Case Report

Solitary Intraorbital Schwannoma in Zaria, Nigeria: A Case Report and Review of Literature

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Abstract

Solitary Intraorbital Schwannomas are rare tumours constituting only 1% of orbital neoplasms. They are benign, truly encapsulated slow growing peripheral nerve sheath tumours originating from Schwann cells. We present the first diagnosed case of Intraorbital Schwannoma in our department. A 45-year-old woman who presented with right eye proptosis of 8 years duration and visual loss of 3 years duration. Examination showed a space-occupying lesion in the right orbit that was firm to palpation, adherent to deep planes, painless and displaced the right eye inferiorly. Computerize Tomographic scan revealed a homogenous soft tissue mass which was isodense to the extraocular muscle within retro-orbital region and eyelid with associated significant proptosis. Pathological examination revealed schwannoma. Patient had exenteration of the right orbital content after counselling and did well 6 months post treatment. Overall, intraorbital Schwannomas are infrequent tumours arising from the orbit. In this review, we shall discuss the clinical and histopathologic findings of this rare presentation of the tumour at this uncommon site and review the relevant literatures.

Keywords: Orbit, Schwannoma, Proptosis, Exenteration, S100, EMA

INTRODUCTION

Schwannomas, also known as Neurilemmomas, are slow growing benign tumors that arise from Schwann cells in the peripheral nerves and were first identified by Verocay in 1908 [1]. Schwannoma is one of the few neoplasms that is completely encapsulated [2]. Intraorbital schwannomas are uncommon, making up less than 1% of all orbital neoplasms and often occur in adults aged 20 to 70years [3,4]. A case series done by Rootman *et al* revealed a predilection for the male gender [1]. These tumors usually arise from sensory nerves traversing the orbit, most frequently from the supraorbital and supratrochlear nerves [2]. However, Optic schwannomas are exceedingly rare because the optic nerve is myelinated by central nervous system oligodendrocytes rather than Schwann cells and to our knowledge, only six cases of intraorbital schwannomas have previously been reported [5]. Intraorbital schwannomas are usually asymptomatic when the tumors are small but the key-feature of any orbital tumor over time, is exophthalmos [6]. Though, clinical presentation ranges from insidious proptosis, visual field loss, retro-orbital pain and headaches, to in rare cases blindness. We present the first case of intraorbital Schwannoma diagnosed in our department.

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CASE REPORT

A 45-year-old female patient ~~presented~~ to our hospital with history of progressive proptosis of the right eye of 8 years duration and progressive loss of vision of 3 years duration. There was associated history of periorbital pain and discomfort as the proptosis progressed. She gave no medical history of trauma, instrumentation, or blunt injury to the face. There were no other contributory complaints.

Clinical Examination showed a space-occupying lesion in the right orbit that was firm to palpation, and adherent to deep planes. It was non tender and displaced the right eye inferiorly. There was nil perception of light in the right eye. Other systemic examinations were unremarkable.

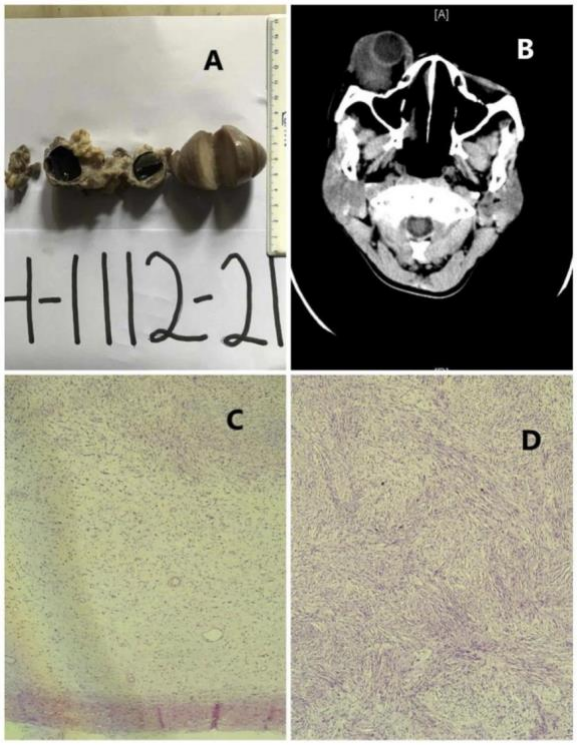

Fig 1 Show the cut sections through the eyeball showed unremarkable anterior and posterior chambers and an ovoid that is nodular gray-white and in appearance (A). Brain CT (B) showing a homogenous soft tissue mass (HU=32) measuring 4.5cm x 2.6cm x 2.3cm (CC x AP x TR). Sections from (C & D) showed an encapsulated tumour composed of spindle cells proliferation disposed in whorls and swirling pattern forming concentric circles with intervening collagen surrounding thin wall vascular channels in areas (X 4 and X 10 respectively).

