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CASE REPORT

Unusual intramandibular plexiform shwannoma

R.M. Bavle, L.J. Thambiah, H. Nanda Kumar, N. Srinath, K. Paremala & M. Sudhakara

Krishnadevaraya College of Dental Sciences and Hospital, Department of Oral Pathology, Hunasamaranahalli, (via) Yelahanka Bangalore, Bangalore Karnataka, India

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Correspondence to:
Dr R Bavle
Krishnadevaraya College of Dental Sciences and Hospital, Department of Oral Pathology
Hunasamaranahalli (via) Yelahanka Bangalore
Bangalore Karnataka
India 562157
Tel.: +919844244822
Fax: +918028467084
email: radix69@gmail.com

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Abstract

Background: Schwannoma is a benign neoplasm originating from the neural sheath of peripheral soft tissues, but to occur within the jaw bones is exceptional. Plexiform schwannoma is a rare variant of Schwann cell tumour having plexiform pattern of intraneural growth with multinodularity.
Case report: This report documents a case of a plexiform intraosseous schwannoma located in the mandible of a fifty-four-year old male.
Conclusion: Although intraosseous schwannomas of the jaw bones and plexiform schwannomas of soft tissues of head and neck are documented in the recent literature; to the best of our knowledge, an intraosseous variant of plexiform schwannoma has not been reported.

Introduction

Neoplasms of peripheral nerve in the head and neck region are of common occurrence, but origin in the oral and para-oral tissues is uncommon and they rarely occur centrally within the jaws[1]. Schwannoma is a benign, encapsulated nerve sheath tumour composed of primarily Schwann cells in a poorly collagenised stroma. It has a vast deal of literature on its nomenclature and cell of origin. There is enough evidence to suggest that schwannomas arise from the Schwann cells and hence, are appropriately named as schwannoma[2].

Schwannoma generally occurs in an age group ranging from 10 to 40 years. They most commonly occur in the head and neck area and flexor areas of the extremities. They present as a discrete, freely movable, smooth-surfaced, usually painless soft tissue swelling[2]. Intraosseous schwannomas are rare and comprise 1% of all bone tumours in the entire body. A review of literature confirmed the mandible to be the most common site for the occurrence of intraosseous schwannomas, with only 43 acceptable gnathic cases previously reported[3]. It presents as an asymptomatic, well-defined unilocular radiolucency with only three cases being reported as multilocular radiolucency in the jaw bones[3].

Histologically, classical schwannoma is an encapsulated neoplasm having two components – Antoni type A and Antoni type B tissue in variable proportions. Antoni A tissue is cellular and consists of monomorphic spindle-shaped Schwann cells with poorly defined eosinophilic cytoplasm and pointed basophilic nucleus. These cells commonly show nuclear palisading, and parallel arrays of such palisades with intervening eosinophilic cell cytoplasm are known as Verocay bodies. Antoni B areas are also composed of Schwann cells, but the cytoplasm is inconspicuous and nuclei are suspended in copious myxoid and often microcystic matrix[1–3].

The common variants of schwannoma are cellular, epitheloid, melanotic and ancient. The recent and newer variant of schwannoma being the plexiform schwannoma[3].

Plexiform schwannoma is an extremely rare and unique variant of schwannoma having predilection for superficial soft tissues in the head and neck region[3]. It occurs as a conventional[4–6] or cellular variant[7],

characterised clinically and/or histologically by a plexiform or multinodular growth pattern (multiple discohesive nodules). There are no reports of this variant occurring in an intraosseous location. Sometimes, such multinodular growth arises in patients showing neurofibromatosis-2 (NF-2) and schwannomatosis[8,9].

Here, we present a unique case of plexiform schwannoma arising intraosseously in the mandible. Our search in English literature did not reveal any conspicuous report of a similar case, so this may be the first case to be documented.

