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INFRATEMPORAL FOSSA SCHWANNOMA IN YOUNG CHILD BOY: CASE REPORT

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Abstract

Background: When a schwannoma affects the mandibular branch from the trigeminal nerve, the solid mass enlarges the foramen ovale, with components below, in the masticatory space, and above, in the middle cranial fossa. Atrophy of the masticator muscles is common. the pterygopalatine fossa and infratemporal fossa are adjacent and continuous spaces, masses that arise in one space can easily spread to the other. the tumors of the infratemporal fossa are uncommon and rarely, they may arise from within the infratemporal fossa itself. endoscopic access to this area often begins with surgical access to the pterygopalatine fossa, with more lateral dissection allowing entry to the infratemporal fossa. Schwannomas are very uncommon in children (except when associated with neurofibromatosis type 2. The advantages of the endoscopic technique are numerous, including Relatively straightforward access is achieved to the pterygopalatine fossa and infratemporal fossa, areas traditionally considered difficult to reach.

Case Description: A.Q.K, an 8-year-old boy, presented with a three-month history of right eye painless protrusion, blurring of vision and a one-week history of swelling over the right cheek. On examination; There was a noticeable right-sided facial swelling with right eye protrusion, globe displacement Antero superiorly, A radiological examination showed large soft tissue mass involved Right pterygopalatine fossa & infratemporal fossa, Histopathological examination confirmed the diagnosis of schwannoma Complete excision was achieved using modified endoscopic danker's approach.

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conclusion: Right. Maxillary sinus. The post-operative period was uneventful.

The target for our case report is highlighting the rarity of this lesion (Infratemporal schwannoma of the mandibular branch of the trigeminal trunk in an 8years old boy), Emphasizing the significance of an accurate diagnosis and including this tumor in the differential diagnosis of facial asymmetry.

The endoscopic technique provides a magnified and multiangled view for more precise discrimination of the dissection planes between the tumor and the adjacent structures. This technique obviates the need for external facial incisions or intra-oral incisions and helps in good surgical outcomes and quick healing and recovery.

Keywords: Schwannoma, infratemporal fossa, Pterygopalatine fossa, Endoscopic denker's excision.

Introduction

SCHWANNOMAS

- A peripheral nerve's axon, a lengthy projection of the neuronal cell body with a specific function of conveying nerve impulses, is its principal structural component. Schwann cells encircle each axon, wrapping around in several layers to produce the protective myelin sheath.
- Each axon and its Schwann cells are surrounded by a layer of loose connective tissue called the endoneurium, which forms a nerve fiber.
- A neural fascicle is made up of several nerve fibers that are encased in strong connective tissue, called the perineurium.
- The epineurium, a dense, irregular connective tissue that envelops every nerve fascicle, forms a peripheral nerve.
- Neoplasms made up of cells exhibiting nerve sheath differentiation are known as nerve sheath tumors. They account for 10% to 12% of all neoplasms in soft tissues.
- Schwannomas are slowly developing neoplasms classified as benign nerve sheath tumors by the World Health Organization in 2013. They are made up only of Schwann cells embedded in a collagenous matrix, and they start in a nerve.

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EPIDEMIOLOGY

Approximately 5% of benign soft tissue neoplasms are schwannomas.
o Although they are rare in youngsters, they can happen to anyone at any age. According to the majority of published data, schwannomas are most common in people between the ages of 20 and 50, peaking in the third and seventh decades of life. The effects are the same for males and women. Schwannomas linked to neurocutaneous disorders typically affect younger individuals.

TOPOGRAPHY

o the head, neck, and limbs' main nerve trunks are most frequently affected by schwannomas. Consequently, the cervical plexus, vagus nerve, and spinal roots are the topographical sites of schwannomas that occur most frequently. Schwannomas have been reported less commonly in the tongue, mouth, and larynx.

