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Baranović, Marijan; Macan, Darko; Begović, Elvis Anđelko; Lukšić, Ivica; Brajdić, Davor; Manojlović, Spomenka

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Schwannoma with secondary erosion of mandible: case report with a review of the literature

Marijan Baranović, DMD,[a] Darko Macan, DMD, PhD,[b] Elvis Anđelko Begović, DMD,[c] Ivica Lukšić, MD, MSc,[d] Davor Brajdić DMD, MSc,[e] Spomenka Manojlović MD, PhD,[f] Zagreb, Croatia

[a] Senior resident, Department of Oral and Maxillofacial Surgery, University Hospital Dubrava.

[b] Associate Professor, Department of Oral and Maxillofacial Surgery, University Hospital Dubrava, School of Dental Medicine, University of Zagreb.

[c] Senior resident, Department of Oral and Maxillofacial Surgery, University Hospital Dubrava.

[d] Assistant, Department of Oral and Maxillofacial Surgery, University Hospital Dubrava, School of Medicine, University of Zagreb.

[e] Senior resident, Department of Oral and Maxillofacial Surgery, University Hospital Dubrava, School of Dental Medicine, University of Zagreb.

[f] Associate Professor, Department of Pathology, University Hospital Dubrava, School of Medicine, University of Zagreb, Croatia

Correspondence: Prof.dr.sc. Darko Macan, Department of Oral and Maxillofacial Surgery, University Hospital Dubrava, Av. G. Suska 6, 10000 Zagreb, Croatia. Phone: +385 91 4664 075; Fax: +385 1 2864 250; e-mail: darkom@kbd.hr

Running title: Schwannoma with mandibular erosion

Schwannoma with secondary erosion of mandible: case report with a review of the literature

Abstract

Schwannoma (Neurilemmoma) is a common, histologically distinctive, benign, usually encapsulated, peripheral nerve tumor of Schwann cell origin. We report a case of schwannoma arising from soft tissue near the mandible. A 53-year-old female presented at our department with painless swelling of lingual mucosa of the mandible. The first molar was extracted 20 months before. Panoramic radiograph showed suspected residual cyst. It was impossible to determine prior to surgery that this was a peripheral nerve sheath tumor. The lesion was completely removed; the tumor appears to have originated in soft tissue and caused secondary erosion of the mandible.

Key words: schwannoma; *neurilemmoma*; mandible; bone, erosion

Introduction

Schwannoma (also known as neurilemmoma, neurolemoma, neurinoma, perineural fibroblastoma, peripheral glioma, and peripheral nerve sheat tumor) is a slow-growing, benign neoplasma, derived from Schwann cells which are sheath cells that cover myelinated nerve fibers. Schwannomas may be encapsulated and can appear anywhere in the body but are more frequently located in the head and neck.[1-5] Most commonly, the tumor appears in the paravertebral region of the retroperitoneum, pelvis, mediastinum, extremities, nasal cavity, nasopharynx, orbit, parapharyngeal space, larynx and oral cavity.[6] Intraoral development is uncommon (only 1%). In this area in a decreasing order of frequency the mobile position of the tongue, the palate, the cheek mucosa, the lip and gingiva are the most frequent locations.[7] In the tongue, the tip is the least affected part.[8-12]

This lesion has been widely reported, but it was only hypothesized that one of the mechanisms by which they involve the bone is through secondary erosion from a soft tissue or periosteal tumor.[13-15] We present a case that confirms this hypothesis.

Case report

A 54-years-old female presented at our department complaining of swelling on the lingual mucosa of the mandible (Fig. 1). The swelling was noticed ten months earlier and the patient did not report pain or paresthesia. During the examination, a firm, nodular, soft mass, measuring 10 mm in diameter was observed at the lingual mucosa of the mandible. The nodule was bounded to part of the mandible, covered by intact mucosa.

A Panoramic radiograph showed a well-defined, unilocular radiolucency with a thin, uniformly sclerotic margin in the body of the mandible. A soft tissue mass was evident above the alveolar crest. The radiolucency was above the mandibular canal which was not involved and no teeth were present in the area (Fig. 2). Under local anesthesia, a lingual mucoperiosteal flap was reflected and round, pale yellow encapsulated mass at the lingual part of the mandible was identified. The mass was removed *in toto* (Fig. 3). Beneath the mass we noticed secondary erosion of the mandible, which surface was smooth (Fig.4). During the surgical removal we could not identify the nerve from which the tumor derived. There was no relationship between the tumor and the underlying impressed bone.

