Pediatric intraosseous schwannoma in maxillary sinus: A case report with review of literature

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Abstract

Neurilemmoma has been defined as a benign, encapsulated neoplasm that arises in the nerve fiber. It originates from the proliferation of Schwann cells in the perineurium causing displacement and compression of the adjacent nerve. This neoplasm is composed primarily of Schwann cells in a poorly collagenized stroma. It can occur in any age group. Neurilemmoma occurs all over the body including the head and neck region. In the head and neck region, 25%–40% of schwannoma cases have been reported. Occurrences of intraoral schwannomas are rare with reported prevalence being 1%. In the present article, we report a case of a 19-month-old baby complaining of pain and swelling. On clinical, radiological and histopathological features it was diagnosed as pediatric intraosseous schwannoma of the maxillary sinus.

Keywords: Neurilemmoma, pediatric jaw lesions, schwannoma

INTRODUCTION

Schwannoma (neurilemmomas) are benign, usually solitary, encapsulated neoplasm’s that arise from the Schwann cells of the nerve sheath derived from the neuroectoderm.¹ Schwann cells are supporting cells of Periperal Nervous System which in turn develop from neural crest cells. They produce a lipid rich layer called the Myelin Sheath that surrounds the axons. The myelin sheath isolates the axons from surrounding extracellular compartment of endoneurium. The conduction of nerve impulses is faster in myelinated axon from one node of ranvier to other by salutatory conduction. There are two types of schwannomas (a) Peripheral: Located in the soft tissues and (b) Central (intraosseous): Located within the bone.²

In the present article a case report of a 19-month-old baby presented with an innocuous mild swelling of the maxilla which was later, on radiographic and histopathological feature examination was diagnosed as central type of schwannoma of the maxillary sinus.

CASE REPORT

A child of 19-month along with her parent, presented to the outpatients clinic of GITAM Dental College and Hospital with the chief complaint of pain and swelling since 2 months. The swelling initially started small and gradually enlarged to the present size. The swelling was not associated with paresthesia, pus discharge or any other secondary changes. On extra oral examination the face was asymmetrical with mild diffused swelling on middle
one-third of face below the infraorbital margin on the left side. On intraoral examination, swelling was seen in the labial mucosa extending from 62 to 65 region with obliterated vestibule.

On extraoral examination of the specific lesion revealed an ill-defined diffused swelling seen on the middle third of the face in the region of zygomatic buttress below the infraorbital margin. This was round to oval in shape measuring about 3 cm × 3 cm in size with smooth surface. Extending anteroposteriorly from ala of nose to 3 cm away from tragus of ear. Superior-inferiorly from the floor of orbit to the alveolar process of maxilla [Figure 1]. The color and skin temperature was similar to that of surrounding.

On palpation, it was mild tender and firm in consistency. The regional lymph nodes were not palpable. A provisional diagnosis of fibrous dysplasia was made. Following axial and coronal computer tomography (CT) and incisional biopsy was performed under general anesthesia.

**Radiographic features**
CT scan of nose and paranasal sinuses showed a mass in the left maxillary antrum [Figure 2]. In the coronal CT scan, a mass in the left maxillary sinus eroding the floor of the orbit giving the mass a dumb-bell shape [Figure 3]. Cone beam CT shows excessive bone destruction extending into floor of orbit into ethmoid sinus and maxillary sinus [Figure 4].

**Histopathological examination**
The hematoxylin and eosin stained soft-tissue section revealed a capsulated circumscribed Schwann cells arranged in short bundles or fascicles. They are arranged in a radial pattern below a ciliated stratified squamous epithelium resembling the maxillary sinus lining. The tissue also exhibited variable amounts of Antoni A and hypocellular Antoni B areas with horizontal rows of palisaded nuclei separated by acellular rows of eosinophilic processes resembling verocay bodies [Figure 5]. Areas of cystification were also seen [Figure 6]. Other section of slide shows cystic spaces lined by basophilic cells resembling ductal or glandular pattern [Figures 7 and 8]. The hematoxylin and eosin stained soft-tissue section also exhibits areas of reactive bone formation and Hyalinization [Figure 9].

Overall radiographic and histopathological features are suggestive of ANCIENT TYPE OF SCHWANNOMA with pseudoglandular changes.

