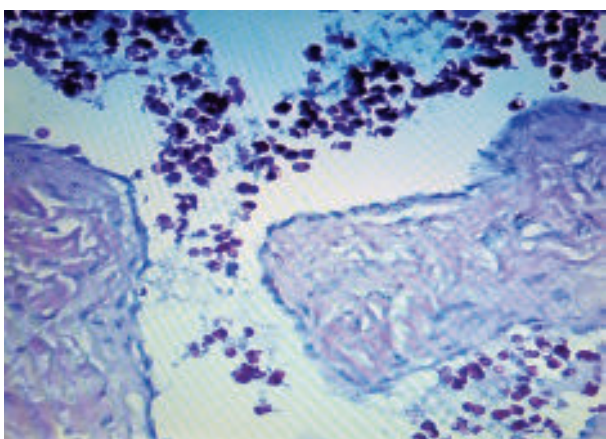


Fig.-4: *High magnification of the Fig 3.*

Discussion:

Orbital cavernoma is more common in female. It is also supported in the literatures^{1, 10}. Our patient was also female. The major presenting symptom is proptosis, resulting from the mass effect¹¹. When the tumour is removed the symptoms improves as well. In our patient it took about a month for the ptosis to improve. Her diplopia had improved immediately following surgery.

Lateral orbitotomy provides excellent exposure of the temporal compartment of the orbit and it is indicated for well defined periorbital and intraconal tumors which are located lateral, dorsal and basal to the optic nerve. It is useful for lacrimal gland tumors, retrobulbar lesions, such as cavernomas, and can be extended for posterior lesions¹².

Lateral orbitotomy provides a wide surgical field in the orbit while providing an efficient access to superolateral cone. Lesions located superior, lateral and inferior to the orbital cone could be removed best via lateral

orbitotomy⁹. Some surgeons preferred mass removal by using a lateral canthotomy and lateral orbital approach in 5 (35%) patients with lesions located anteriorly and laterally⁷. The aim of this incision is to expose and resect the lateral wall and orbital rim⁵. This provides an excellent corridor for the laterally placed tumours. The scar is small and it is cosmetic also. The tumours usually pop out of the orbit after the lateral orbitotomy. The lateral orbital rim, which needs to be removed for surgery, is fixed with miniplates and screws, so there is no disfigurement of the face.

In one study, there was only one case of cavernoma out of ten cases⁹. In another study none of the seven (out of 74 cases) patients with an orbital lesion had worsened after operation: improvement of exophthalmos was seen in six patients, with improved visual acuity in five; the one asymptomatic patient remained intact¹⁰. Our patient had no visual symptoms. She had improved after the surgery. Her visual acuity had had not changed.

Lateral orbital approach is safe for the patients who had tumours at the lateral part of the orbit in Margalit et al series of 41 cases¹³. Boari et al performed eight lateral orbitotomy cases and it was safe and acceptable to the patient¹.

In 1941, W. E. Dandy, one of the pioneers of neurosurgery, postulated that all intraorbital lesions can be attacked by means of a transcranial procedure¹⁴. Today we reserve the transcranial approach for those lesions superior and medial to the optic nerve, especially if they involve the orbital apex. The lateral orbitotomy is widely accepted as the technique of choice for lesions confined to the lateral aspect of the orbit. First proposed by Kronlein in 1889, technical advances and modifications have made the lateral orbitotomy an effective, safe procedure¹⁴.

Conclusion:

Orbital cavernoma is a benign tumour of the orbit. This results in proptosis of the patient. Lateral orbitotomy is a safe and convenient approach for the removal of the laterally placed intraorbital cavernoma. It is cosmetically acceptable to the patient and technically easy for the surgeon.

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