Case report

A 54-year-old male patient reported to the Krishnadevaraya College of Dental Sciences and Hospital, Bangalore, India, with a chief complaint of swelling in the left cheek region for the past 6 months. His medical history was non-significant. Extraoral examination revealed slight asymmetry on the left side of the face in the preauricular and mandibular area. The swelling extended inferio-superiorly from the lower border of the mandible to the ala-tragal line and antero-posteriorly from 2.0 cm posterior to the angle of the mouth to the angle of the mandible. The swelling measured 3.0 × 3.0 cm in dimension, was well-defined, hard, non-tender, with normal overlying skin (Fig. 1). Marked paraesthesia was present on the left half of the lower lip and angle of the mouth. Solitary submandibular lymph node was palpable, mobile and tender. Intraoral examination revealed a polypoid mass, measuring about 1.0 × 2.0 cm in relation to the left retromolar area. The mass was well defined, soft and non-tender, obliterating the buccal sulcus. Overlying mucosa showed a large central area of yellowish gray mucinous covering (Fig. 2).

The orthopantamograph showed multilocular radiolucency with faint trabeculae, suggesting loculation, with smooth and distinct borders extending from the region of 36 to the left ramus of the mandible. The lower border of the mandible was intact and root

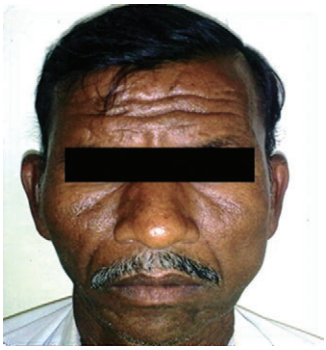

Figure 1 Clinical photograph showing extraoral swelling of the left side of the face in the preauricular and mandibular area.

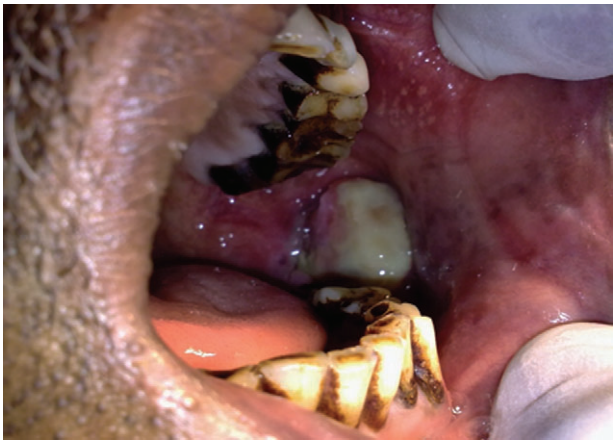

Figure 2 Intraoral clinical photograph showing well defined mass obliterting the left buccal sulcus having a large central area of yellowish gray mucinous covering.

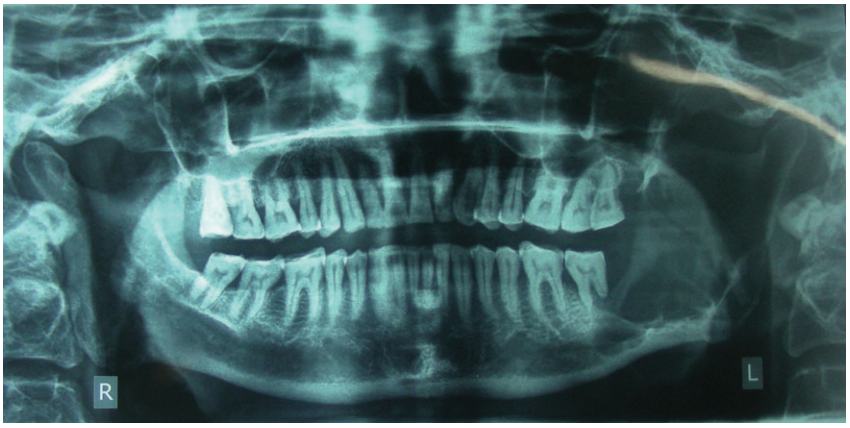

Figure 3 Orthopantogram showing faint multilocular radiolucency with distinct borders extending from the region of 36 to left ramus of the mandible. The lower border of the mandible was intact. Root resorption of distal root of 37 is seen.

