

## CASE REPORT

### A RARE CASE OF INTRA OSSEOUS SCHWANNOMA IN RETROMOLAR AREA AND REVIEW OF LITERATURE

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#### ABSTRACT:

Schwannomas are one of the benign neoplasms originating from the neural sheath. Soft tissues of the head and neck are the most common sites of occurrence; however, intraosseous schwannomas are rare. Intraoral lesions are uncommon, however, and intraosseous schwannomas are even rarer. In the Mayo Clinic series of 11,087 primary bone tumors, 14 cases of intraosseous schwannoma were identified, accounting for less than 1% of these benign primary bone tumors. The most common site of occurrence is the mandible, a characteristic traditionally attributed to the long intraosseous path of the inferior alveolar nerve. In this article, we describe an additional case occurring in the mandible of a 18-year-old male patient.

Keywords: Intraosseous schwannoma, mandible, neurilemmoma.

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#### INTRODUCTION:

Schwannomas (neurilemmomas) are slow growing, benign neoplasms derived from Schwann cells, the sheath cells that cover myelinated nerve fibres.<sup>1-3</sup> The first reported case in literature of a schwannoma or neurilemmoma was by Verocay in 1910.<sup>4</sup> This tumor often exists in association with pre-existing neurofibromatosis or von Recklinghausen disease.<sup>5,6</sup> It has been suggested that patients in whom the lesion is associated with neurofibromatosis have a poorer prognosis, attributed to a tendency toward larger tumors, poorer differentiation, higher rate of metastases, and multifocality. These tumors most common arise in the soft tissues of the head and neck, as well as on the flexor surfaces of the upper and lower extremities.<sup>7,8</sup> Intraoral lesions are uncommon, however, and intraosseous schwannomas, are even rarer.<sup>1</sup> The most common site of occurrence is the mandible characteristic traditionally attributed to the long intraosseous path of the inferior alveolar nerve.<sup>9</sup> In the current medical literature, there are 44 acceptable cases of intraosseous schwannoma of the jaws, 39 of the mandible and five in the maxilla, representing less than 1% of the primary tumors of the bones. Other sites reported include the sacrum, vertebra, clavicle, ribs, humerus, radius, ulna etc.

Documented cases : 83 only

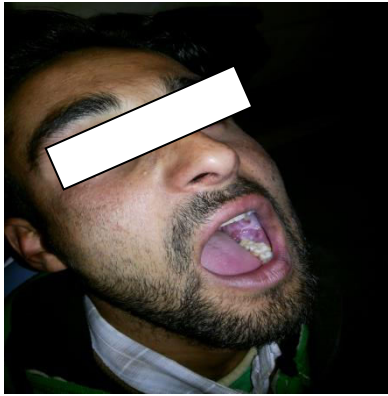
Sex predilection : Slightly female

Jaw predilection : Mandible (84.3%)

Often affects : Posterior segments

Mean patient age : 30.8 years

**CASE REPORT:** A 18 year old male patient presented with a diffused swelling in lower left third molar area measuring 2.5x2 cms with surface ulcerations caused by the opposing third molar. (Figure 1, 2) The patient reported no history of pain and paresthesia. Panoramic radiography showed a multilocular radiolucent area with distinct borders extending mesially upto the apical area of the distal root of the first molar and distally involving the body of ramus. Third molar was displaced distally and inferior alveolar nerve displaced inferiorly. (Figure 3,4) The differential diagnosis included odontogenic cysts (especially odontogenic keratocyst), odontogenic tumors (such as unicystic ameloblastoma and ameloblastic fibroma), non odontogenic central tumors (such as central giant cell granuloma), and traumatic bone cyst. Because neurilemmoma of the jaw is a rare entity, it was not considered in our differential diagnosis list. An incisional biopsy was obtained under local anesthesia. Histopathologic examination of the H&E stained section of the incisional biopsy showed areas of Antoni A tissue including bundles of spindle shaped cells with long basophilic nuclei palisaded around an acellular eosinophilic areas known as Verocay bodies. Antoni B tissue with randomly arranged spindle cells, within a loose myxomatous stroma was also seen therefore a preliminary diagnosis of neurilemmoma was made. Subsequently the patient underwent total removal of the tumor under local anesthesia.



