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Infra-orbital Nerve Schwannoma with Sino - Orbital Involvement : A Case Report

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ABSTRACT: 52 yearS old lady was referred to our centre with complaint of left eye proptosis to upward gaze for past 6 months. Computer tomography (CT) brain and orbit showed heterogenous mass arising from left maxillary sinus extending into extraconal space of the left orbit. The floor of left orbit was eroded and the mass cause expansion and bowing of the walls of left maxillary sinus. Biopsy performed via sublabial approach revealed benign neural tumor. The mass was removed via open medial maxillectomy approach and orbital floor was reconstructed using titanium mesh. We report a case of schwannoma originating from infra-orbital nerve and review the literature.
Key Words : Schwannoma, Infraorbital nerve, Medial maxillectomy.

INTRODUCTION : Schwannomas are rare tumor arising from the schwann cells, the outermost layer that sheath the peripheral nerve. It can occur in all parts of the body. It constitutes 1-8% of all head and neck tumors and 1-4% of the orbital tumors. Mostly occur in 2nd to 4th decade of life[1].

CASE REPORT : A 52 years old lady was referred from ophthalmology department complaining of progressively worsening left eye proptosis to left upward gaze for the past 6 months associated with intermittent numbness over left cheek. No history of double vision or color blindness. No reported sinonasal symptoms. There was no history of trauma. Clinical examination showed proptosis of left eye anterolaterally and the globe is displaced superiorly, slight fullness over left cheek, non-tender and not inflammed. Vision was normal. No nystagmus and all cranial nerves were intact. No neck nodes palpable. Rigid nasal endoscopy performed and showed no evidence of sinusitis or intranasal mass. Systemic examination revealed no signs of hyperthyroidism or connective tissue disease. Other examinations were unremarkable. The CT orbit and brain showed heterogenous enhancing mass with epicenter within the left maxillary sinus. It measures 3cm x 3.5cm x 5cm. There was no calcification seen within. The mass cause expansion and bowing of the walls of the left maxillary sinus. The mass extend superiorly into the extraconal space of left orbit. There was erosion over floor of left orbit. The left osteomeatal complex was obliterated. The optic nerve were not affected bilaterally. Biopsy was taken via sublabial approach, reported as benign neural tumor, with differential of schwannoma originating from infraorbital nerve. She underwent open medial maxillectomy to remove the tumor with reconstruction of left orbital floor with titanium mesh. Intraoperatively there was a well capsulated mass occupying maxillary antrum extending to infraorbital region with partial erosion of orbital plate. The mass was not adherent to periorbita and left orbit was intact. Maxillary mucosa and membrane were normal. Lateral wall of left nose was left opened to create a connection with maxillary sinus. Post operatively, there was numbness over left cheek, ala of left nose and left upper lip and minimal skin dimpling inferior to left orbital floor. The left eye was less proptosed and extraocular movements were normal with no diplopia. Patient was under our follow up for 24 months post-operatively and was disease free without evidence of recurrence.

DISCUSSION : Schwannomas account for 8 % of all intracranial tumors and most frequently arise from vestibular division of vestibular cochlear nerve. Other cranial nerves are less frequently involved. They can develop anywhere in the body, arising from myelin sheaths of peripheral motor, sensory, sympathetic and cranial nerves. Between 38 to 45% of all schwannomas occur in head and neck region[2]. Presenting symptoms are varied and non-specific depending on the site of tumor. They are often not detected in the early stage due to the lack of early symptoms and because of their slow growing nature.

Orbital schwannomas also known as orbital neurilemmomas are rare tumors. They constitute 1-4% of orbital tumor[1]. It usually arises from branches of the supra-orbital or supra-trochlear nerves, less commonly arises from infra-orbital, ciliary, oculomotor, trochlear or abducent nerves[3]. Schwannomas from infra-orbital nerve are very rare, only seven cases were reported up to date. They may occur at any age with no gender predilection[4]. They are localized and well encapsulated, usually unilateral and rarely become malignant[5]. In most cases, the nerve of origin is unable to be identified during surgery in almost 50% of cases[1-6]. In this case, the nerve from which the tumor originate can be identified according to the pre or post-operative neural deficit. Patient complaint of numbness over the left cheek, left ala of nose and upper lip post-operatively thus confirmed that infra-orbital nerve is the nerve from where schwannoma originated. Most of the tumor originate from sensory nerves, so, they do not interfere with eye movements or vision unless they are located in the orbital apex and compress the optic nerve[1,7,8]. Similar to our patient, she only complaint of left eye proptosis with upward gaze.

Schwannomas are benign, painless, slow growing and

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can affect adjacent structures by erosion of bone and direct extension. They push and erode walls by pressure effect. In this case, the orbital floor was pushed inferiorly, the tumor occupied the left maxillary antrum and extend superiorly to the extraconal space of left orbit. Post-operatively, it left a large space between the contents of the orbita and the orbital floor, thus, we constructed a new orbital floor using the titanium mesh.

Histopathological characteristics of orbital schwannomas are the same with other schwannomas of the head and neck. Antoni A and B (hypercellular and hypocellular area) with Verocay bodies are characteristic of these tumor and their differential diagnosis with neurofibromas is based on these histopathological findings[7]. Neurofibroma may be distinguished by the occurence of calcification that is infrequently seen in schwannomas[5]. In CT scan, orbital schwannomas are seen as homogenous smooth bordered solitary, and are iso-intense with the brain, very rarely cystic masses[1,9].

Treatment of choice is surgical excision and the approach depends on the location and extension of the tumor. Imaging helps in deciding the approach route. In this patient open approach choosed as from the imaging showed that the tumor located in left maxillary sinus with superior extension to the left extraconal space and erosion of floor of the left orbit. Total removal of the tumor is usually ample to prevent recurrence[1,2,9]. The prognosis for this benign tumor is good and recurrence is rare if the tumor is removed completely.

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