https://doi.org/10.36570/jduhs.2022.1.1095

JDUHS

CASE REPORT

Schwannoma- A Rare Cause of Proptosis

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ABSTRACT

Schwannoma a tumor of cranial and peripheral nerves is rarely found in the orbit and if present seldom causes proptosis. We report a rare cause of unilateral proptosis. A 15 year's old boy reported with a 10-month history of unilateral proptosis. Ophthalmic and slit lamp examination revealed no other abnormality except for a dystopia, 3 mm inferiorly and 1.5 mm medially of left eye. On palpation, a small, non-tender rubbery nodular swelling in the left supero-temporal region was found. Magnetic Resonance Imaging (MRI) was suggestive of a lacrimal gland mass with a hyper intense area in the region of left lacrimal gland. Complete excision biopsy was done which reported a Schwannoma of lacrimal nerve. This case emphasis on two important points. First how to approach and manage a rare cause of unilateral proptosis and also that Schwannoma of lacrimal nerve has a very good prognosis after complete surgical excision and a good follow up.

Keywords: Schwannoma, Lacrimal Gland, Proptosis, Orbital Swelling.

INTRODUCTION

Schwannomas (also called neurileimoma) are rare tumors, which may arise from cranial or peripheral nerves.[1] They account for 1% of all orbital tumors in adults and are considered a rare cause of proptosis.[2]

They usually occur in isolation but maybe associated with neurofibromatosis. They are categorized as benign tumors but may undergo malignant transformation if related to neurofibro-matosis. Benign tumors are asymptomatic if small but larger tumors may cause painless and progressive proptosis. A variety of signs and symptoms may occur due to the variable origin and location of the tumor in the orbit.[3]

Displacement of the eyeball is related to the site of the schwannoma. Diagnosis can be confirmed only on histopathological report. Prognosis is excellent if complete excision of the tumor is carried out. Because of its rare occurrence at this site and being the very rare cause of proptosis, this case is being reported in which we are reporting (with the consent of the patient) this rare case of proptosis which was caused by Schwann-oma of the lacrimal gland.

CASE REPORT

A 15 year's old male presented to the ophthalmology outdoor department with 10 months history of gradual, progressive, painless protrusion of the left eye. There was no history of pain, fever, vomiting, double vision, weakness, palpitations, tremors, cold/heat intolerance. He had not sustained any trauma or undergone any ocular surgery. He was not using glasses.

On examination, a young man of average built with stable vital signs and unremarkable systemic examination. Ophthalmic examination revealed, visual acuity of 6/6 in both the eyes, with normal optic nerve functions. There was dystopia of 3mm inferiorly and 1.5mm medially of the left eye, with non axial proptosis. Exophthalmometry by Hertel revealed a difference of 5mm between the two eyes. There was no lid lag/ lid retraction and extra ocular muscles had a normal range of movement. A small nodular swelling was palpable in superotemporal region of left orbit, which was non tender, rubbery in consistency, mobile, 1mm x 1mm in size and its posterior margins were not palpable. Orbital margins were palpable. Para nasal sinuses were non

tender. Thyroid gland was not enlarged. Lymph nodes were not palpable. There was no resistance to retropulsion, and auscultation revealed absence of bruit. The anterior and posterior segment examination of both the eyes was normal.

Blood complete picture, blood sugar random, chest x-ray and thyroid function tests were all normal. Computed tomography (CT) scan showed hyper dense, lobulated soft tissue lesion, in left supra orbital region. There were no calcifications within the mass and no erosions of surrounding orbital wall. Magnetic resonance imaging (MRI) showed well defined hyper intense area in region of left lacrimal gland with significant inferior displacement of the globe and proptosis (Figures 1 & 2). Significant contrast enhancement showing intra and extraconal involvement and displace-ment (Figures 3 & 4).

A provisional diagnosis of lacrimal gland tumor was made and lateral orbitotomy and excisional biopsy of left orbital mass under general anesthesia was done after taking an informed consent (Figure 5).

The biopsy however revealed schwannoma of lacrimal nerve (Figure 6). The patient was followed up after 1,3 and 9 months and remained symptoms free.

