CASE REPORT

Rare occurrence of intraosseous schwannoma in a young child, its review and its pathogenesis

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ABSTRACT

Schwannoma is a benign, encapsulated, perineural tumor that arises from the Schwann cells. Approximately 25% of the reported cases originate from the head and neck region. Of these, approximately 1–12% occurs intraorally. The intrabony lesion accounts for less than 1% of the central neoplasms. We report a rare case of intraosseous schwannoma in an 8-year-old male patient characteristically originating from the mental nerve. Radiographic examination followed by histopathological evaluation was further confirmed by immunohistochemical markers, S-100 protein, and GFAP that stained intensely positive for the tumor. Thus, confirming the diagnosis of intraosseous schwannoma.

Key words: Antoni type A, Antoni type B, immunohistochemistry, intraosseous schwannoma, neural tumor, S-100 protein

INTRODUCTION

The general organization and distribution of nervous tissue (the third basic tissue) are to a large extent determined by its course of evolution. The specialized cells that constitute the functional units of nervous system called neurons supported by their connective tissue neuroglia constitute the main bulk of the nervous tissue in the body. Neurons are derived in embryogenesis from primitive neuroblasts. They are terminally differentiated cells that do not regenerate in the event of cell death. Any disorder to these, leads to devastating benign or malignant tumors of the nerves in the oral cavity.[1]

Virchow's original classification emphasized the relationship of the nerve tumors to the neuron proper by dividing the tumors into true and false neuromas. False neuromas comprised nerve sheath tumors (e.g., neurofibroma, neurilemoma). This group has been accepted as the more common and significant group of tumors of peripheral nerves and it is this group that forms the principal focus. Despite the common nature of peripheral tumors, they continue to pose problems in classification and nomenclature.[2]

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Neurilemmoma also referred to as Schwannoma, neurinoma, and perineural fibroblastoma is an encapsulated nerve sheath tumor.[2] A benign neoplasm derived from Schwann cells was first described by Verocay in 1910. He called it "Neurinoma" then. In 1935, the term "Neurilemmoma" was coined by Stout.[3] Eversole and Howell[4] reported the first intraoral case of ancient schwannoma, although Cherrick and Eversole[4] presented two additional cases in the same year but detailed clinical information was not given. Although the occurrence of schwannomas in the head and neck area is relatively high, an intraosseous schwannoma is rare, presenting in less than 1% of benign primary bone tumors.[5]

Schwannoma is usually a solitary soft tissue or intrabony lesion that is slow growing, encapsulated and is often associated with the associated nerve attached peripherally.[6] Oral schwannomas are found to exhibit two types. The common type is the submucosal module, which is encapsulated, well defined, firm in consistency, thus resembling a cyst. The second type is the nonencapsulated, where the tumor is found below the basal layer of the mucous membrane.[7]

The preoperative diagnosis is often difficult and is made by computed tomography and/or magnetic resonance imaging to evaluate the extent and determine infiltration of the surrounding structures.[8] If the nerve of origin is identified, as in the case described by Yamazaki *et al*.[9] of schwannoma associated with the mental nerve, it is possible to diagnose a peripheral nerve sheath tumor, which at least allows the patient to be given appropriate information regarding the risk of nerve lesion during surgery. Kun *et al*.,[10] were able to

make a correct preoperative diagnosis in only four cases located in the neck of the 49 cases studied, they concluded that it is very difficult to make the diagnosis based on diagnostic imaging techniques. Kawakami *et al*.,[11] in the case of a tumor in the floor of the mouth, with computed tomography and magnetic resonance a malignant tumor of the sublingual gland. A differential diagnosis is needed with other neurogenic tumors such as neurofibroma, neuroma, myoblastoma of granular cells, neurogenic sarcoma, malignant schwannoma, neuroepithelioma, and melanoma.[12]

In this article a rare clinical case of intraosseous schwannoma is reported whose diagnosis was established upon clinical, histological, and imunohistochemical findings.

CASE REPORT

An 8-year-old male child patient named Binod murmu reported to the department of oral and maxillofacial pathology with a chief complaint of swelling in the lower left region of the face since 1 year.

No contributory medical, family, or personal history was elucidated.

On extra-oral examination, patient presented a diffuse, firm, nontender, nonpulsatile, nonreducible, noncompressible, nonfluctuant swelling in the lower left one third of face [Figure 1].