An oval, sharply demarcated, encapsulated, firm nodule measuring 11 mm in diameter was submitted. The cut surface was yellowish-white and smooth. Microscopic analysis revealed the tumor mass composed of the interlacing fascicles of compact spindle cells with twisted nuclei (Fig. 5). The nuclear palisading formed the Verocay bodies (Fig. 6). A fibrous capsule surrounded the tumor nodule.

Discussion

Oral peripheral nerve tumors include schwannoma, neurofibroma, nerve sheath myxoma, palisaded encapsulated neurinoma, mucosal neurinoma associated with multiple endocrine neoplasia III, traumatic neuroma, and granular cell tumor.[16]

There are 3 mechanisms by which schwannomas may involve bone: (1) a tumor may arise centrally within bone, (2) a tumor may arise within a nutrient canal and produce canal enlargement, or (3) a soft tissue or periosteal tumor may cause secondary erosion and penetration into bone.[13-15]

This case demonstrates an example a schwannoma secondarily involving bone.

Schwannomas most often occur in the fourth and fifth decade of life with a 1.6:1 female predilection. The duration of symptoms varies from a few months to a few years. The majority of these tumors have a long duration because of their lack of symptoms and slow growth.

A review of the English literature revealed 3 cases similar in clinical and radiological features as ours. In 1954, Bruce[17] described a tumor, which was located on the left edentulous mandibular alveolus. Dental radiographs revealed some osteolysis in the mandibular alveolus subjacent to the tumor. Surgical excision and enucleation of the tumor left a smooth concavity in the body of the mandible which was not associated with the mandibular canal or nerve, but histologically it was neurofibroma. Worth[18] describes neural sheat tumors that arise subperiosteally causing saucerization of the bone. These tumors are radioluscent and may or may not have a cortical outline. One such neurofibroma was reported by Schneider[19] et al. The central portion of the lesion was described as mottled, and at surgery, the lesion appeared to be covered with bone. Mortada[20] and Sciubba and Sachs[21] reported cases of schwannomas with secondary penetration into the bone, but they could not determine if the lesion arose centrally or from the periosteum. Kun[22] et al. reported that preoperative diagnosis was correct in only 4 out of 49 cases in their study. They concluded that it was difficult to make a confirmed diagnosis on the basis of imaging findings.

Radiographically, schwannoma is commonly unilocular and associated with bone resorption.[23] It may resemble many benign conditions such as odontogenic or periodontal cyst, ameloblastoma, angioma, and benign odontogenic tumor. If the tumor is large with destruction of cortical bone it may resemble a malignant lesion.[24] Some schwannomas have reportedly turned malignant,[25,26] and mandibular malignant schwannoma has been reported.[27] When degenerative changes are very pronounced,, calcifications, hyalinizations, haemorrhages and atypical nuclei will appear as well as cystic formations, but these changes do not lead to malignancy.[28,29] The preoperative diagnosis of the schwannoma is rare,[28] although with digital intravenous subtractionangiography, CT scans and MRI, the probabilities are increased.[30] Magnetic resonance patterns for neurofibromas are characteristic: low-to-intermediate signal intensity on T1-weighted images; enhancement of the solid component of the tumor after administration of contrast medium; heterogeneity on T2-weighted images; multiple target signs due to a central collagen area (some patients).[31] MRI findings of intraosseous schwannoma of the mandible help in differentiating solid from purely cystic lesions (eg, dentigerous cysts, periodontal cysts).[32] Yamazaki et al.[33] reported that ultrasound and MRI were effective in preoperative imaging diagnosis of schwannoma originating in the mental nerve. They also suggested that the compatibility of photographic parameters in MRI techniques for identifying nerves, particularly the final branch with a short diameter in the extracranial region, requires careful discussion in the future. On MRI, tumor can be delineated as solid, cystic, or mixed based on its pathological characteristics. Because the imaging findings are variable, it is difficult to arrive at a confirmed diagnosis based only on such findings. In their case, malignancy could not be completely ruled out by preoperative imaging findings. Wakoh et al.[34] reported two cases of schwannoma displaying marked cystic changes; one in the temporalis muscle and one in the submandibular space. They concluded that MRI should depict the nerves and allow identification of the origin of a schwannoma. MRI can show not only the tumor and the capsule but also, in certain cases, the nerve from which it has developed.[35] Only a few cases of schwannoma in the oral floor have been reported. However, when the characteristic findings are

