Case Report

Diagnostic imaging of a Rare Intraosseous Mandibular Schwannoma using Cone Beam Computed Tomography – A case report

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ABSTRACT

Intraosseous schwannoma is a very rare (<1%), slow-growing, benign tumor of Schwann cells. The mandible is the most common site for intraosseous schwannoma. Here, we report the case of a 35-year-old female reported with extraoral and intraoral swelling with bicortical expansion on the left side of the mandible for the past 6 months. Two-dimensional imaging modalities (intraoral periapical radiograph, orthopantomogram, and occlusal radiograph) showed giant well-defined false multilocular radiolucency appearance in the left body of the mandible and external root resorptions of the premolars and molars. Aspiration was non-productive. Cone-beam computed tomography (CBCT) study showed a bicortical expansile unilocular hypodense lesion in continuity with the widened mandibular canal suggesting a tumor of neural tissue origin. Radiological diagnosis of intraosseous schwannoma was given and histopathological examination after an excisional biopsy confirmed the same. The postsurgical phase was uneventful and the patient is under follow-up. This article highlights the importance of three-dimensional modality like CBCT in narrowing the diagnosis of such rare clinically misleading presentations

Key words: Cone-beam computed tomography, Neurilemmoma, Schwannoma

Schwannoma (neurilemmoma, neurinoma, or perineural fibroblastoma) is a benign tumor of Schwann cells, which forms the inner layer of nerve sheath (World Health Organization grade-I, International Classification of Diseases-O 9560/0). It is classified as Central schwannoma (intraosseous) and Peripheral schwannoma (in soft tissues). It is relatively common in the head and neck region (25%) [1,2]. Intraorally, it is rare (1–12%) with the tongue as the most predilected site. Intraosseous schwannomas are even rarer, <1% of primary bone tumors [2-5].

Here, we report a case with detailed cone-beam computed tomographic (CBCT) image findings of the intraosseous mandibular schwannoma of inferior alveolar nerve (IAN) and becomes first of its kind in the literature [2,6] to the best of our knowledge

CASE REPORT

An otherwise healthy 35-year-old female patient reported to our Department of Oral Medicine and Radiology with complaints of pain and swelling on the left side of the lower jaw for the past 6 months. There was a history of swelling which was gradual in onset and slow in progress, also associated with dull and continuous pain in the left body of the mandible region for the past 6 months, and paresthesia of the lower lip for the past 1 month. Past medical and surgical histories were non-contributory.

The patient was conscious, alert, and oriented. No signs of pallor, icterus, cyanosis, clubbing, edema, and lymphadenopathy were present. The pulse rate was 72/min, respiratory rate was 16/min, blood pressure was 120/80 mmHg, and the temperature was 98.6°F. On extraoral examination, an ill-defined, diffuse swelling, approximately 5×2 cm on the left side of the mandible extending from the angle to the parasymphysis medially was evident (Fig. 1a). On palpation, the swelling was non-warmth, non-tender, and hard in consistency. On neurosensory examination, two-point discrimination, and light touching tests showed decreased sensation on the lower lip and the labial mucosa. Intraorally, an ill-defined, hard, non-tender, and non-pulsatile tumefaction with bicortical expansion obliterating the left mandibular buccal vestibule in relation to 33–38 was evident (Fig. 1b). Electric pulp vitality testing showed a negative response in those teeth. Teeth 35, 36, and 37 were inclined lingually with grade-I mobility.