HISTOLOGY

GENERAL HISTOLOGIC FEATURES

o They are typically firm, bounded, and enveloped in epineurium. Schwannomas cause the nerve's regular components to shift to one side as they grow eccentrically. Given that schwannomas may be separated from the underlying nerve fibers, it follows that surgical removal can typically preserve the parent nerve.
o grossly, the sliced surface appears tan or gray, with sporadic yellow patches and cysts, especially in larger tumors. Schwannomas are less frequently (partially) hemorrhagic. While larger tumors might be oval, sausage-shaped, or irregularly lobulated, smaller schwannomas are often spheroid.

In schwannomas, two different tissue types can be identified under a microscope: The Antoni Tissue is made up of densely packed cells that are frequently grouped in rows and divided by distinct hyaline bands, a pattern known as Verocay bodies. A less cellular

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Antoni B pattern, in which the tumor cells are divided by a soft honeycombed eosinophilic matrix, is typically present in all or part of a schwannoma. Schwannoma vessels are typically susceptible to spontaneous thrombosis, which can result in necrosis and occasionally bleeding in the surrounding tissues. The frequently severe amplification of schwannomas on imaging tests is indicative of their robust vascular supply.

Schwannoma histologic characteristics can be summed up as follows and may be indicative of their sporadic non-uniform appearance on computed tomography (CT) or magnetic resonance imaging (MRI) exams:
The regions with lower cellularity (Antoni B) next to those with higher cellularity (Antoni A)
Degeneration of the cystic wall brought on by arterial thrombosis and the necrosis that follows Xanthomatous areas, which have clumps of foamy, lipid-filled cells.

CLINICAL PRESENTATION

o small sized schwannomas typically don't cause any symptoms. When the tumor grows to a size where it compresses the affected nerve or other structures, symptoms such as pain, paresthesias, muscular atrophy, or other problems may manifest. They can be moved side to side during a physical examination, but not along the nerve's long axis.

IMAGING CHARACTERISTICS

COMPUTED TOMOGRAPHY

o Schwannomas appear as well-circumscribed, homogeneous lesions on unenhanced CT images. In comparison to muscle, they are hypo- to isodense. Larger tumors often have an inhomogeneous look with low-density patches.
o Most schwannomas become iso- or hyperdense to muscle on contrast-enhanced CT scans; big schwannomas usually contain non-enhancing cystic or necrotic regions.

MAGNETIC RESONANCE IMAGING

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Schwannomas might present as harder to detect due to their somewhat higher signal intensity on T1-weighted imaging (T1-WI) compared to muscle.
o They are hyperintense to muscle on proton density-WI.
o There is a noticeable increase in signal intensity on T2-WI, which makes the tumor and the surrounding fat and muscle stand out sharply. (1) First of all

SURGICAL ANATOMY

Because the infratemporal and pterygopalatine fossas are contiguous and connected areas, masses that form in one can readily transfer to the other.
o The endoscopic method has many benefits, such as relatively easy access to the pterygopalatine and infratemporal fossas, which were previously thought to be difficult to reach. (2) o The infratemporal fossa tumors are rare and infrequently, they can originate from the fossa itself. (3) o Surgical access to the pterygopalatine fossa is frequently the first step towards endoscopic access to this region; further lateral dissection grants entry to the infratemporal fossa. (4) o Schwannomas are quite rare in children, except for those connected to neurofibromatosis type 2. (5)

A.Q.K, an 8-year-old boy, presented to an ENT specialist with a three-month history of *right* eye painless *protrusion*, *blurring of vision*, and a one-week history of *swelling over the right cheek.* There was no history of associated trauma, and he had not experienced any sinonasal or dental symptoms. His past medical history, surgical, and drug history were none.

On examination; There was a noticeable right-sided facial swelling that was smooth and non-tender, with right eye protrusion, *globe displacement anterosuperiorly*, no widening of medial canthus space, (see Fig.1). Endoscopically examination was not done as the child was irritable. Examination of the oral cavity was unremarkable. Ophthalmological exam.; Optic disc was pale and compromised by tumor, very poor visual acuity. There was no focal neurological deficit

On imaging; CT scan and MRI were done for the patient with the following results: (see Fig.2,3,4,5 and 6).