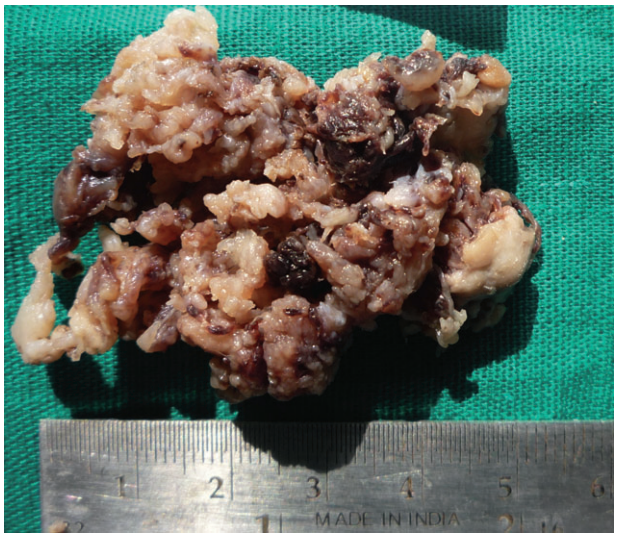

Figure 4 Macroscopic photograph showing multiple nodules being held by tenacious fibrous intervening tissue.

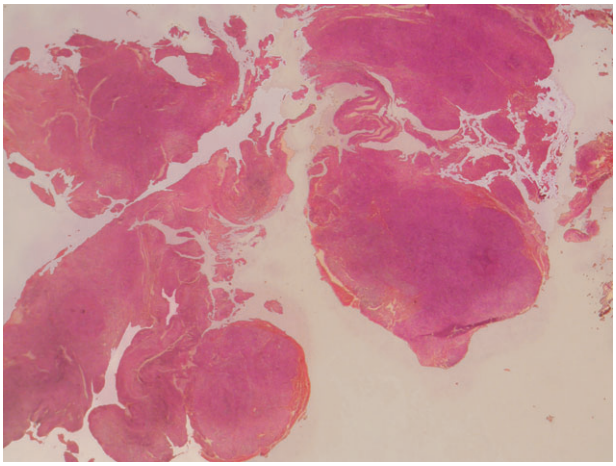

Figure 5 Photomicrograph showing clearly segregated multiple nodules and surrounded by fibrous connective tissue. ×10.

resorption of 37 was seen (Fig. 3). The occlusal radiograph showed marked expansion of the cortices, more so, on the lingual aspect.

Surgical note

The lesion was enucleated and curetted under general anaesthesia. The lesion separated out from the bone easily. Lesion was removed by transecting the attachment to the nerve, leaving the nerve trunk intact. Extremely thinned out buccal cortical plate in the region of 37 and 38 was removed along with the lesion. The uninvolved condyle and coronoid processes were left intact. The 37 was extracted and it showed a moderate amount of resorption of the distal root and apical resorption of the mesial root.

Pathology

Macroscopic findings

The lesional tissue measured 5.5 × 4.5 cm in dimension, soft in consistency and had a matted appearance with multiple nodules being held by tenacious fibrous intervening tissue (Fig. 4). Sections of multiple areas were taken for histopathological diagnosis and the tissues were sent for routine processing. Resorption on the roots of 37 was confirmed on gross examination.