Intraoral periapical radiograph (IOPA), lateral mandibular occlusal, and the panoramic radiograph showed a

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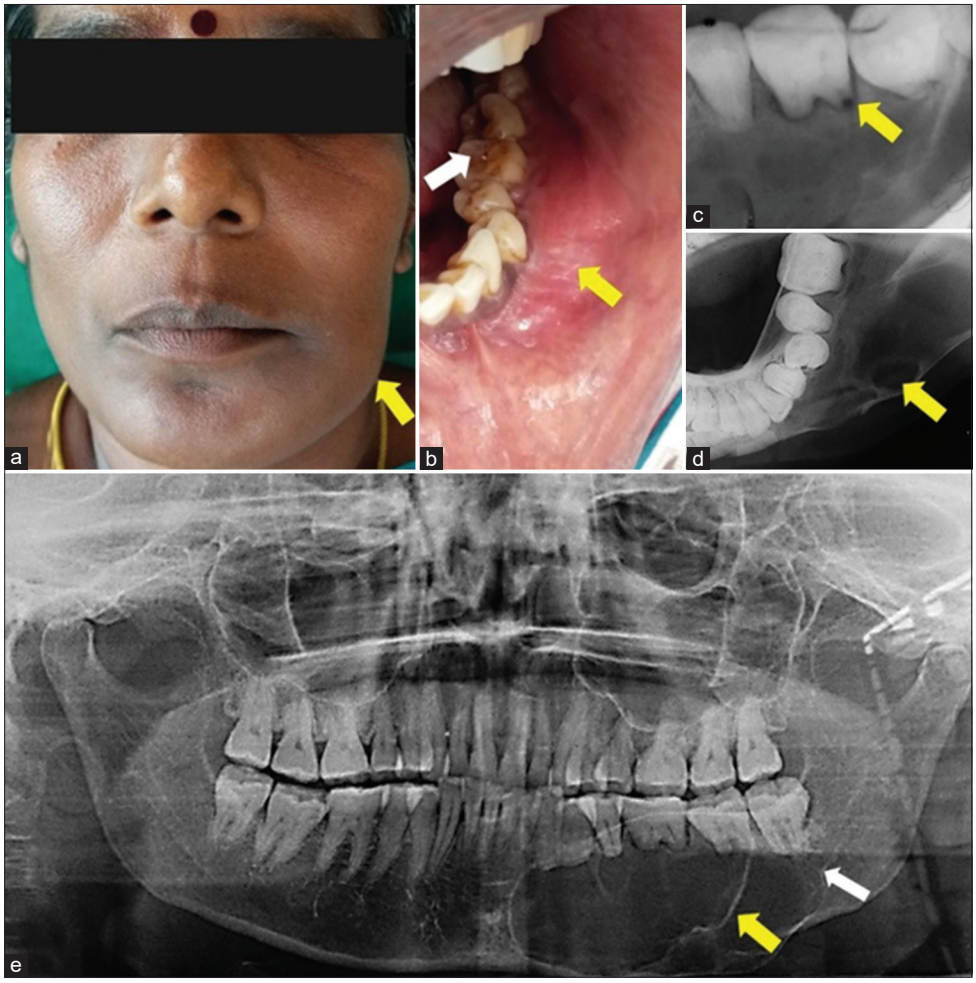

Figure 1: (a) Extraoral swelling in the left side of the mandible (yellow arrow); (b) intraoral swelling obliterating the left buccal vestibule (yellow arrow) and lingual inclination of teeth (white arrow); (c) periodontal ligament space widening, external root resorption in IOPAR; (d) Mandibular occlusal radiograph showing bicortical expansion, multilocular radiolucent appearance (yellow arrow); (e) orthopantomogram showing multiple radiopaque internal septa (yellow arrow), multilocular appearance and inferior alveolar nerve canal not well appreciated (white arrow)

well-circumscribed expansile multilocular radiolucent lesion separated by thin septa as internal architecture and with sclerotic border. External root resorption of 35, 36, and 37 evident with the widening of periodontal ligament spaces (Fig. 1c-e). Aspiration was non-productive. A provisional diagnosis of multicystic ameloblastoma was given.

A CBCT scan was done using CS 9300 select CBCT machine (Carestream Health, Rochester, NY) with isotropic voxel size (slice thickness) of 180 m, 10×10 cm field of view with the mandible as a region of interest at 90 kVp, 4 mA, 8 s. Multiplanar reconstruction and panoramic reformatting along with tracing of the IAN canal (Fig. 2) were done in the region of interest. The axial section showed a massive bicortical expansile hypodense lesion measuring 5.58 cm anteroposteriorly and 3.33 cm mediolaterally in its greatest dimension, with cortical thinning and perforation. A thin hyperdense septum was evident without separating the internal structure into at least 2 hypodense compartments suggesting its unilocularity in contrast to IOPA, panoramic, and occlusal findings. Widening of mental foramen was evident (Fig. 3a and b).