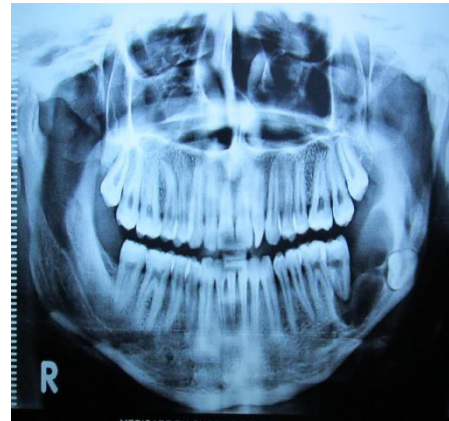
**Figure 1:** Clinical Photograph



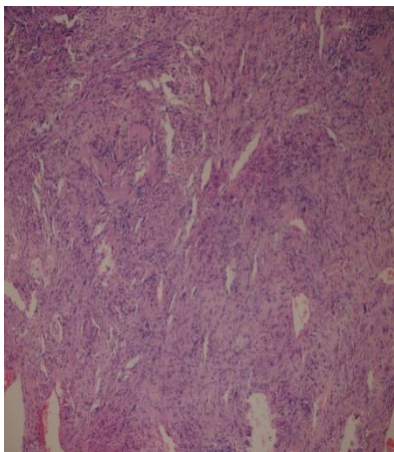
**Figure 2:** Diffuse swelling in left mandibular third molar region



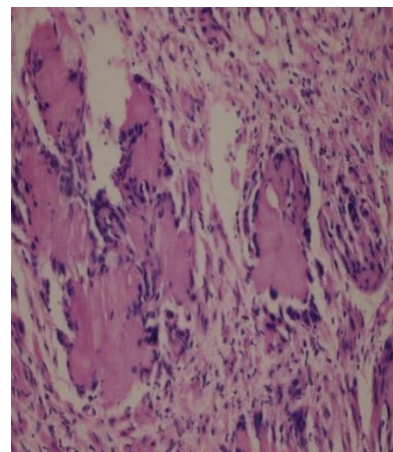
**Figure 3:** OPG showing multilocular radiolucent area



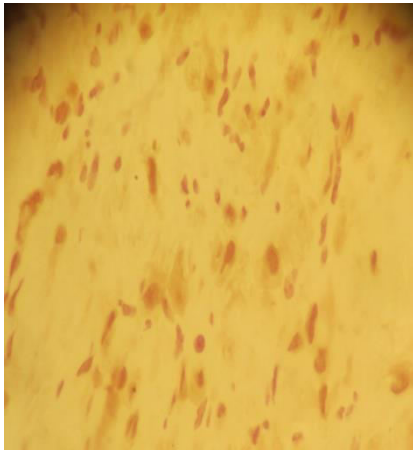
**Figure 4:** Multilocular radiolucent area in left mandibular area



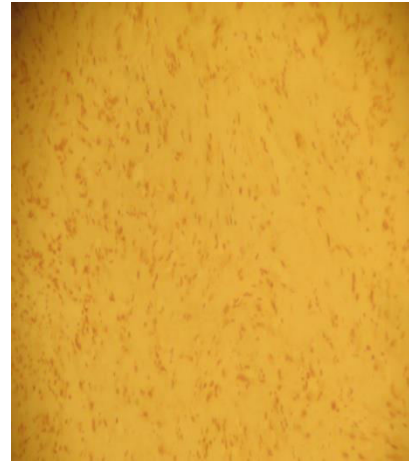
**Figure 5:** Photomicrograph, H and E stained section Antoni A and Antoni B pattern



**Figure 6:** Photomicrograph, H and E stained section showing verrocay bodies



**Figure 7:** S-100 immunostaining



**Figure 8:** S-100 protein was diffusely positive

**INVESTIGATIONS:** OPG showed a multilocular radiolucent area with distinct borders extending mesially upto the apical area of the distal root of the first molar and distally involving the body of ramus. Third molar was displaced distally and inferior alveolar nerve displaced inferiorly.

