Central Mandibular Nerve Sheath Myxoma

[1]Narges Ghazi, Jahanshah Salehinejad[2], Shapoor Yaripour[3], Arash Dehghan[4], Elham Baradaran Nasseri[5], Mehdi Shahabinejad[6]

[1]Oral & Maxillofacial Diseases Research Center, Mashhad University of Medical Sciences, Mashhad, Iran
[2]Department of Oral and Maxillofacial Pathology, Mashhad Dental School, Mashhad, Iran
[3]Department of Oral and Maxillofacial Surgery, Hamedan Dental School Hamedan, Iran
[4]Department of General and Clinical Pathology, Hamedan University of Medical Science, Hamedan, Iran
[5]Department of Medicine, Mashhad Dental School, Mashhad, Iran
[6]Department of Oral and Maxillofacial Pathology, Mashhad Dental School, Mashhad, Iran

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Abstract

Nerve sheath myxoma has been described as a rare neural tumor arising from Schwann cells. It is observed most frequently in the central area of the face, neck and upper extremities. In the past the term neurothekeoma was used as synonym for nerve sheath myxoma but according to new reports, they are separate entities which can be confirmed by immunohistochemistry as in our case. Oral involvement of this tumor is extremely rare. Here, we present an unusual case of nerve sheath myxoma in the mandible of a 22-year old female patient. This case appears to be the first myxomatous variant which is centrally located in the mandible.

Keywords: nerve sheath myxoma, schwann cells, neurothekeoma, mandible.

Introduction

Nerve sheath myxoma (NSM) is an uncommon, benign nerve sheath tumor which was described in 1969 as a distinct pathologic entity (1-2). In the past the term neurothekeoma was used as synonym for nerve sheath myxoma but according to new reports they are separate entities which can be confirmed by immunohistochemistry (3). The microscopic diffrential diagnosis of nerve sheath myxoma include myxoid neurofibroma and myxoid neurothekeoma.

neurothekoma is S100 negetive while nerve sheath myxoma is S100 positive as in our case.

It is observed most frequently in the central area of the face, neck and upper exterimities. A slight female predilection was reported (1.8:1) with the mean age of 25 years (4).

Histologically, it is characterized by the presence of epithelioid and spindle cells arranged in well-formed micronodules. Myxoid, mixed and cellular are the three histologic variants of NSM (4).The treatment of choice is wide local exision with free surgical margins and it has local recurrence rate of 50% but no metastasis or malignant transformations has been reported.

Oral involvement is extremely rare. Here, we present an unusual case of nerve sheath myxoma centrally located in the mandible of a young patient.

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Case report

A 22-year old female presented with a chief complaint of left mandibular swelling in the area of tooth #38 that had been started two weeks ago.
The patient also had numbness of left side of lower lip and pain during chewing.

Clinical examination revealed a firm swelling, present on the left mandibular angle which was extended intraoraly in the area of partially erupted tooth #38 (Fig. 1).

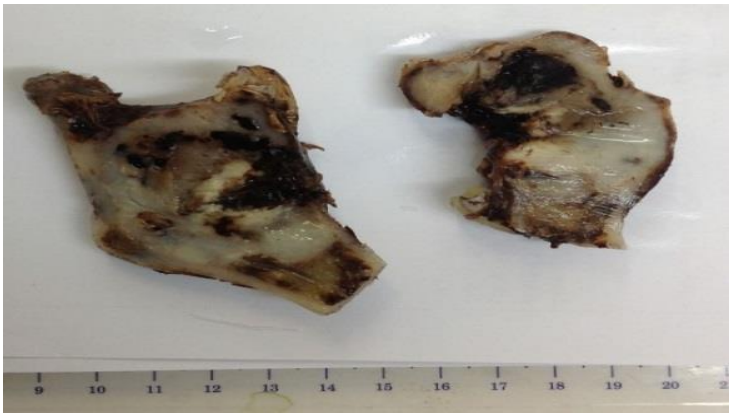

Figure 1. Gross appearance of resected mandible

Panoramic radiograph demonstrated a well- defined unilocular radiolucency extending from distal of tooth #37 up to the inferior border of condyle and coronoid causing obliteration of mandibular canal and thinning of inferior border of mandible (Fig. 2).