Investigations included a Brain CT (Figure 1b) which revealed a homogenous soft tissue mass (HU=32) measuring 4.5cm x 2.6cm x 2.3cm (CC x AP x TR). The mass was isodense to the extraocular muscle in the right retro-orbital region and eyelid with associated significant proptosis Homogenous enhancement in the delay series (HU=40) post IV contrast was noted. The optic nerve was difficult to delineate from the mass, however the optic canal was not expanded and there was no infiltration of the eyeball. There was no adjacent bone erosion or remodeling. The rest of the cerebral hemisphere, brain stem, cerebellum, ventricles, cortical sulci, and basal cisterns showed normal morphology and density. No intracranial extension was seen, and the left orbit, retro-orbital structures and paranasal sinuses were normal in outline and density.

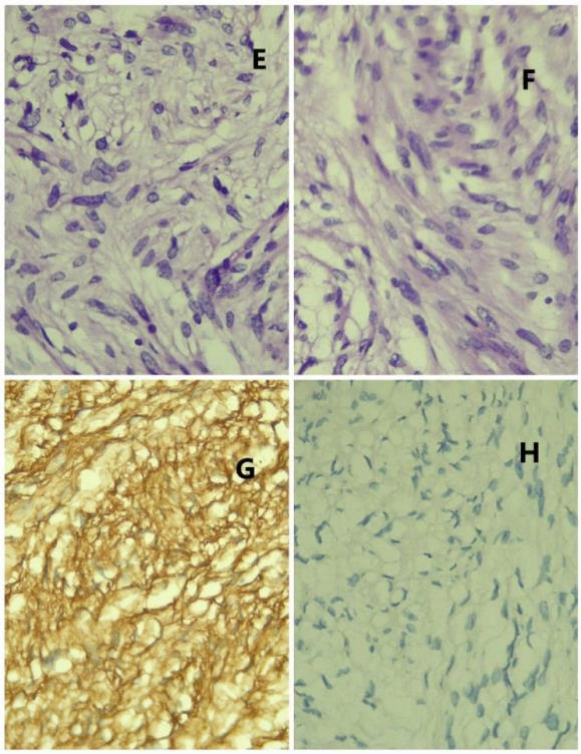

Fig 2: Sections from (E & F) the tumour cells are wavy with oval having bland nuclei features. Few scattered random pleomorphism were noted. The stroma is loose to myxoid. Other areas show schwannian-like stroma and shredded carrot appearance supporting a diagnosis of schwannoma. Sections from the globe, optic nerve and intra-orbital content were unremarkable.

A clinical diagnosis of leiomyoma with differentials of neuroma was entertained. Based on the visual loss, proptosis, and orbital inflammation due to late presentation, patient was counselled and had exenteration of the right orbit. The post-operative period was uneventful, and the patient has been regular on follow-up with no new complaints.

The exenterated orbital contents were sent to the pathology laboratory (Figure 1a). Two specimens were received: a distorted right globe with lens opacity accompanied by a well-circumscribed ovoid grey mass. The globe measured 4x4x2cm and weighed 20g while the accompanying mass measured 5x4x3cm and weighed 19g. Cut sections through the eyeball showed unremarkable anterior and posterior chambers containing aqueous and vitreous humour respectively. The optic nerve was unremarkable measuring 0.5cm in length. Cut sections through the ovoid mass showed white nodular to homogenous-surfaces.

Histologic sections from the ovoid mass showed an encapsulated tumour composed of spindle cell proliferations disposed in whorls and swirling pattern forming concentric circles with intervening

collagen surrounding thin wall vascular channels in areas. The tumour cells are wavy to oval having bland nuclei features. Few scattered random pleomorphism were noted. The stroma is loose to myxoid. Other areas show schwannian-like stroma and shredded carrot appearance supporting a diagnosis of schwannoma (Figure 1c-d). Sections from the globe, optic nerve and intra-orbital content were unremarkable. Immunohistochemical stains for S100 and EMA performed revealed uniform diffuse and strong positivity for S100 while EMA and silver stains were negative, thus confirming a diagnosis of solitary intra-orbital Schwannoma.