observed on CT and MRI, schwannoma should be added to the differential diagnosis.[36] In their presented case, based on the preoperative CT and MRI findings, a malignant tumor derived from the sublingual gland was suspected. Intraoperatively, adhesion of the mass to circumferential regions was not observed, but nerves penetrated into the mass at several places. Based on operative findings, the mass was thought to be a tumor derived from the lingual nerve. Almeyda et al.[37] reported a case of submandibular schwannoma (3.5 x 2 x 2 cm) misdiagnosed pre-operatively. The differential diagnosis of adenolymphoma (Warthin's tumor) was based on the clinical examination, US and two fine needle aspirations. Intraoperatively the surgeon noted adherence of the tumor to a branch of the lingual nerve. The mass and submandibular gland were excised "en block". Even with advances in imaging, the diagnostic dilemma remains.[37] MRI has been of greater use, with a distinctive target pattern demonstrated by most, but not all schwannomas.[38] In the case presented by Asaumi et al.[39] the US, CT, and MRI appearance of schwannoma of the upper lip (3.8 x 1.8 x 1.4 cm) correlated well with the histologic features. MRI was particularly helpful in showing the internal characteristics of the encapsulated mass. They concluded that, however, because most tumors of the upper lip present as relatively small lesions, establishing the differential diagnosis using US, CT, and MRI should not be considered as routine or necessary. We completely agree with this attitude because in our case, the tumor measured 1 cm in diameter.

Although schwannomas originate from the nerve tissue, locating the nerve of origin exactly can be impossible. Direct relation with a nerve can be demonstrated in approximately 10 - 50% of cases.[7,28,40-43] Tumors arising from the small nerves are freely mobile but mobility is restricted along the axis in those arising from large nerves.[44] The growth of these lesions will cause the displacement and compression of the surrounding normal nerve tissue.[12] Yamazaki et al.[33] stated that the lesions are diagnosed as peripheral nerve sheat tumor when the tumor is connected directly to the nerve, even though the nerve itself can not be identified. However, in their presented case the possibility of malignancy could not be ruled out from the preoperative imaging findings and the clinical course. The nerve of origin is often not identified at the time of surgical excision, although

if presented it is displaced to the side by the expanding tumour.[37] Arda et al.[45] presented a schwannoma arising from the parasympathetic fibers of the lingual nerve. They found only a few nerve fibers, which were thought to be the parasympathetic nerve of the sublingual gland, attached to the tumor. Results of CT imaging did not help them preoperatively for the diagnosis of the mass, and FNAB was not useful. Garcia de Marcos et al.[46] presented nine cases of schwannomas; five of them were localized intraorally, and for these no preoperative test were carried out. For seven out of nine schwannomas determining of the nerve of origin was not possible.

In the differential diagnoses neurofibroma, granular cell tumors, lipoma, fibroma, leiomyoma, rhabdomyoma, nerve-sheat myxoma, adenoma, neuroma, granular cell tumor, neurothekeoma and perineurioma should be considered.[8,31,47,48] The differentiation between schwannoma from neurofibroma is essential because an apparently "solitary" neurofibroma may be a manifestation of neurofibromatosis. Fifteen to sixteen percent of patients with neurofibromatosis will present malignant transformation in one or more lesions, contrary to schwannoma.[49] The recurrence rate of a schwannoma is lower than that of a neurofibroma because of encapsulation.[50] The differentiation between these two neoplasms is imperative because neurofibromas tend to recur frequently and have the potential for malignant transformation. It is difficult, however to differentiate an intraosseous schwannoma from an ameloblastoma associated with a substantial solid component.[51] A small and slow-growing mass in the tongue with positive history of tongue bite is first suggestive of schwannoma, as well as neurofibroma, lingual cyst, and minor salivary gland tumor.[52] Neurofibromas lack the thick collagenous capsule of schwannomas and instead are surrounded by a variably thickened perineurium and epineurium. Neurofibromas also lack the Antoni type A and B patterns and Verocay bodies typical of schwannomas. Immunoreactivity for S-100 protein is observed in only a portion of the cells comprising a neurofibroma, as opposed to uniform reactivity throughout an schwannoma.[5] Neurofibroma is generally non-encapsulated and lobulated, with an irregular surface, and unlike the schwannoma which pushes away the associated nerve, it becomes intertwined with the nerve of origin.[28-30] Neurofibroma is difficult to remove, it