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On histopathology study: Sections showed hypercellular area (Antoni A) composed of a proliferation of spindle cells with nuclear palisading alternating with the myxoid area (Antoni B) (see Fig.7).

On immunohistochemical stain: S100……. positive. (see Fig.8, 9)

On surgical treatment: He underwent modified endoscopic Denker's approach under general anesthesia with oral endotracheal intubation, after scrubbing and draping the patient with supine position and head elevation, started with decongestant packs for nasal mucosa preparation, endoscopic examination of nasal cavity and mass assessment, after that began to do middle meatal antrostomy, cauterization and shaving all the soft tissues on the lateral nasal wall (medial wall of rt. Maxillary sinus) then elevate of periosteal flap from pyramidal aperture to exposed the ant. Maxillary wall and remove it, after remove the medial wall of the maxillary sinus with Sharpe cutting of NLD and partial resection of the middle turbinate, continued to remove the posterolateral maxillary wall then reached to PPF and ITF and started to remove all parts of tumor (sphenoidal, orbital, and ITF part) then hemostasis was done and cover the raw area by surgical. Resection of the middle turbinate and inferior turbinate with wide middle meatal antrostomy was done. The specimen was sent for histopathological examination which revealed a neoplasm of benign schwannoma. The patient had an uneventful post-operative recovery.

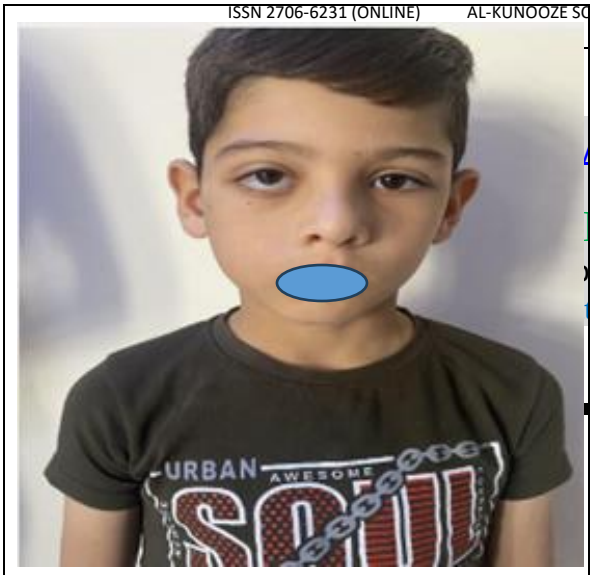

Figure 1: right-sided facial swelling with right eye protrusion, globe displacement anterosuperiorly.

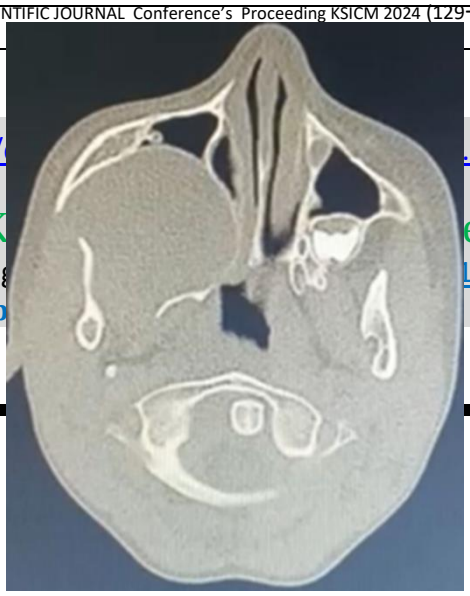

Figure 2: native CT scan, the bone window showed soft tissue density mass, well-demarcated, homogenous with bone remodeling involved Rt. ITF and widening foramen ovale (red ring) with pushing Rt. Maxillary sinus anteriorly.