Intra-oral examination revealed a smooth, firm, nontender, nonpulsatile, nonreducible, noncompressible, nonfluctuant swelling with ulcerated surface (due to trauma from antagonist teeth) extending from lower left lateral incisor to first permanent molar. Expansion of both buccal and lingual cortical plates and mobility of regional teeth was also present [Figure 2].

The following investigations were performed: Aspiration, routine hemogram, radiology, light microscopy, and immunohistochemistry.

An x-ray OPG of jaws revealed mixed dentition with teeth in various stages of development and eruption and root resorption of 73, 74, 75, displacement of permanent tooth buds 33, 34, 35, extrusion of 73, 74, 75 and increased vertical height of lower left side of mandible. The radiograph presented a large round unilocular radiolucent area with moderately defined borders [Figure 3].

All routine blood examination was conducted before the surgical procedure.

An incisional biopsy was performed under LA followed by the primary closure of the wound with 3–0 silk suture. The excised tissue submitted to the department of oral pathology for histopathological examination. A differential diagnosis of neurofibroma, benign fibrous histiocytoma, FIbro sarcoma, leiomyoma.

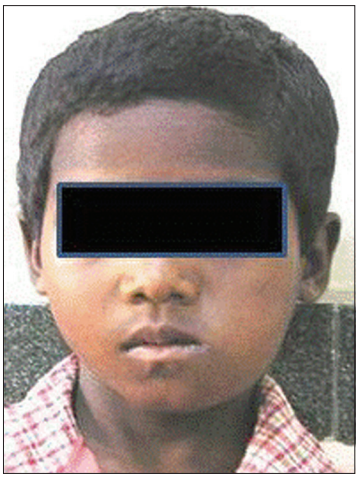

Figure 1: Photomicrograph showing swelling in the lower left region of the face

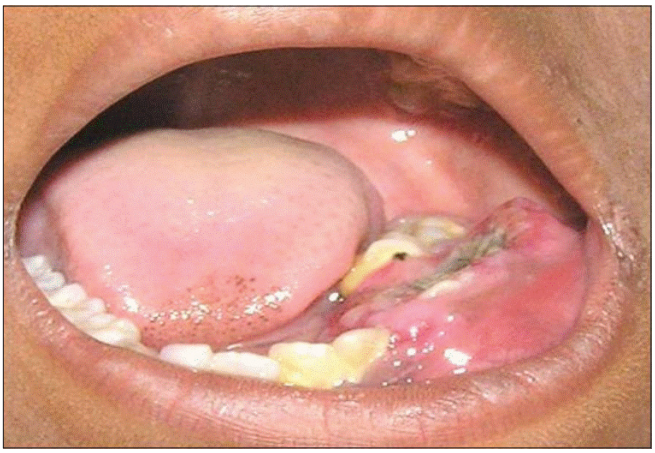

Figure 2: Photograph showing swelling from lower left lateral incisor to first permanent molar

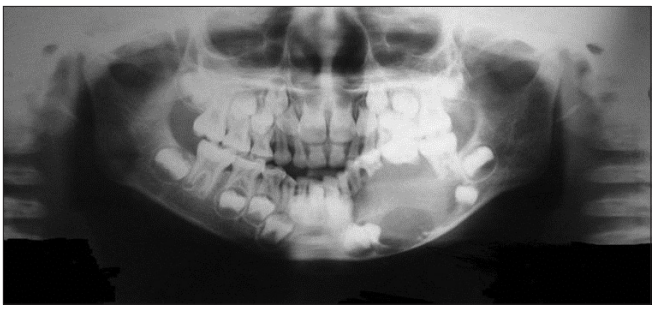

Figure 3: An orthopantomograph revealing a large round unilocular radiolucent area with moderately defined borders

The histopathology of the submitted H and E stained sections showed a thin fibrous capsule with Antoni A type and Antoni B type tissue arrangements. Antoni A type being characterized by Schwann cells that are closely packed, arranged in bundles or rows with palisading nuclei. While in the Antoni B