Microscopic features

The haematoxylin- and eosin-stained sections showed clearly segregated multiple nodules in lower magnification (×40) (Fig. 5). On higher magnification, these nodules (×100 and ×400) showed highly cellular stroma at many areas. The cells were spindle in shape with eosinophilic cytoplasm and basophilic oval nucleus exhibiting palisading. Parallel arrays of such palisades with intervening eosinophilic cell cytoplasm – Verocay bodies – were evident, depicting Antoni type A tissue. These densely cellular areas were alternating with less cellular, loose, oedematous areas, containing numerous small blood vessels exhibiting perivascular hyalinisation and a mild amount of collagenisation representing Antoni type B areas (Fig. 6). Ill-defined nerve fascicles were seen in a loose, less cellular, collagenous tissue and a large, mature nerve fascicle/nerve trunk at the periphery of the lesion was seen.

S-100 immunostaining showed strong positivity in the nerve fascicles, Antoni type A and Antoni type B areas of the section, while the capsular and the intervening areas showed negative staining (Fig. 7). A very mild positivity was seen for Ki-67 in the lesional tissue.

The patient has been kept under a post-surgical follow-up since 14 months. He is showing good health and absence of any clinically detectable local alterations.

Discussion

Schwannoma is a peripheral nerve sheath tumour commonly defined as a benign, encapsulated neoplasm arising from a nerve sheath composed primarily of Schwann cells in a poorly collagenised stroma, and is thought to arise from Schwann cells[1–3].

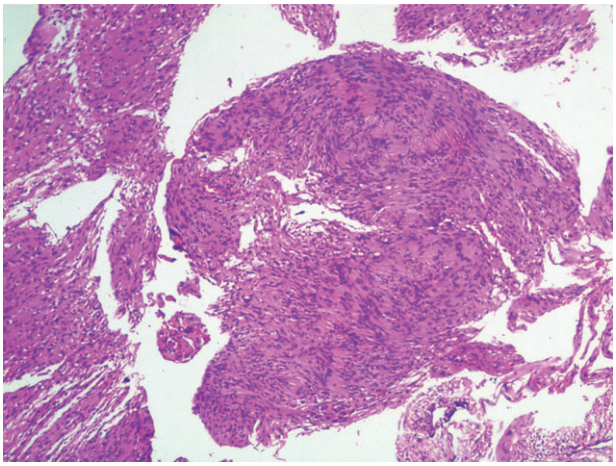

Figure 6 Photomicrograph showing densely cellular areas with verocay bodies depicting Antoni type A tissue alternating with less cellular, loose, oedematous areas containing numerous small blood vessels and mild amount of collagenisation representing Antoni type B areas. H & E stain. ×100.

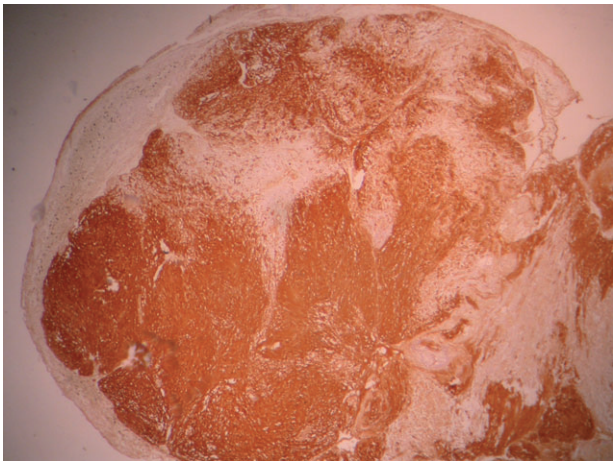

Figure 7 Photomicrograph showing S-100 immunostaining with strong positivity in the Antoni type A and Antoni type B areas of the section and negative in the collagenous capsular areas. Multinodularity of the lesion is evident. ×40

Clinically, schwannoma is seen commonly in adults of the age group ranging from 10 to 40 years, with a female predilection of 2:1[1]. To date, approximately 146 cases of soft tissue schwannomas were reported in the oral cavity usually involving the tongue, vestibule and palate. Most of them present as smooth surface, usually painless swelling with an intact overlying epithelium[2]. Approximately 44 cases of intraosseous schwannomas are reported to date in the English literature, accounting for <1% of all benign intraosseous tumours[3]. Clinically, they typically present as painless swelling of the mandible and radiographically, they show a well-demarked, unilocular radiolucency with external root resorption of the associated tooth[1,3].