recurs or persist when resection has been incomplete, and in cases of neurofibromatosis, it can transform into a malignant tumor.[29] Malignant transformation of schwannoma is in contrast to neurofibroma, an exceptionally rare event and for practical purpose can be discounted.[53]

In conclusion, a rare case of schwannoma with secondary erosion of the mandible was reported. The tumor may have originated from a branch of mandibular nerve in the mucoperiosteum of the alveolar gingiva and extended into the body of the mandible, creating a bony defect.

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Figure 1. Well-encapsulated tumor at the lingual part of the mandible.

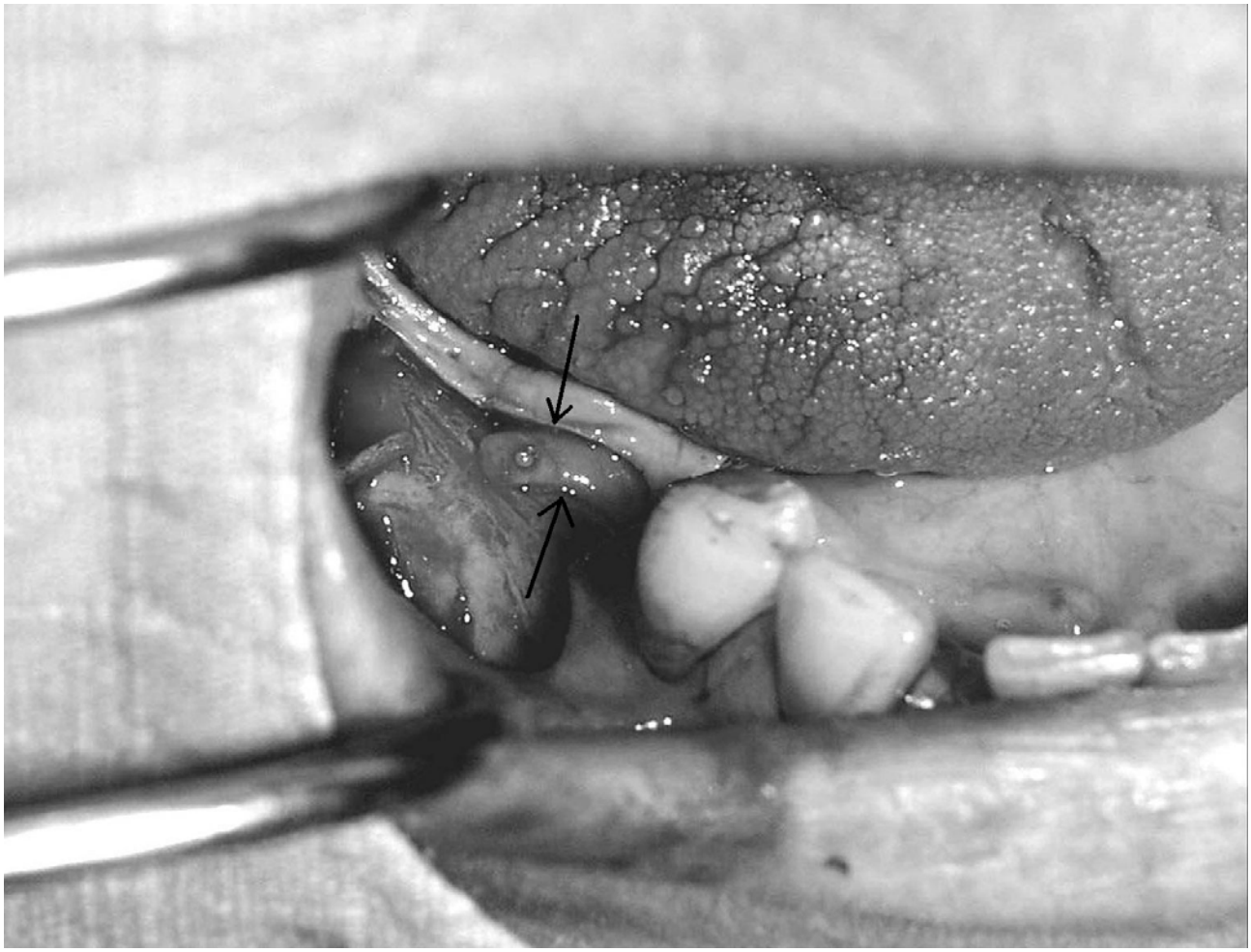

Figure 2. Panoramic radiograph showed the radiolucency above the mandibular canal lifting the soft tissue.