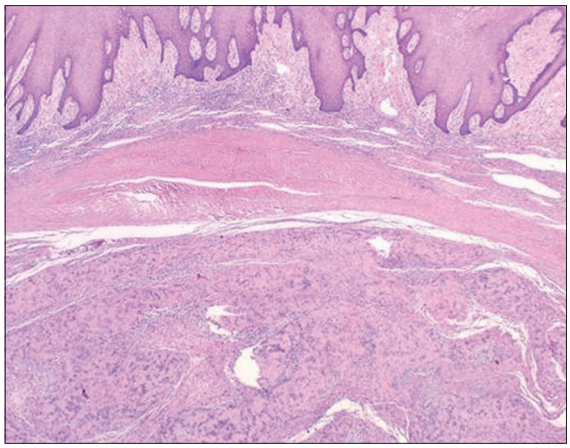

Figure 4: Photomicrograph showing a well circumscribed tumor showing a thin well-defined fibrous connective tissue capsule and proliferating schwann cells (H and E, ×4)

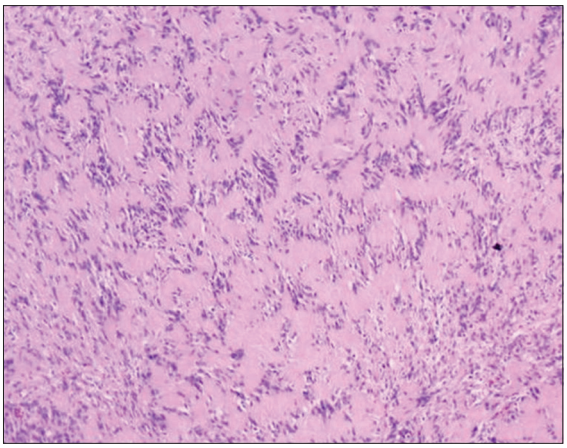

Figure 5: Photomicrograph showing proliferating schwann cells arranged in a palisading pattern (H and E, ×10)

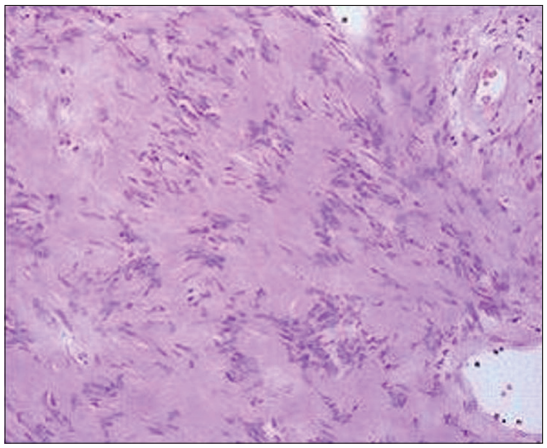

Figure 6: Photomicrograph showing Antoni type A cells interspersed with some verocay bodies (H and E, ×40)

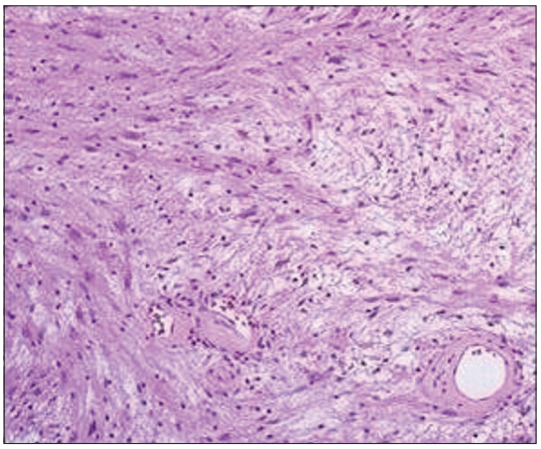

Figure 7: Photomicrograph showing Antoni type B loosely arranged tumor tissue with no characteristic palisading appearance