DISCUSSION

Primary Schwannomas rarely occur in the orbit and when they do, they generally arise from intra orbital sensory nerves. The most frequent being the supraorbital and supratrochlear branches of the frontal nerve [7]. Orbital schwannomas are mostly asymptomatic when small, but as they gradually grow, progressive painless ocular proptosis can develop and prompt patients to seek medical attention [8]. The index case presented with history of progressive proptosis and visual loss which are common findings as documented from seen in other reports [2,3,9,10]. Our patient presented in the 5th decade which is in keeping with other case reports showing common occurrence in adults from 3rd to 7th decades with a median age of 40 years [11,12].

A variety of imaging techniques are available for orbital schwannoma assessment and monitoring. CT is best for detecting bony involvement and planning for surgery, whereas MRI best describes the nature of the tumour and its involvement of nearby soft tissue structures.

Orbital schwannomas present on CT imaging as smooth, spherical, elongated, and homogeneous lesions with a density like extraocular muscle. (12) Furthermore, CT can detect calcification in tumours when present. [13] They tend to show significant augmentation with CT contrast and appear as smooth, well-circumscribed tumors that adapt to the contour of the cavity and develop along the orbital axis. When compared to other peripheral nerve sheath tumours (PNSTs), orbital schwannomas are more oval or spindly in form. They are often extraconal, in contrast to haemangiomas and other masses on the differential diagnoses that are frequently intraconal. [14] In comparison to meningiomas, schwannomas are more likely to infiltrate through the superior orbital fissure. [15,16] Furthermore, CT will demonstrate their distinctive development into bone without eroding fissures, which occurs less frequently in neurofibromas.

On MRI, Schwannomas often exhibit hypointense signaling on T1 and hyperintense signaling on T2. MRI can detect homogeneous or heterogeneous enhancement that correlates with tumor histology and morphology. Antoni A areas exhibit intermediate intensities with T1 and T2, whereas Antoni B regions are hypointense on T1 and hyperintense on T2. There is no contrast enhancement, however in around 41% of schwannomas, there may be cystic degeneration; these areas may correspond to Antoni B regions. MRI can identify schwannoma separate from lymphoma because lymphoma tend to show intermediate T2 and molds around neighbouring structures, but schwannomas intrude and distort the surrounding tissue. Dermoid cysts have a similar structure but are hyperintense on T1 and lack gadolinium enhancement. Solitary fibrous tumours have comparable T1 and T2 intensities and positions, however dynamic contrast enhanced MRI can tell the difference. Solitary fibrous tumours exhibit a smoother washout curve in dynamic contrast MRI due to their dense cellular stroma, whereas schwannomas have a plateau washout due to a nonuniform and loose cellular organization. Cavernous haemangiomas will be distinguished by dynamic MRI enhancement because they will exhibit increasing enhancement in later scans. Schwannoma characteristically show encapsulation and is one of the truly encapsulated human tumours.

The classic histology is that of hypercellular (Antoni A) areas and hypocellular (Antoni B) area, with the former showing nuclear palisading and Verocay bodies formation. This classical histological appearance may not be present in all cases as shown by this case and this raise important differential diagnostic considerations. The important differential diagnoses include other PNSTs, particularly neurofibroma. However, neurofibroma usually lacks a capsule and is composed of spindle cells with shredded carrot collagen and mast cells. Hypocellular, myxoid areas without hypercellular areas are usually seen. It shows a weaker and patchy S100 positivity, focal calretinin, stronger CD34, and is also factor XIIIa positive.

Leiomyomas may show palisading and extensive degenerative changes in the form of hyalinization, calcification, myxoid changes and ancient nuclear atypia. However, they are S100-, SMA+ and desmin+. [17,18] Malignant peripheral nerve sheath tumors (MPNST) have an infiltrative growth pattern with hypercellular, pleomorphic nuclei and high mitotic activity. Areas of geographic necrosis and divergent differentiation may be seen. Solitary circumscribed neuroma (palisaded encapsulated neuroma) is another important differential diagnosis of this tumour. It is most often an encapsulated dermal or subcutaneous tumor which may be seen with club-like extension in the subcutaneous tissue and may show nuclear palisading. However, silver stains show the traversing axons withing the lesion; however, these axons are located near the capsule in schwannomas. The late presentation of this case which was associated with a complete loss of sight on the right eye necessitated a more radial management approach (exenteration).

CONCLUSION

The orbit is an uncommon and a rare site for a Schwannoma and a high index of suspicion should be entertained in patients with inexplicable proptosis/ exophthalmos and visual loss Prompt medical intervention is required to avoid visual loss.

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