DISCUSSION

Proptosis is an abnormal protrusion of the globe, which may be caused by infection, inflammation, tumors(benign/malignant), varices or bony anomalies.[4] It may be classified as unilateral/bilateral, axial/non axial, pulsatile/non pulsatile, depending on the underlying cause.

Inferomedial dystopia may be caused by lacrimal gland tumors which are responsible to give rise to non axial proptosis, along with the dystopia. Majority lacrimal gland masses are idiopathic inflammatory dacryoadenitis. Among the tumors 50% are benign mixed (Pleomorphic adenoma) and 50% are carcinomas.[5] Benign mixed cell tumors account for 25% of all lacrimal fossa lesions.[6]

Rarely neural tumors occur in association with lacrimal gland (e.g., neurofibromas, schwannomas). These lesions account for 6-8% of intracranial neoplasms but do not usually cause proptosis, and account for only 1%

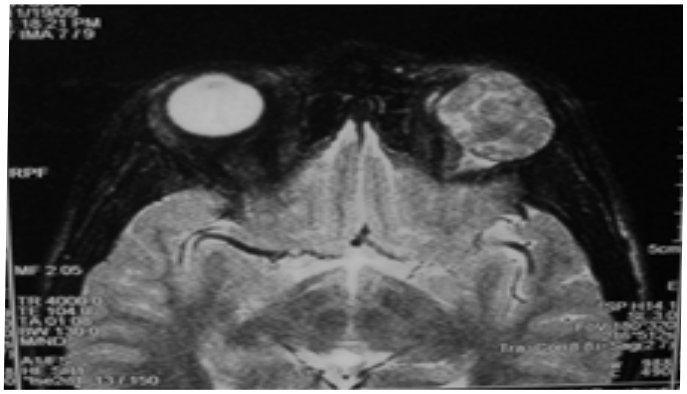

Figure 1: MRI ,T2 Axial image showing hyperintense lesion in the superolateral aspect with of the orbit causing proptosis