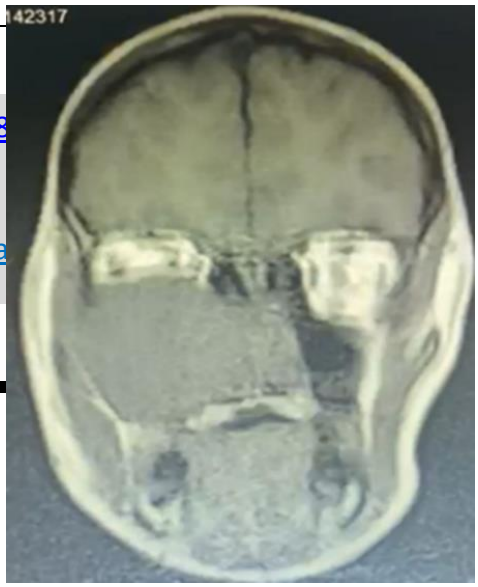

*Figure 3: T1WI MRI; showed a soft tissue intensity (47*48*51mm) involved Rt. PPF & ITF with extension to the orbit and sinonasal area.*

Figure 1: right-sided facial swelling with right eye protrusion,

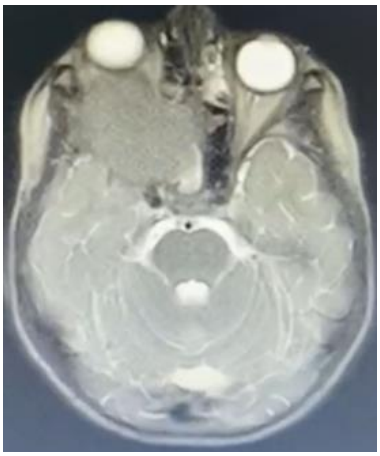

Figure 4: T2WI MRI; Showed A Soft Tissue Intensity (Intermediate Signal) Mass Involved Rt. Infraorbital Components

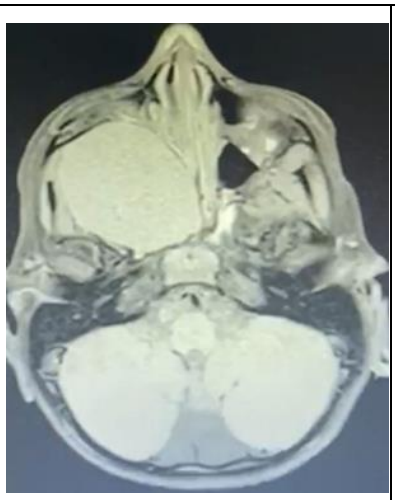

Figure 5: t1wi MRI with gadolinium, the mass was enhanced.

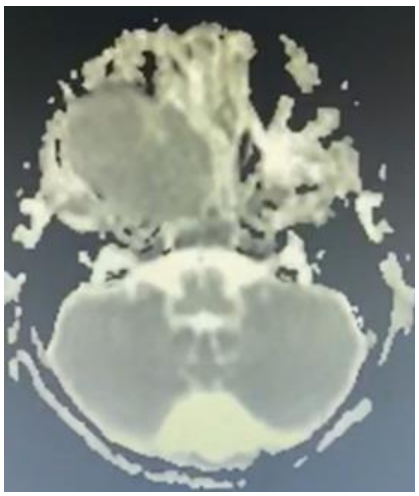

Figure 6: diffusional weighted image MRI; showed hypointense area indicated tumour rather than inflammatory lesion.