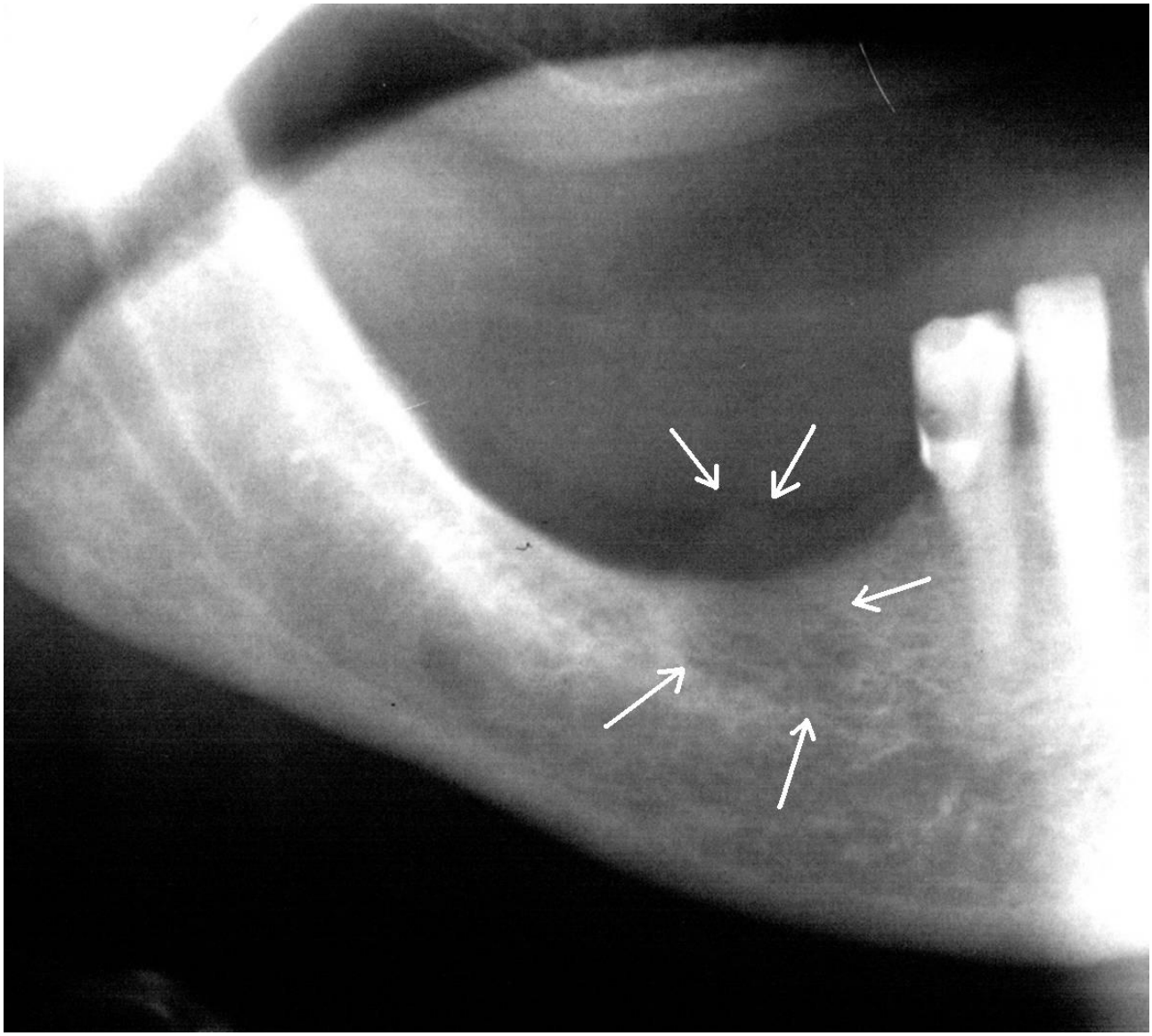

Figure 3. The tumor removed in toto.