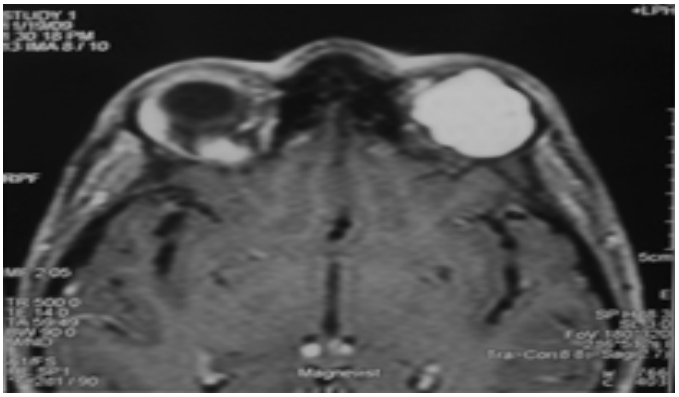

Figure 3: MRI: Contast enhanced image showing hypointense orbital mass

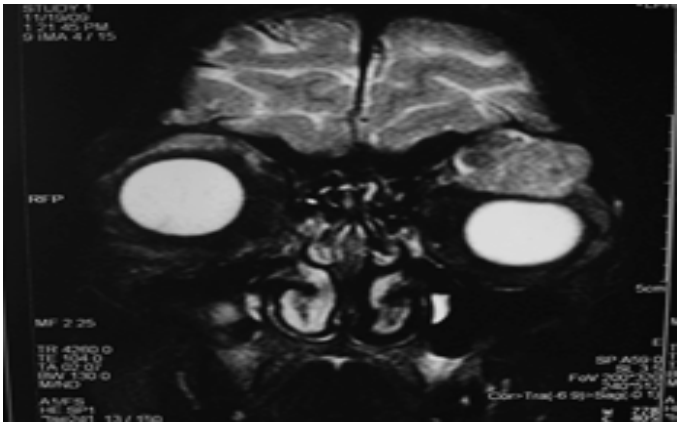

Figure 2: MRI, T2 Coronal image showing inferior displacement of the orbit

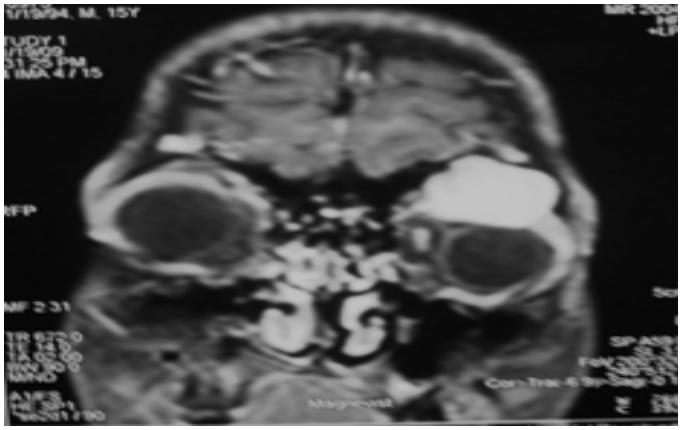

Figure 4: MRI : Contrast enhanced coronal view showing displacement of the globe.

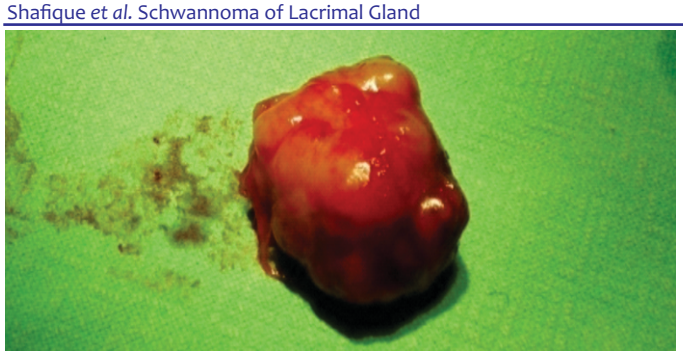

Figure 5: Mass after Excision biopsy

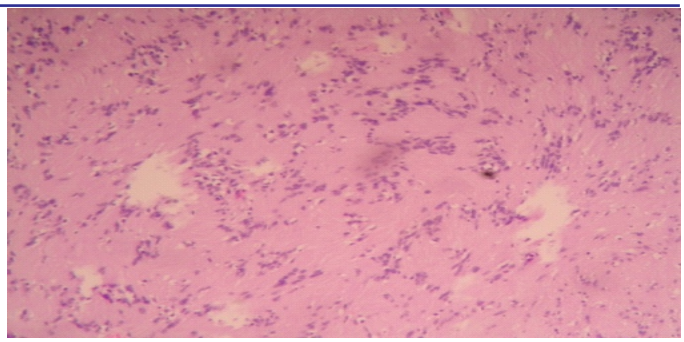

Figure 6: Microscopic image of a Schwannoma

of orbital tumors.[6] Schwannomas also known as a "neurinomas", "neurilemoma", and "Schwann cell tumors" are benign nerve sheath tumors composed of Schwann cells, which normally produce the insulating myelin sheath covering peripheral nerves. They develop on the outside of the nerve, but as they increase in size, they may push aside adjacent structures and may cause propstosis and dystopia, as was in our case.[7] They present between 20 to 70 years of age. They may occur in isolation or in association with neurofibromatosis.[8] These are benign tumors if not associated with neurofibromatosis.

They cannot be diagnosed on clinical examination alone. They may be detected by CT scan/MRI but need confirmation by histopathology. The neurilemomas are characterized by two basic tissue types.[9] Antoni type A with Verocay bodies, and Antoni type B.

There is no role of medical therapy in treatment of Shwannomas but they may be treated by conservative surgical excision, with minimal risk of recurrence.

They carry excellent prognosis after complete excision. Gamma Knife surgery sometimes referred to as stereotactic radio surgery (SRS), has largely replaced surgical resection for the treatment of schwannomas, particularly when the lesions do not compress the brainstem.[10]

CONCLUSION

The histopathology report of Schwannoma of lacrimal gland was a pleasant surprise as it is rare in the orbit and since the examination and investigations suggested otherwise it was not considered in the differentials diagnosis either. It is benign and also has a good prognosis after complete excision and no recurrence. Hence the reason to report this case is that even with a common presentation a rare cause can be found and a timely biopsy is pivotal and can lead to a happy patient and a happy surgeon at the end of the day.

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