Our case lies largely typical in the age group and site of involvement, arising in a 54-year-old male in the posterior part of the mandible, though such occurrences are predominantly seen in females. In contrast to the conventional radiographic picture of a smooth, unilocular, well-defined radiolucency, our case showed a well-demarcated multilocular radiolucency, causing different degrees of resorption in relation to 37 and an intact lower border inspite of large blown-up appearance of posterior one-third and ramus of the mandible.

Pathological examination of the resected specimen revealed evident tumoural multinodularity, which were of varying sizes ranging from 0.5 cm to 1.0 cm of variable number and are matted by fibrous intervening strands. The above presentation completely differed from conventional intraosseous schwannoma, pointing towards an anatomically unique variant characterised on gross examination and/or microscopically by intraneural, plexiform, and often multinodular growth called as plexiform schwannoma[4,10]. Plexiform schwannoma was first described in 1978[10]. It often arises in superficial soft tissues, particularly of the head and neck region which are abound with superficially situated peripheral nerve. This lesion represents 4.3% of all schwannomas, 23% of schwannomas of head and neck soft tissues, with only 28 reported cases[4]. The present case, a true central tumour with multinodular growth, seems to be among the first reported cases.

Microscopically, a confirmation for plexiform schwannoma was reached, as the tissue in lower magnification (×40) showed multinodularity in almost all the areas. A higher magnification showed that the nodules were covered by fibrous and cellular connective tissue capsule. The nodules showed features of conventional schwannoma with compact cellular arrangement with palisading nuclei, forming characteristic Verocay bodies (Antoni type A tissue) and occasional loose texture areas, with cells having multiple processes and capillaries showing perivascular hyalinisation (Antoni type B tissue). A histopathological diagnosis of schwannoma was easy to reach. The tissue showed a strongly positive immunohistochemical staining with S-100 in the nodules, confirming the nature of the lesion to be of neural tissue. S-100 was completely negative in the superficial capsular areas and in the areas which were intervening and fibrous in nature. Epithelial membrane antigen was seen to be positive in only the periphery. Collagen IV was seen to be moderately positive uniformly. Ki-67 was very mildly positive in a few areas.

Plexiform schwannoma is a benign tumour with no malignant potential[4,11], but may recur when incompletely excised. Generally, they are solitary and sporadic in occurrence[1,4]. Multiple lesions are seen in association with NF-2 and schwannomatosis[4,8,9,12]. Syndrome-associated plexiform schwannoma accounts for 11% of plexiform schwannomas and are generally seen occurring in a paediatric setting[4]. The present case showed no evidence of increased cellularity or cellular changes like epitheloid-like cells linked with malignant changes[7]. No nuclear pleomorphism and mitotic figures were seen, indicating the absence of features of malignancy at cellular or tissue level. A detailed case history, examination and diagnostic set up revealed no other swellings in the body which ruled out the association of NF-2 and schwannomatosis.

In summary, we report a case of intraosseous plexiform schwannoma, a rare and unique variant of Schwann cell tumour at a central location in the mandible and can be the first of its kind to be documented in the English literature to date.

Clinical relevance

Though both plexiform neurofibroma and plexiform schwannoma appear similar on clinical and macroscopic examination, the prognosis of the two lesions varies greatly. Plexiform neurofibroma is generally associated with syndromes and has a higher malignant transformation rate, while plexiform schwannoma is less frequently associated with syndrome. The tumour also has a different behavioural pattern, rarely becoming malignant. Hence, it is very important to diagnose the lesion correctly as the treatment modality of these two lesions differs.

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