Histological examination showed hypercellular “Antoni-A” pattern of spindle cells with elongated palisading nuclei surrounding pink acellular zones (Verocay bodies). “Antoni –B” pattern with a looser stroma and fewer cells were also seen. (Figure 5, 6)

An Immunohistochemistry for S-100 protein was diffusely positive. (Figure 7,8)

**DISCUSSION:** The neurilemmoma is a benign neoplasm of Schwann cell. These tumors commonly arise in the head and neck soft tissue, but intraoral lesions are unusual, especially centrally in the jaws.<sup>10-14</sup> There are three mechanisms by which schwannomas may involve a bone

1. A tumor may arise centrally within a bone,
2. A tumor may arise within a nutrient canal, or
3. A soft tissue or periosteal tumor may cause secondary erosion and penetration into bone.

This case demonstrates an example of schwannoma centrally within a bone. In the review of the literature, Chi et al.<sup>13</sup> reported 44 intraosseous schwannomas of the jaws including 39 mandibular and 5 maxillary tumors. Most of mandibular tumors were placed in the posterior areas of the body and ramus. It can be attributed to the long intraosseous pathway of the inferior alveolar nerve within the mandible.<sup>14-16</sup> The average age was 34 years, with peak prevalence in the second and third decades of life,<sup>13</sup> however our case was an 18-year-old which was younger than the mean age. Intraosseous schwannoma is a slow-growing tumor that frequently produces expansion of the affected bone and causes swelling. Pain and paresthesia may be found in 50% of the patients.<sup>16,18</sup> Our patient had a large swelling with no pain or paraesthesia. He reported 1 month’s duration, but based on the tumors large size and histopathologic degenerative changes, true

duration of the lesion seems to be more. Radiographically, Intraosseous schwannoma is a well-circumscribed unilocular radiolucency, with thin sclerotic borders commonly found in the posterior areas of mandible that can be like the other benign cystic lesion such as OKC or a benign tumor, like ameloblastoma.<sup>14,19</sup>

Microscopically, the features were found to be characteristic of neurilemmomas including encapsulation, Verocay bodies, palisading nuclei, and Antoni A and B tissues. These tumors’ cells typically showed a diffuse positive immunoreactivity for S-100 protein.<sup>18,20</sup> In addition to the classic schwannoma, there are several histopathologic variants, including the cellular, plexiform, epithelioid, ancient, and melanotic types.<sup>20</sup> Ackerman and Taylor<sup>21</sup> first found the schwannoma presented with clear areas of hypocellular tissues and attributed the changes to the long standing degenerative changes. Ancient schwannomas tend to be large tumors of long duration and are characterized by degenerative alterations including cystic changes, calcification, hemorrhage, and hyalinization. Macrophages and hemosiderin deposits are common. Although nuclear atypia may be striking, mitotic activity is absent.<sup>13,17</sup>

Reviewing the literature, only eleven cases of ancient schwannoma had been reported that were placed in the anterior portion of tongue, palate, mandibular and maxillary vestibule region, floor of the mouth and buccal mucosa.<sup>17,20</sup> The most common site of this tumor among the cases seen in the literature was floor of the mouth, with mean age of 30.5 years and female sex predilection.<sup>17</sup>

Malignant transformation of the schwannoma is almost unknown although one acceptable example has been reported. (Carstens and Schrodt, 1969). However, recurrence of the schwannoma is possible if it is inadequately excised.<sup>22</sup> Schwannomas are resistant to radiotherapy.<sup>23</sup> Because it is a well-encapsulated lesion, the treatment of choice is conservative surgical enucleation with periodic follow-up.

**CONCLUSION:** Schwannoma treatment consists of surgical enucleation with periodic follow-up examinations. Recurrence is uncommon and in this case,

the patient was followed up for three months with no clinical or radiographic signs of recurrence.