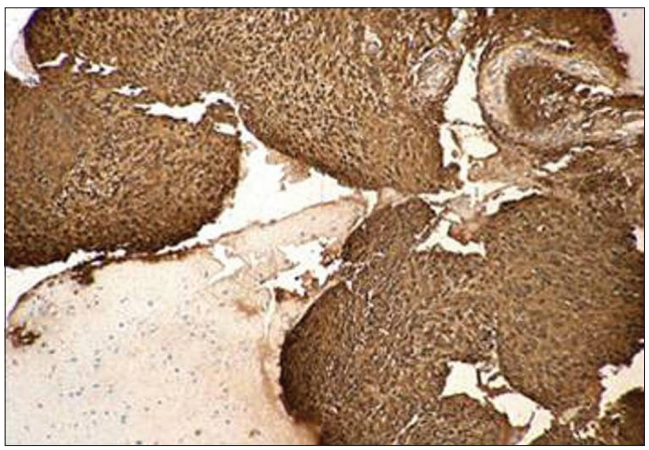

Figure 8: Tumor cells shows positive stain for S-100 protein

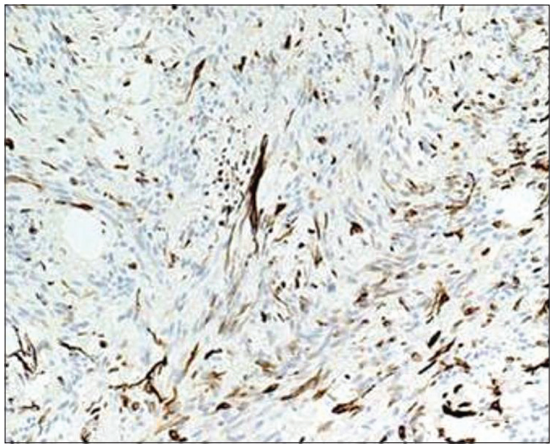

Figure 9: Tumor cells shows positive stain for glial fibrillary acidic protein

type, there is less number of cells and disorganization of fusiform cells dispersed in a loose and random fashion. Free bands of amorphous substance between the rows of nuclei constitute the Verocay bodies. The vasculature is not that prominent.

Considering the nonorganization of the tumoral Schwann cells forming the Verocay bodies, immunohistochemical analysis was done using S-100 and glial fibrillar acidic protein (GFAP) that stained intensely positive [Figures 4–9].

Table 1: Reported cases on intraosseous schwannoma in the oral cavity

Reference	Age	Gender	Location	Size(cm)	Duration	Follow-up
Kusama *et al.*	31	Female	Mandible	2.5	Unknown	NED (12 months)
Barrett *et al.*	53	Male	Mandible	1	N/A	NED (9months)
Morton	58	Male	Mandible	N/A	N/A	N/A
Koutlas *et al.*	14	Male	Anterior mandible	2	N/A	2 years
Koutlas *et al*.	24	Female	Posterior mandible	1.1	Unknown	10 years
Koutlas et al	48	Female	Posterior mandible	2	1 Year	2years then LF

On the basis of clinical, radiological, and histological features a provisional diagnosis of Schwannoma was given.

Immunohistochemistry was done S-100: Positive, glial fibrillary acidic protein: Positive, smooth muscle actin: Negative, desmin: Negative.

Confirmative diagnosis of intraosseous schwannoma was given.

Management of the lesion was done by surgical resection along with preservation of nerve and on follow up no malignant transformation was reported.

DISCUSSION

Schwannoma, is a benign tumor derived from the peripheral nerve sheath. It is a rare, benign neural tumor arising from the neural sheath Schwann cells of the peripheral, cranial, or autonomic nerves.[13,14] It can also arise from any nerve covered with a Schwann cell sheath, which include the cranial nerves (except for the optic and olfactory), the spinal nerves and the autonomic nervous system.[14] The origin of the Schwann cell has also long been debated. Some say that they arise directly from the neural tube whereas others insist their origin from the neural crest, (a group of cells that lie lateral to the neural tube and beneath the ectoderm of the developing embryo).[2] Some put its etiology to originate from a proliferation of Schwann cells in the perineurium causing displacement and compression of the adjacent nerve.[15]

In the parapharyngeal space, it usually derives from the vagus nerve and the cervical sympathetic chain. It does not arise from cranial nerves I and II because they lack Schwann cells.[16] The tumor is solitary, with a smooth surface and slow asymptomatic growth although the clinical symptomatology depends on the nerve of origin.[15]

Two types are distinguished (central and peripheral schwannoma) located in bone or in soft tissues respectively.[15] Approximately 25% of the reported cases originate from the head and neck region.[14] Of these, approximately 1–12% occur intraorally. The intrabony lesion accounts for less than 1% of the central neoplasms.[16] In our case, the tumor was intraosseous suspected to have a close association of its origin with the mental nerve. In the past reviews, very few cases of intraosseous schwannomas in the oral cavity have been reported [Table 1].

According to the literature, there have been reports of 44 cases of intrabony neurilemmomas that occurred in either of the jaws. Neurilemmomas of the jaw occurred in the age range of 8–72 years, with the average age of 34 years and a definite female predilection.[17] Contrary to the above literature, ours is one such unusual case cited in the first decade in very young male patient of 8 years.