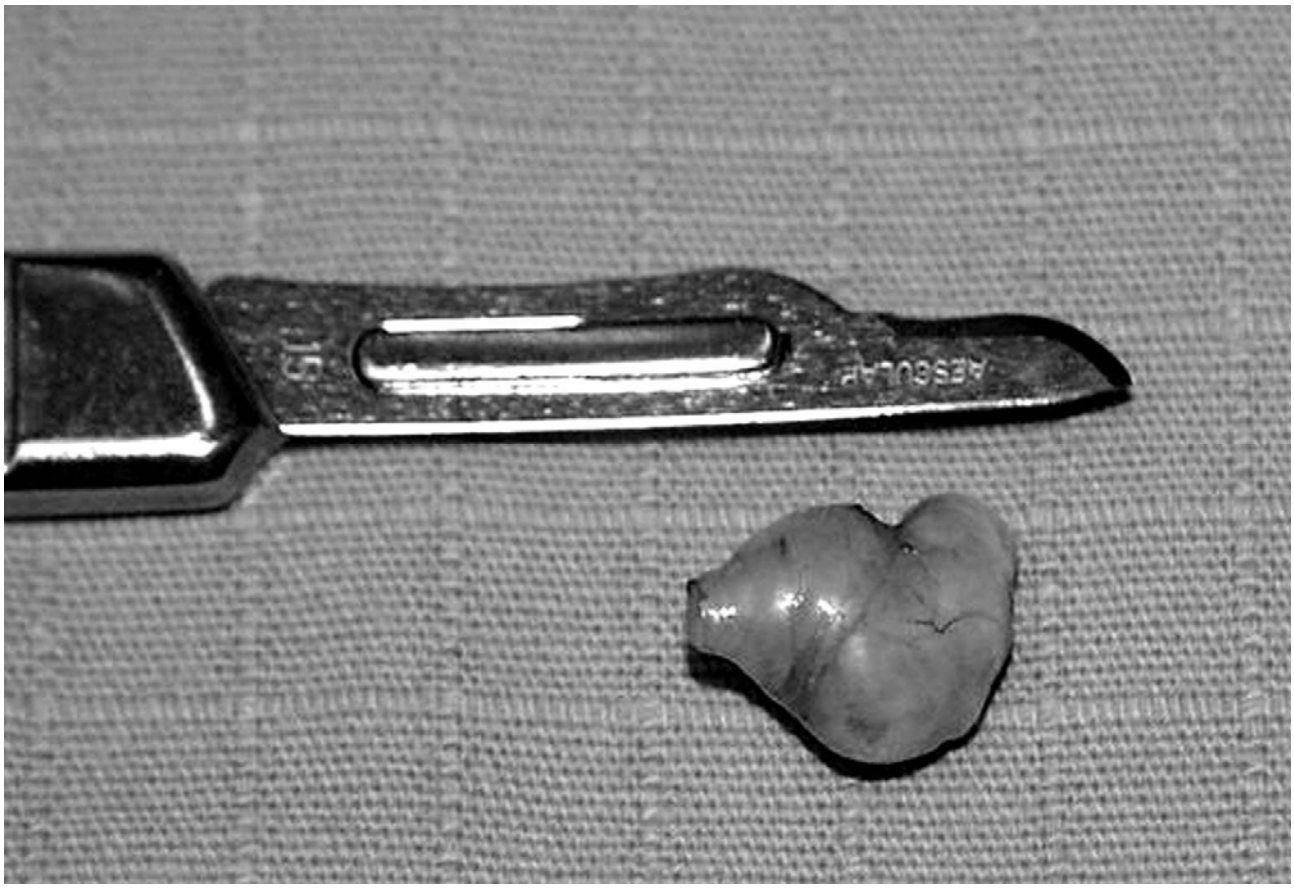

Figure 4. Secondary erosion of the mandible beneath the tumor.