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FIGURE 2: NATIVE CT SCAN, BONE WINDOW SHOWe

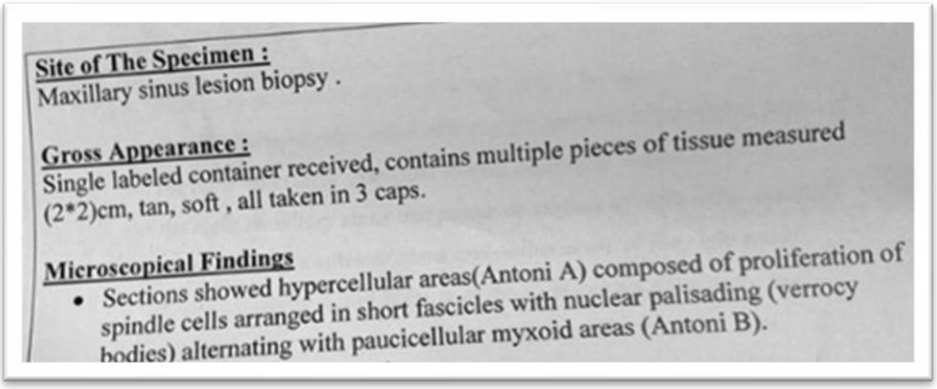

Figure 7: histopathology study showed; figuration of hypercellular area (Antoni a) with myxoid area (Antoni b).

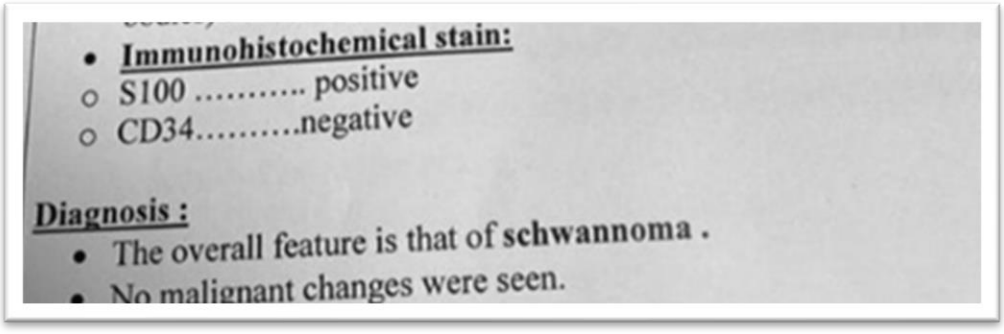

Figure 8: immunohistochemical stain showed result of +ve s100.

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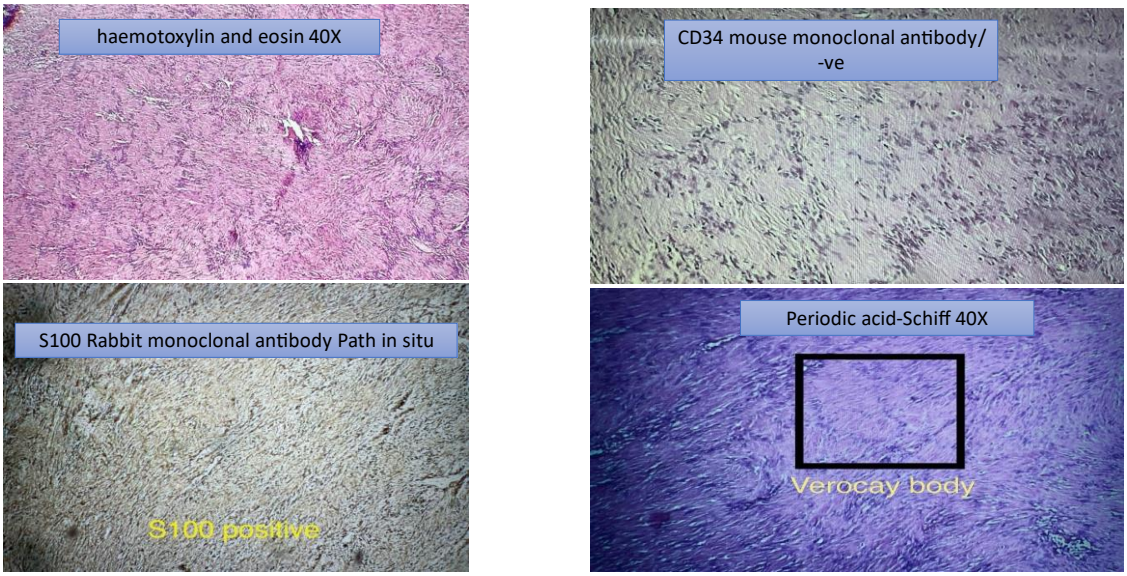

Figure 9: immunohistochemical stain showed result of +ve s100.