#### REFERENCES:

- Murphy J, Giunta JL. Atypical central neurilemmoma of the mandible. *Oral Surg Oral Pathol* 1984; 59:275-8
- Villanueva J, Gigoux C, Sole F. Central neurilemmoma of the maxilla. A case report. *Oral Surg Oral Pathol Oral Radiol Endod* 1995;79:41-3.
- Belli E, Becelli R, Matteini C, Iannetti G. Schwannoma of mandible. *J Craniofac Surg* 1997;8:413-6.
- Verocay J. Zur Kenntnis der Neurofibrome. *Beitr Path Anat* 1910;48:1-68.
- Elias MM, Balm AJ, Peterse JL, et al: Malignant schwannoma of the parapharyngeal space in von Recklinghausen's disease: A case report and review of literature. *J Laryngol Otol* 107:848,1993.
- Uehara T, Matsubara O, Kasuga T: Malignant schwannoma arising in Recklinghausen's disease: Report of 4 cases. *Gan-no-Rinsho* 31:308, 1985
- Ackerman LV, Spjut HJ. Tumors of bone and cartilage. Washington, DC: Armed Forces Institute of Pathology; 1962. P.247-9.
- Enzinger FM, Weiss SW. Benign tumors of peripheral nerves. In: *Soft tissue tumors*. 4<sup>th</sup> ed. St Louis: Mosby -year Book, Inc; 2001. p.1111-1200.
- Celli P, Cervoni L, Colonnese C. Intraosseous schwannoma of the vault of the skull. *Neurosurg Rev* 1998;21:158-160.
- Neville B, Damm DD, Allen CM, Bouquot J. 3rd ed. Philadelphia: Saunders; 2008. *Oral and Maxillofacial Pathology*; pp. 526-7.
- Regezi JA, Sciubba JJ, Jordan RCK. 5th ed. Philadelphia: Saunders; 2007. *Oral Pathology: Clinical Pathologic Correlations*; p. 170.
- Gnepp DR. 2nd ed. Philadelphia: Saunders; 2009. *Diagnostic Surgical Pathology of the Head and Neck: Expert Consult - Online and Print*; pp. 245-6.
- Chi AC, Carey J, Muller S. Intraosseous schwannoma of the mandible: a case report and review of the literature. *Oral Surg Oral Med Oral Pathol Oral Radiol Endod*. 2003;96(1):54-65.
- Hsieh MY, Chen CM, Lin LM, Chen YK. Intraosseous Neurilemmoma (Schwannoma) of the Mandible - Report of a Case. *Cases Journal Taomfr*. 2009;1(2):1-6.
- Gallego L, Junquera L, Rodriguez-Recio C, Fresno MF. Intraosseous mandibular schwannoma mimicking an odontogenic keratocyst, with a postsurgical pathological fracture. *J Laryngol Otol*. 2009;123(5):560-2.
- Nakasato T, Katoh K, Ehara S, Tamakawa Y, Hoshino M, Izumizawa M, et al. Intraosseous neurilemmoma of the mandible. *AJNR Am J Neuroradiol*. 2000;21(10):1945-7.
- Amirchaghmaghi A, Salehinejad J, Basirat M, Delavarian Z, Javadzade A, Forouzanfar A. Gingival Ancient Schwannoma: Review of Literature and a Case Report. *Journal of Applied Sciences*. 2010;10(23):3137-40.
- Ito S, Mandai T, Ishida K, Deguchi H, Hata T, Irei I, et al. Intraosseous schwannoma of the mandible: A case report. *Kawasaki medical journal*. 2009;35(3):249-52.
- Jang KY, Moon WS, Park HS. Intraosseous Neurilemmoma of the Mandible - A Case Report. *The Korean Journal of Pathology*. 2009;43(1):88-91.
- Subhashraj K, Balanand S, Pajaniammalle S. Ancient schwannoma arising from mental nerve. A case report and review. *Med Oral Patol Oral Cir Bucal*. 2009;14(1):E12-4.
- Ackermann LV, Taylor FH. Neurogenous tumors within the thorax. A clinicopathological evaluation of 48 cases. *Cancer*. 1951;4:869.
- Williams HK, Cannell H, Silvester D, Williams DM. Neurilemmoma of the head and neck. *Br J Oral Maxillofac Surg*. 1993;31:32-5.
- Yusuf H, Fajemisin OA, McWilliam LJ. Neurilemmoma involving the maxillary sinus: A case report. *Br J Oral Maxillofac Surg*. 1989;27:506-11.
- Parhar S, Singh HP, Nayyar, Manchanda A. A Intra-Oral Schwannoma- A Case Report. *J Clin Diagn Res*. 2014 March; 8(3): 264-265.

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