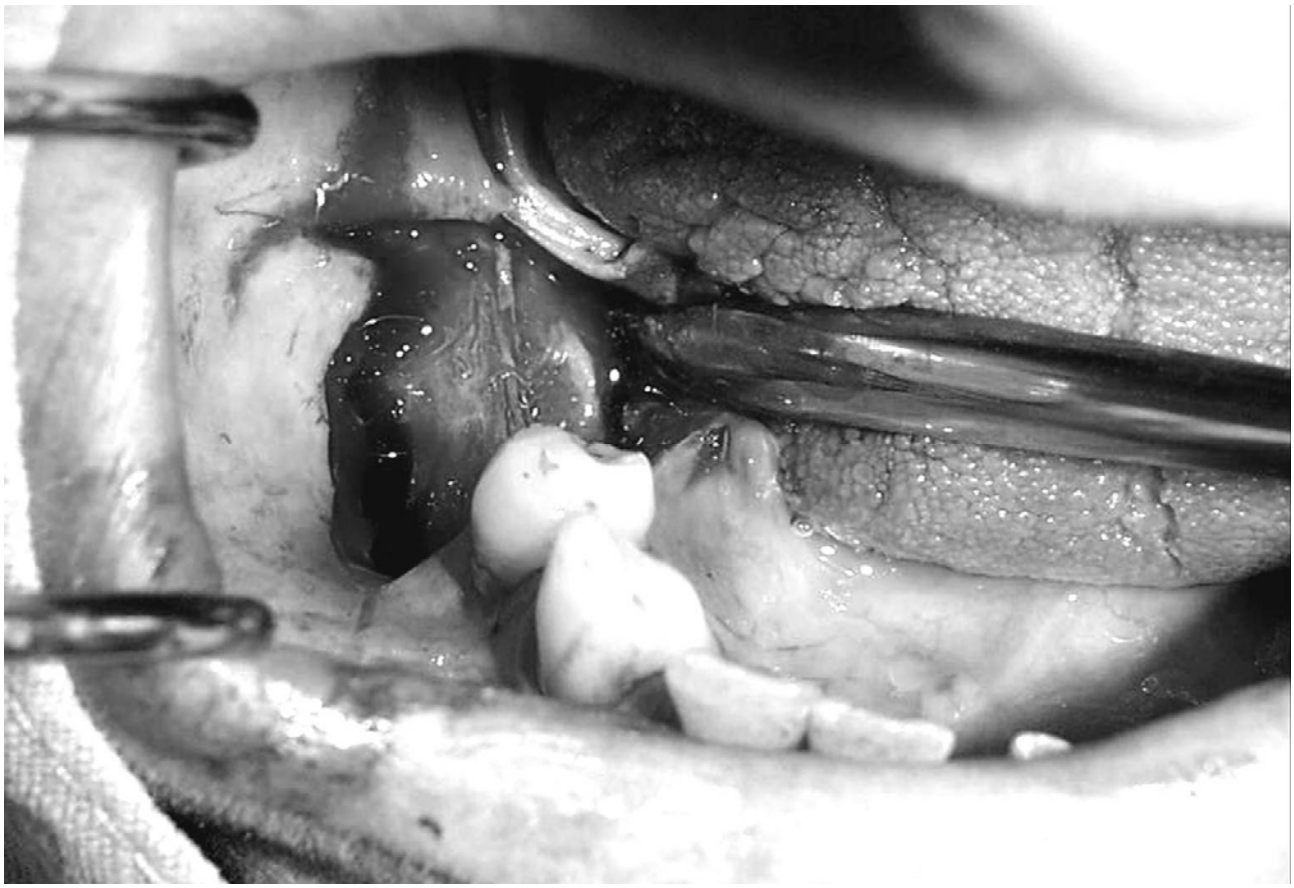

Figure 5. Interlacing fascicles of compact spindle cells with twisted nuclei (haematoxylin and eosin stain x 100).