**DISCUSSION**
Schwannoma (neurilemmoma, neurinoma, and perineural fibroblastoma) benign neural tumor usually solitary with slow growth rate. It was first described by Verocay in 1908. Schwannomas generally develop in all body
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Figure 4: Cone beam computed tomography shows excessive bone destruction extending into floor of orbit

Figure 5: Verocay body showing horizontal rows of palisade nuclei separated by acellular rows of eosinophilic processes

Figure 6: Cystification with areas of ciliated cells in the lower boarder

Figure 7: Large foamy histiocytes with fibrillary stroma

Figure 8: Contains cystic spaces lined by basophilic cells resembles the ductal or glandular pattern

Figure 9: Round to oval basophilic cells are radially oriented in the collagenous stroma

Antoni A with verocay bodies

Area of cystification

Xanthomatous change

Radial cellular arrangement

Schwannoma also reported in ethmoidal sinus, maxillary sinus, nasal fossa and sphenoid sinus.\[9\]

Three mechanisms of bone involvement: (a) arising within bone, (b) arising within the nutrient canal and (c) arising from the periosteum.\[10,11\] Maxillary schwannomas are extremely rare only 13 cases reported in literature in post 70 years.\[7,11,12\]

The clinical features of all those cases together with the current one has been summarized in the Table 1.

parts, head and is the most common site which accounts 25%–45% of all benign schwannomas.\[5\]

In the oral cavity tongue is the common sit followed buccal mucosa, floor of the mouth, palate and lip.\[6\] On the contrary; intraosseous (central) schwannomas are uncommon in head and neck area constituting <1%. Mandible is also most favored site due the inferior alveolar nerve supply and its long course within the jaw.\[7,8\]
Schwannomas develop from any peripheral, cranial or autonomic nerve that has a Schwann sheath. In sinonasal schwannomas, Schwann cells trigeminal nerve mainly ophthalmic and maxillary branches.\[26-28\] Oral schwannomas are usually present in the soft tissue but may have clinical features similar to other benign lesions such as mucocele, fibroma, neurofibroma, lipoma and benign salivary gland tumor.

Radiographically, intraosseous schwanna occurs as well demarcated, unilocular radiolucency with additional features such as external room resorption, cortical plate thinning, cortical expansion, and peripheral scalloping can be seen. As a diagnostic tool, ultrasonography, computed tomography and magnetic resonance imaging (MRI) help in estimating the tumor margin as well as infiltration into the surrounding structures. However, the present case, it was intra osseous variant with margins extending into the nasal and floor of the orbit.

In our case study, histopathologically Schwannomas characterized by a mixture of two patterns of tissue growth, namely, Antoni type A and B. Antoni A type with...
verocay bodies. The cystic spaces lined by basophilic cells resembling ductal or glandular pattern. Xanthomatous change forming large foamy histiocytes in fibrillary stroma and areas of cystification are also seen. Reactive bone formation and Hyalinization is observed. Ancient schwannoma shows degenerative changes such as cystic, myxoid, edematous and fibrotic areas. Vascular abnormalities and nuclear pleomorphism can also be seen. Cellular schwannomas can be classified on microscopic examination and immunostaining. It differs from classic schwannomas by its cellularity, nuclear pleomorphism and hyperchromatism, the absence of verocay bodies and increased mitotic activity.[29]

Histologic criteria to differentiate schwannoma, cellular schwannoma and malignant peripheral nerve sheath tumor were previously described and are summarized [Table 2].[30,31]

As shown in Table 2 and overall histopathological features came to final diagnosis as ANCEINT/CONVENTIONAL TYPE OF SCHWANNOMA DIAGNOSTIC CRITERIA FOR SCHWANNOMATOSIS.

Molecular diagnosis
1. Two or more schwannomas and genetic studies of at least two tumors showing loss of heterozygosity at chromosome 22 and NF2 mutation. The presence of a common SMARCB1 mutation defines SMARCB1-associated schwannomatosis
2. One schwannaoma and germ line pathogenic SMARCB1 mutation.

Clinical diagnosis
1. Two or more nonintradermal schwannomas and absence of vestibular schwannoma on MRI
2. One schwannoma affected first degree relative
3. Possible diagnosis if two or more nonintradermal schwannomas and chronic pain associated with tumors
4. Exclusion criteria
5. Germ line pathogenic NF2 mutation, fulfill criteria for NF2, first-degree relative with NF2, schwannomas in radiation field only.

The treatment of choice is pericapsular excision. The schwannoma should be extirpated in its entirety to avoid tumor recurrence even if the nerve of origin cannot be preserved.[9] The prognosis is very good and malignant transformation is rare although some authors have mentioned a malignant transformation rate of 8%–13.9%.[30]

CONCLUSION
The youngest patient that has been documented was just 10 years in the literature. To the best of our knowledge, this is the only case reported in a 19 months old baby. Upcoming treatment option like CO2 laser surgery is used to decrease complications.

Declaration of patient consent
The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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Conflicts of interest
There are no conflicts of interest.

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