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Case Report

Endoscopic Transmaxillary orbitotomy for Cavernous Malformation: A Case Report with Technical Considerations

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Abstract:

Cavernoma is a benign tumour of the orbit. It results in proptosis, visual disturbance, diplopia and ptosis of the eye. It is amenable to surgery without recurrence. We report a case of left orbital cavernoma in a female, who presented with non-pulsatile painless proptosis, without ptosis and no perception of light. Her MRI revealed a cavernoma of the left orbit inferior to the optic nerve. She was operated by the endoscopic transmaxillary orbitotomy. This easily removed the tumour, avoided craniotomy and was cosmetically acceptable.

***Key Words**: Cavernous malformation, proptosis, transmaxillary approach, endoscopic maxillary approach, Caldwell-Luc approaches.*

***Abbreviations:** CM- Cavernous Malformation, CSF- Cerebro Spinal Fluid*

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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 cavernomas[1].

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 presentation[2]. 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 years[3]. Easy to remember are the "Six P's" of orbital lesions: proptosis, pain, progression, pulsation, palpation, and periorbital changes[4].

The maxillary sinus is a door that opens several important surgical corridors: medially and superiorly providing access to the inferomedial wall of the orbit, posteriorly and laterally to the infratemporal fossa, and posteriorly and medially to the pterygopalatine fossa and pterygoid processes[5]. The transpterygoid approach is the first step in localizing the petrous

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internal carotid artery and provides access to the ventral skull base superior (suprapetrous space) and inferior to the petrous internal carotid artery (infrapetrous, parapharyngeal space)[5].

Because of the close arteries and nerves, surgery of the orbit is difficult and demanding. There are various microsurgical approaches to the orbit, generally classified into transcranial and transorbital[6]. Transorbital approaches are extracranial and include anterior, lateral, medial and combined mediolateral orbitotomy. In addition to the microsurgical approaches, endoscopy is a feature that can be used in orbital surgery. Endoscopy offers excellent illumination of the anatomical structures, and the entry is much smaller compared to microsurgical approaches, leading to a better cosmetic result[6].

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 elements[1].

Case report:

A 30 years old non-diabetic, normotensive female presented to us with progressive dimness of vision of the left eye for three years. She said that she was completely well for three years back. Then she gradually started to have visual disturbance which was difficult to see in the upper field of vision. This had progressed to all fields of vision until she became blind at the left eye. Now in her left eye there is no perception of light. But her eye movements are intact. All other neurological examinations were normal. Her MRI had revealed a non contrast enhancing lesion at the inferomedial part of the left orbit that had pushed the left optic nerve upward and medially (figure 1).

Surgicalconsiderations:

As the tumour was situated inferiorly and medially to the optic nerve, and it had pushed the inferior rectus muscle laterally and the optic nerve superiorly so the decision to approach the tumour through an endoscopic transmaxillary approach was selected.

The patient was placed supine and the head turned to the left and extended. The head was fixed with a three point head fixator. The upper lip of the left side was retracted. The canine tooth was identified and a mucosal incision was given about 2 cm long on either side of the left canine tooth just above the groove. Mucosa was stripped off the bone. An aperture was made at the buccal groove about 1 cm in diameter. This is similar to the Caldwell-Luc approach to the maxillary air sinus (figure 2). Then the endoscope was brought in. With the 30°and 0° telescope the groove of left inferior orbital nerve and maxillary foramen was identified (figure 3). The groove of the inferior orbital nerve is an important landmark. It has to be identified early and clearly. This groove is situated at the roof of the maxillary air sinus and so it is the floor of the orbit. The maxillary foramen is also important as this marks the medial wall. The small triangular space just medial to the groove is the floor of the orbit. If any confusion arises about the position or direction of the endoscope, these two landmarks can help to restore the orientation.The grove of the inferior orbital nerve was kept laterally and maxillary foramen medially. A 30° endoscope was used to visualize the working area. The 0° helps to find the working area but the 30° is more helpful to work with. The roof of the maxillary antrum was drilled. The tumour just popped into view. It was reddish, with a definite capsule and was partly