DISCUSSION

Schwannomas, most commonly involve the ***major nerve trunks*** of the head and neck, the ***most common*** topographical **locations** of schwannomas include the ***spinal roots, cervical plexus and the vagus***, represent approximately 5 % of all benign soft tissue neoplasms. They can occur at all ages, though they are ***uncommon in children***. most literature data indicate that schwannomas are most prevalent between the ages of 20 and 50, with peak incidences in the third and seventh decades. Men and women are equally affected. Regarding our case, the schwannoma was affecting the division (mandibular) of trigeminal nerve trunk, involving the foramen ovale in infratemporal fossa and the age was only

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8years old in boy child. From 2020-2024, there was just one published case report of pediatric schwannoma, Hua Cai et al; (2024) present a case of a 6-year-old girl child exhibiting a sizable soft tissue mass in the left PPF, extending into the inferior orbital fissure, with a recent physical examination indicating reduced vision in the left eye accompanied by a central dark spot for 1.5 months.[6]

While the others: Cao, Zhiwen et al, (2024) received a 69-year-old male patient who had been experiencing headache, dizziness, and numbness from the mastoid region of his left ear to the corner of his mouth for a duration of 22 days.[7] Agosti, E et al, (2022) received 16-year-old male presented with right-sided ear fullness, hearing loss, and nasal congestion, No focal neurological defects, with Rt. giant mandibular schwannoma of the ITF.[8] Samal S et al, (2020) present A 65-year-old male presented to the outpatient department with a progressive swelling over the left parotid for 5 years and pain during chewing for 6 months which was diagnosed as benign spindle cell tumor on cytology.[9] LIU, James K., et al,(2020) demonstrate a case of a 44-year-old female with a giant ITF schwannoma with intracranial extension and erosion of the central skull base.[10] Wendell Dwarika et al,(2021) demonstrate a case of a 46-year-old male who presented with a twelve month history of left-sided facial pain and progressive swelling. Contrast-enhanced computed tomography (CT) and magnetic resonance imaging (MRI) revealed a large soft tissue lesion located in the left pterygopalatine fossa (PPF).[11] Saifi, A.M.et al, (2023) both patients had a history of gradually increasing swelling, the first originating from the Sino-nasal region and the second from the temporal/ infratemporal region.[12] Prabha BB et al,(2020) published a case report of a 26-year-old lady who presented with *pain in the left cheek* for 6 months, she underwent an endoscopic trans nasal trans maxillary approach to the pterygopalatine fossa, and the specimen was sent for histopathological examination which revealed a neoplasm of benign schwannoma. [13]

With all those authors and their kinds of literature from 2020-2024, it was documented that our case represented as a rare presentation regarding the age of the patient who was only 8 years old, and uncommon site of ITF with involvement of mandibular root of trigeminal nerve schwannoma.

CONCLUSION

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The target for our case report is highlighting the rarity of this lesion (Infratemporal schwannoma of the mandibular branch of the trigeminal trunk in an 8years old boy), Emphasizing the significance of an accurate diagnosis and including this tumor in the differential diagnosis of facial asymmetry .The endoscopic technique provides a magnified and multiangled view for more precise discrimination of the dissection planes between the tumor and the adjacent structures. This technique obviates the need for external facial incisions or intra oral incisions and helps in good surgical outcomes and quick healing and recovery.

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