Corrected sagittal section through the course of the inferior alveolar canal showed a widening of the IAN canal into a fusiform

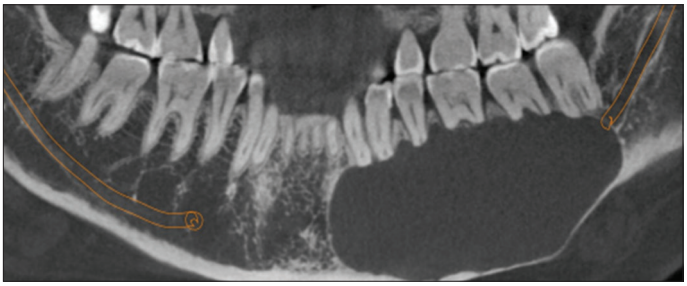

Figure 2: Cone-beam computed tomography panoramic reformatted image (slice thickness 300 μm, minimum intensity projection) with inferior alveolar nerve canal tracing

shaped hypodense lesion measuring 2.42 cm superioinferiorly, with characteristic scalloped cortical margins and thinning of the inferior border of the mandible. The cortical border of the IAN canal was continuous with that of the lesion suggesting the vascular or neurogenic origin of the lesion (Fig. 3c). Radiological differential diagnoses were central schwannoma and neurofibroma of IAN.

Under general anesthesia, marginal mandibulectomy with surgical enucleation of the tumor (Fig. 4) was done. Histopathological examination (hematoxylin and eosin-stained)

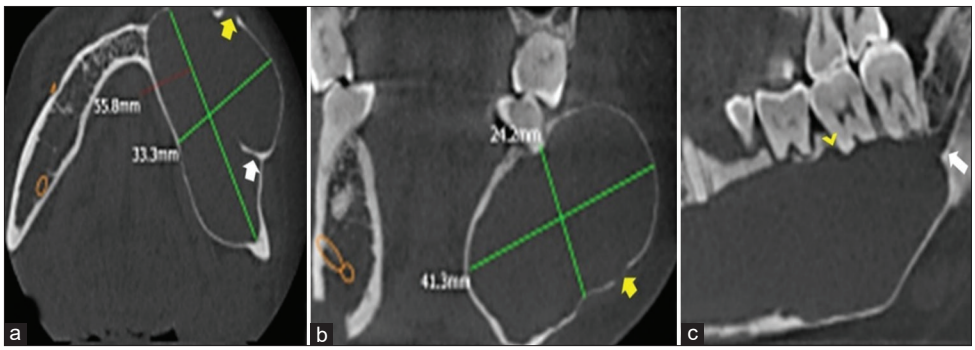

Figure 3: (a and b) Cone-beam computed tomography axial and coronal section showing an expansile unilocular hypodense lesion with cortical thinning and an incomplete hyperdense septum (white arrow) projecting into the hypodense area giving a scalloped appearance. Widening of mental foramen (yellow arrow) is remarkable; (c) corrected sagittal section parallel to the inferior alveolar nerve canal shows the continuity (white arrow) of its cortical border with the hypodense lesion. Scalloped cortical outline (yellow arrow)

revealed streaming fascicles of spindle-shaped Schwann cells in palisaded arrangement around the central acellular eosinophilic area called Verocay bodies, suggesting Antoni type A arrangement (Fig. 5a). Spindle cells arranged randomly within loose myxomatous stroma in few areas resembling Antoni type B showing classic schwannoma type (Fig. 5b). Hence, the above histopathological findings proved our radiological diagnosis of Central Schwannoma of IAN. The post-operative phase was uneventful. A post-operative orthopantomogram after 4 months shows new bone formation (Fig. 6).