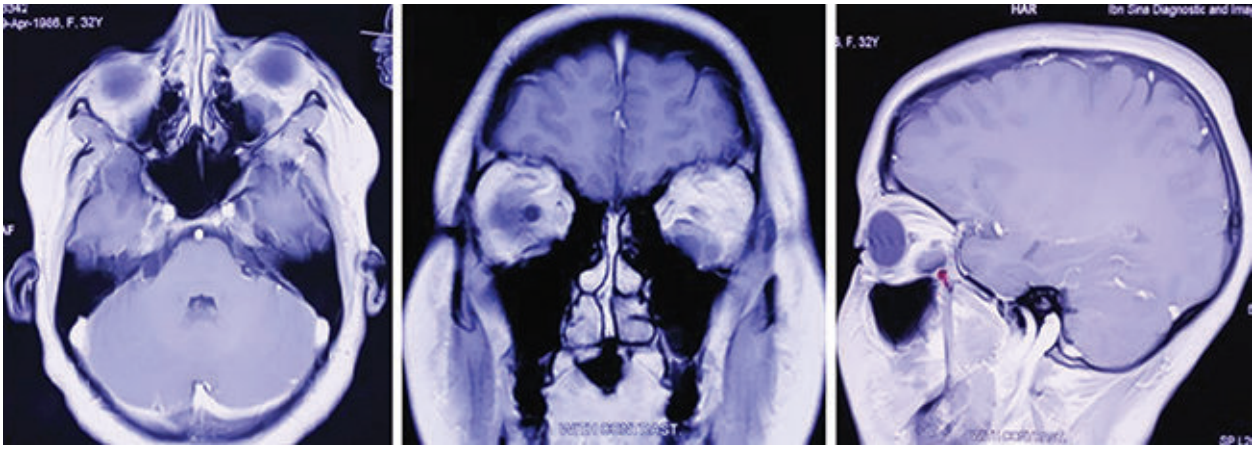

Fig.-1: *Pre operativeT1WI MRI : Non contrast enhancing isointense lesion in the floor of the left orbit*

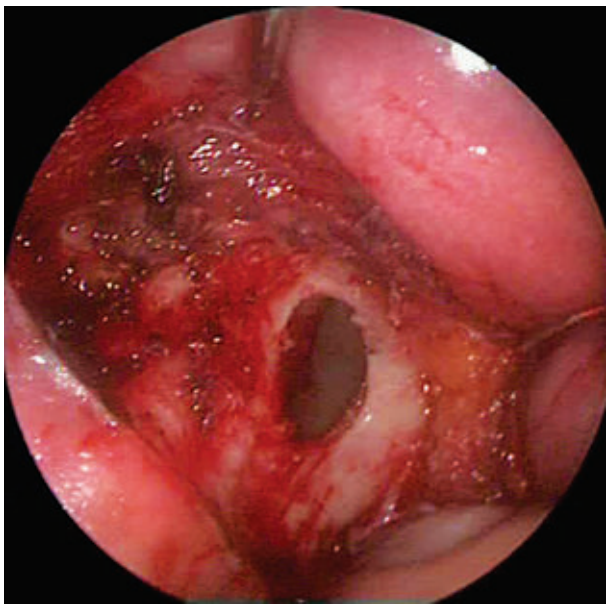

Fig.-2: *Left maxillary antrostomy done*

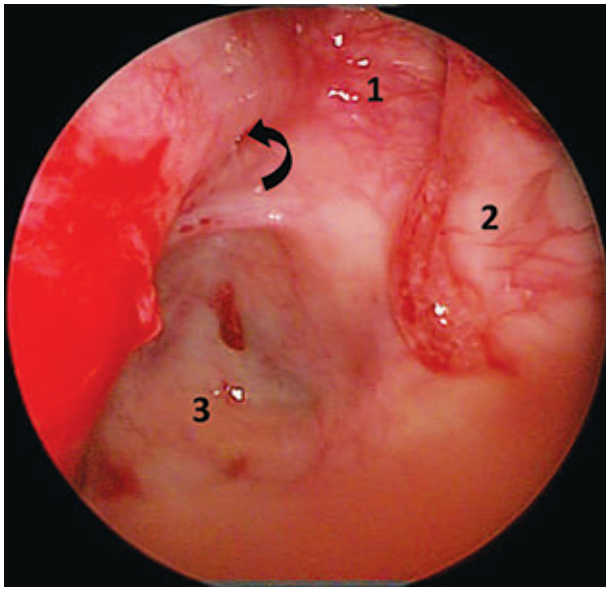

Fig.-3: *endoscopic view of the left maxillary sinus. Curved arrow – opening of maxillary sinus, 1= roof of the sinus, 2= groove of the inferior orbital nerve, 3=posterior wall of maxillary sinus*

suckable. It was moderately vascular. It was removed in piecemeal and suction. Then the orbital fat had herniated down. Haemostasis was secured with bipolar coagulation and Surgicel©. Then antrostomy was covered with miniplate and screws (figure4). Mucosa was sutured with plain 4/0 catgut in round bodied needle.