The radiographic appearance of a well-defined unilocular nonspecific radiolucent lesion, with root divergence (expansive growth) and root resorption only in teeth contacting the lesion, was suggestive of a benign process and thus making a preoperative diagnosis was difficult. The possibility of an intraosseous schwannoma was not considered at first because of the extreme rarity of this location. Along with this, an association with the mental nerve was also present. Radiographic finding from four different sites may suggest three mechanisms by which schwannomas may involve bone: 1) a tumor may arise centrally within the bone, 2) a tumor may arise within the nutrient canal and produce canal enlargement, or 3) a soft tissue or periosteal tumor may cause secondary erosion and penetration into bone.[18] In the case we hereby report, the third mechanism can be excluded because there was no cortical bone perforation of intraosseous schwannomas. Imaging of the lesion comprises of computed tomography (CT) and magnetic resonance imaging (MRI) as it not only determines the extent of the lesion and assesses its resectability but delineates its relationship to the artery, nerve, skull base, or intracranial involvement also.[19] If the nerve of origin is identified, as in the case described by Yamazaki *et al*. of schwannoma associated with the mental nerve, it is possible to diagnose a peripheral nerve sheath tumor, which at least allows the patient to be given appropriate information regarding the risk of nerve lesion during surgery.[12]

However, a definite diagnosis is difficult to make on the basis of radiography alone; a biopsy is usually necessary. Schwannomas only rarely arise in the inferior alveolar canal, but when they do, such lesions become rounded and the wall of the mandibular canal is preserved.[20] On the other hand, neurofibromas tend to grow specifically in the canal, which typically becomes ovoid shaped.[21] The recurrence rate of schwannoma is the same as that of neurofibroma.[21] The

differentiation between these two neoplasms is imperative because neurofibromas have the potential for malignant transformation.[22]

In our case, differential diagnosis was made with neurofibroma, benign fibrous histiocytoma, fibro sarcoma, leiomyoma.

The histological findings of the present case are similar to those reported previously, consisting of a thin fibrous capsule and a tumoral proliferation formed by two types of tissue arrangements: Antoni type A and Antoni type B.[14] This histological picture is dominated by an encapsulated lesion arising from a nerve end composed of an intimate mixture of spindle cells forming highly cellular Antoni A and less cellular, myoxid Antoni B areas.[4] Histopathologically, five schwannoma variants have been described: Common, plexiform, cellular, epithelioid, and ancient schwannomas.[2] In the Antoni A region, nuclear palisading and homogeneous acellular zones known as verocay bodies are sometimes noted. Although verocay body and nuclear palisading have traditionally been linked to neural differentiation, they have been described in association with many other lesions and neoplasms, some of them involving the skin.[23] Hence, to confirm the histogenesis of this lesion we went ahead with the immunohistochemical analysis for the same which revealed the schwannoma cells to be positive for the protein S-100, a marker for the nervous system.

However, the histological differential diagnosis is made with other neural origin lesions, which could be neurofibroma and neuroma or muscular or fibroblastic origin tumor.[24,25] Schwannomas usually present as a solitary lesion. When multiple, however, they can be associated with neurofibromatosis. The differentiation between schwannoma and neurofibroma is essential because an apparently "solitary" neurofibroma may be a manifestation of neurofibromatosis.[26]

Because it is a well-encapsulated lesion, the treatment of choice for schwannomas is the conservative surgical enucleation with periodic follow-up. Recurrence is uncommon.[27] In this case, the patient has been followed up for four years with no clinical or radiographic signs of lesion recurrence. The slow (3 years) and expansive growth (root divergence was more accentuated than resorption), the well-delimited borders, the encapsulation and the lack of invasion signs, significant atypia, necrosis and recurrence after 4 years suggest benignity from both clinical and histopathological standpoints.

CONCLUSION

Schwannoma should not be discarded when observing a tumor in the oral cavity. It can have far reaching immense complications if it involves a nerve or malignant transformation may take place on leaving untreated. Definitive preoperative diagnosis should be carried out with a biopsy and anatomopathologic study. Since a rare case of intraosseous schwannoma of the mandible is reported here with radiographical features suggestive of markedly enhanced solid mass with no cystic parts, hence, a culmination of clinical, radiographic and histopathologic examination was imperative in cases so as to initiate the right approach and correct treatment plan for the patient.

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