DISCUSSION

WHO defined Intraosseous schwannomas as "a benign tumor of nerve sheath origin having the same morphological features as soft tissue neurilemmomas/schwannomas, but originating within the bone" [4]. A schwannoma is a benign tumor of neuroectodermal origin, arising from the Schwann's cells of the spinal nerves, cranial nerves (except optic and ophthalmic nerves), and autonomic nerves [3,7,8]. Perkins *et al*. updated a review in 2018 which documented 88 cases of intraosseous schwannoma of jaws, of which mandibular schwannomas accounted 79 cases, including their two cases [2]. Because of the longest course of IAN along the length of the mandible than any other bone, the mandible may be the most affected one, followed by the spine and the sacrum [3,4,9].

Intraosseous schwannoma is more common in the second and third decades (mean 36.9 years) and females show high predilection than males (M: F=1:1.5) [2,7]. The etiology of this encapsulated tumor is unknown [1], but it typically arises in association with a nerve trunk and pushes aside the nerve along with adjacent structures as they grow slowly and causes asymptomatic swelling, drifting of teeth (14%), and pressure associated root resorption (26%) of the teeth [2]. The site of predilection is the posterior body and the mandibular ramus (78%) [2,3]. Our case also presented with lip paresthesia and tooth mobility which are all less commonly reported findings (<20%) [2,9].

It usually presents as a well-circumscribed unilocular radiolucency (76%) with sclerotic margins. Multilocular

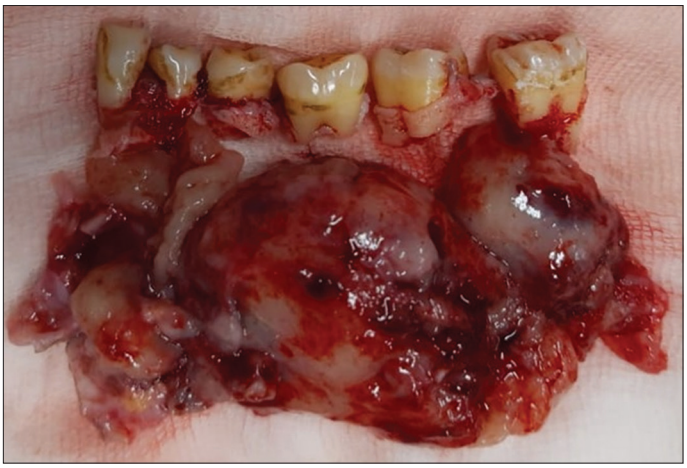

Figure 4: Gross specimen showing grayish-white rounded, smooth, convex growth of the benign tumor

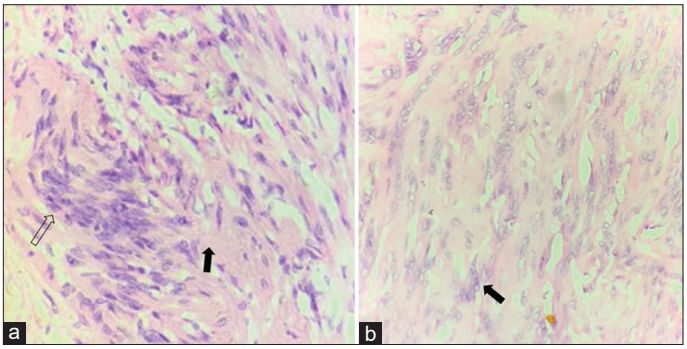

Figure 5: Photomicrograph (100× magnification, hematoxylin and eosin-stained) showing (a) palisaded arrangement of Schwann cells (hollow black arrow), acellular eosinophilic area (solid black arrow); (b) spindle cells arranged randomly in loose myxomatous area

presentation is very rare (16%). The bicortical expansion, multilocularity in two-dimensional imaging modality, external root resorption, peripheral scalloping, negative aspiration mimicked odontogenic tumors like ameloblastoma, and odontogenic keratocyst similar to other literature [2,4,5]. However, in the CBCT, it was clearly unilocular as the internal structure was not separated into compartments by the bony septae [4,7]. Such incomplete septa have been reported previously in computed tomography (CT) study of the same