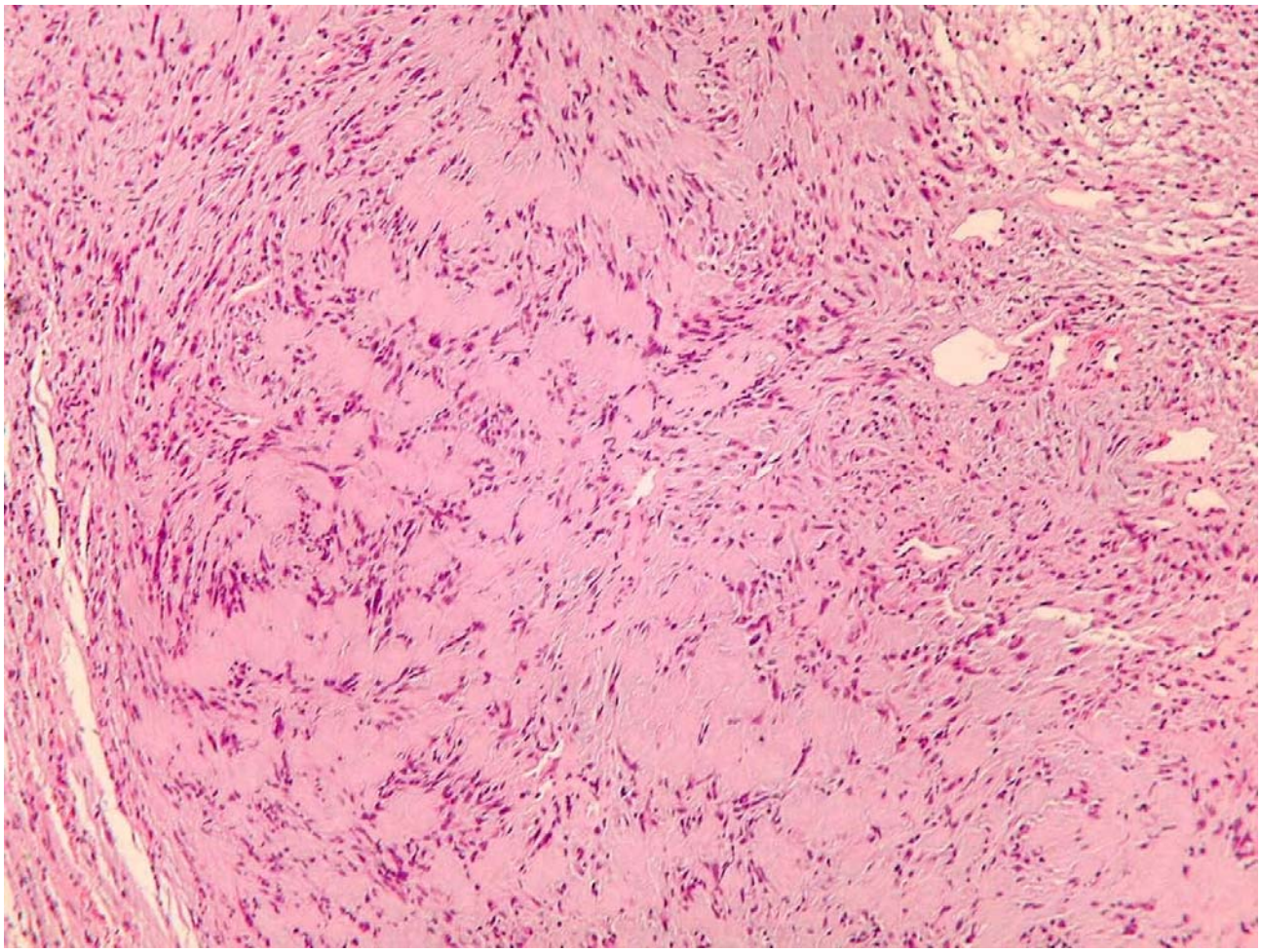

Figure 6. The nuclear palisading formed the Verocay bodies (haematoxylin and eosin stain x 200).