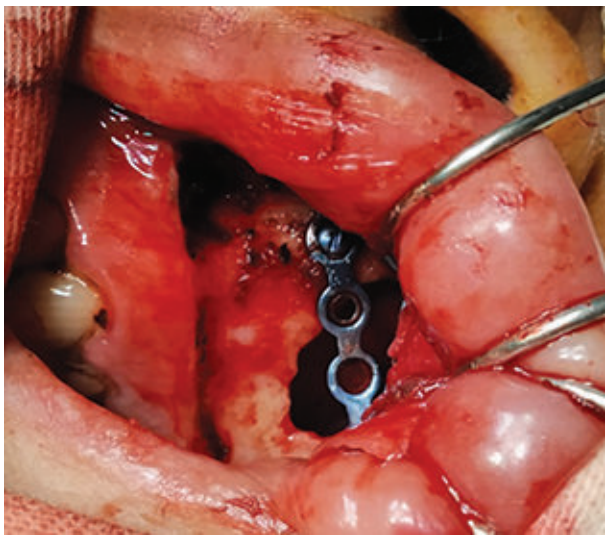

Fig.-4: *Antrostomy partially closed with miniplates*

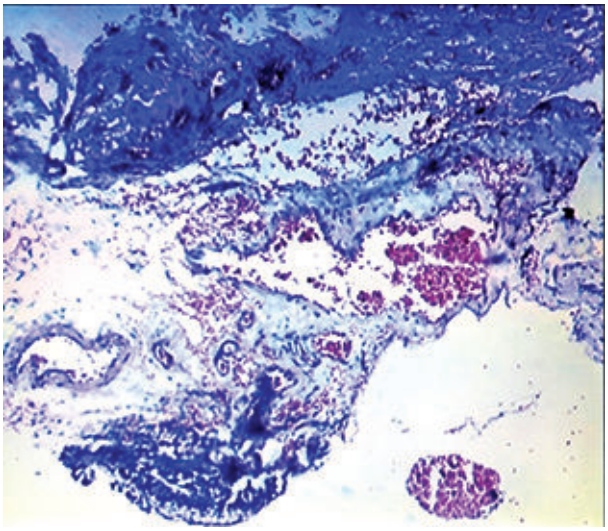

Fig.-5: *Histopathologilcal photomicrograph of the tumour (H&E stain)*

The patient had a smooth recovery.Her wound had healed well. After surgery movement of the left eye was normal.But her vision had not improved. She was discharged from the hospital on the 8thpost operative day. Her histopathological report suggested cavernous haemangioma (Fig 5).

Discussion:

These are low-flow, circumscribed lesions usually situated behind the globe, most commonly within the 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 hemagiomas are readily amenable to surgical treatment and do not recur[7]. Our patient was blind in the left eye. There was no perception of light. This is because the tumour was at the orbital apex where all the nerves vessels and muscles are closely packed together.

In the sublabial-transmaxillary approach, the bony opening of the maxillary sinus provides the medial and lateral limits of surgical freedom. The effective perpendicular cross-sectional area is excellent because the bony opening is almost perpendicular to the angle of attack[8].Intraconal orbital tumors with well-demarcated margins and lateral localization may be removed with lateral orbitotomy[9,10]. Intraoperative navigation remains essential for the safety and maximal effectiveness of this approach as it does on all endoscopic cranial base procedures[11].

During surgery there were some difficulties the team had faced. Firstly the position of the patient had to be carefully placed, so that the orbital floor was vertical to the floor and nearer to the surgeon. The small opening of the maxilla allowed only limited movement of the telescope. Using a 0° and a 30° telescope alternately helped in the procedure. There was haemorrhage from the mucosal wall which was controlled by monoplar coagulation. Sometimes there was disorientation of the anatomy; therefore it was corrected by keeping the two landmarks (e.g. infraorbital groove and maxillary foramen)in view

Conclusion:

Orbital cavernoma is a benign tumour of the orbit. Transmaxillarytransorbital approach is a good approach to the inferior wall of the orbit. It provides excellent visualization of the tumour and avoids craniotomy or orbitotomy. It is cosmetically acceptable to the patient and technically easy for the surgeon.

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