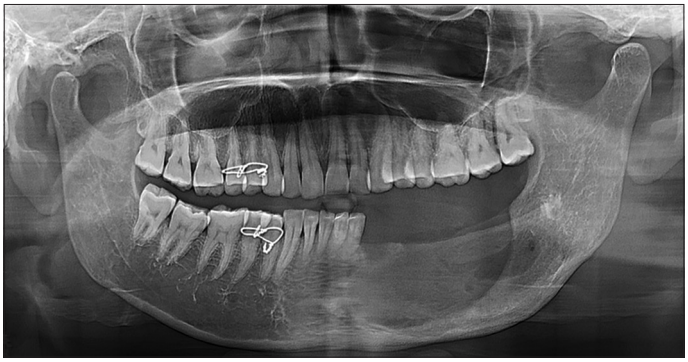

Figure 6: Post-operative orthopantomogram after 4 months follow-up showing new bone formation

lesion which gave an impression of multilocularity in plain radiographs [10,11].

Some authors say multilocularity in the two-dimensional imaging modality might be due to the peripheral scalloping, that is, due to the shadowing of corrugations on the walls of the bone cavity containing the tumor [5,7,12]. Furthermore, distinct asymmetrical distension of the mandibular canal is considered to be a profound indicator of neural pathology [13] and a well-defined sclerotic border suggested benign nature [5]. There was a fusiform widening of the mandibular canal in our case which is rare (10%) [13]. Other main causes of the widening of the mandibular canal are vascular malformations, but it is ruled out in our case due to negative aspiration.

In addition, in the case of these tumors, even if it is expanded, thinned, or perforated, the cortical outline can still be seen in multiplanar reconstructions of CT/CBCT. As malignant tumors of IAN canal contents usually show ill-defined cortical borders with ragged edges, we ruled out the possibility of malignancy [2]. Periosteal reactions are usually rare [4]. Schwannomas usually become rounded when they arise and the wall of the IAN canal is preserved, but in contrast, neurofibromas tend to grow particularly in the canal, which typically becomes ovoid-shaped [14]. The differentiation between these two tumors is important as the latter has a high recurrence rate and potential for malignant transformation.

Establishing a diagnosis based on routine radiographs is of little value because they do not possess any features that distinguish schwannoma from other lesions [10]. However, CBCT, with low radiation dose, can enable better diagnosis of schwannomas of the head and neck region, as in our case, by scrutinizing the continuity and enlargement of the mandibular canal, mental foramen, internal structures details, etc. The need for a magnetic resonance imaging in the present case was ruled out due to the greater radiographic detail obtained using CBCT imaging and the absence of any evidence of extragnathial extension.

Histologically, intraosseous, and soft tissue schwannomas are identical. They are encapsulated tumors composed of both Antoni A and B arrangements with Verocay bodies. It shows strong diffuse cytoplasmic and nuclear immunoreactivity for S-100 protein. This is a classic schwannoma type similar to our case. There are five other types of schwannomas: (1) Ancient schwannoma with degenerative changes, (2) plexiform, (3) epithelioid, (4) melanotic/pigmented, and (5) microcystic/reticular types [2].

DeLeonibus *et al*. recommended the surgical approach for intraosseous schwannomas of mandible based on the location and extent of the tumor and suggested segmental mandibulectomy for large lesions more than 3.5 cm [15]. In our report, the lesion was more than 5 cm in the anteroposterior aspect. In our case, the surgical treatment plan of marginal mandibulectomy from 33 to 38 with surgical enucleation was done to preserve the mandible and for better prognosis. Malignant transformation and recurrence are rare. Prognosis is usually good due to its benign and encapsulated nature [7,9].

CONCLUSION

A rare case of intraosseous schwannoma of the mandible has been reported with CBCT findings. Prompt diagnosis and appropriate treatment should be based on thorough radiological, clinical, and histopathological investigations.

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