Computed tomography (CT) and 3D images showed a 44×22mm lesion with massive bone destruction and thinning of mandibular border which extends medially. A provisional diagnosis of KOT (keratocystic odontogenic tumor), ameloblastoma and myxoma was made (Fig. 2).

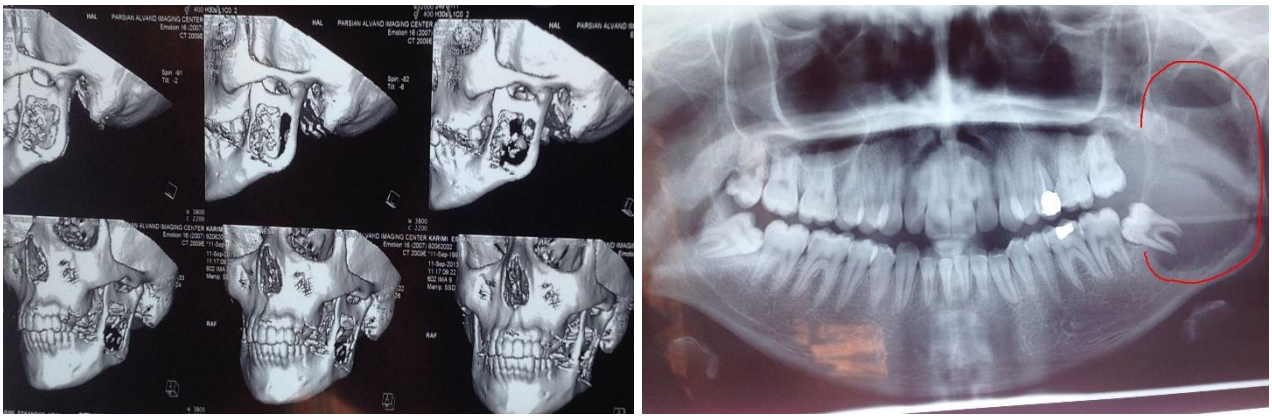

Figure 2. Panoramic radiograph and 3D images demonstrated a well- defined unilocular radiolucency with massive bone destruction and thinning of mandibular border

After incisional biopsy, the specimen submitted to the Pathology Department for histologic examination. Histopathological examination revealed myxomatous tissue consist of fine and scarce collagen fibers and few fibroblast together with sections of blood vessels. According to histopathological findings an initial diagnosis of myxoma was made (Fig 3).

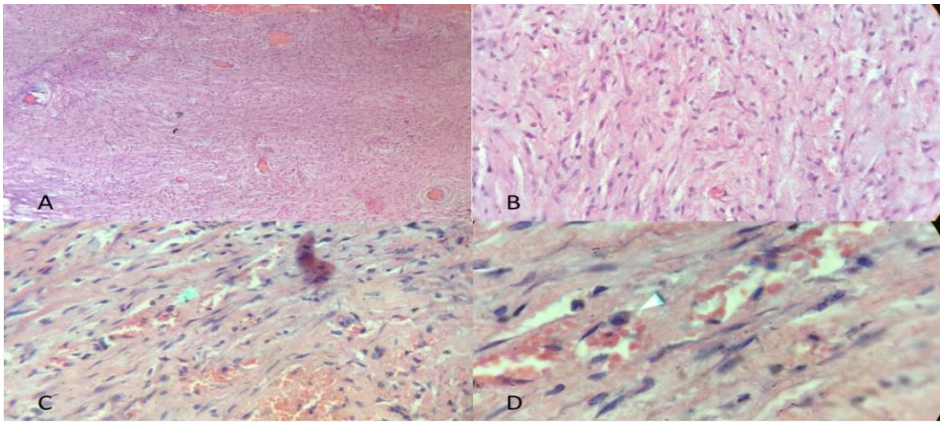

Figure 3. Phothomicrograph showing A, B: Round to oval cells along with spindle cells with wavy nuclei against a myxoid background, H&E, original magnification, A: 100x, B:400x, C: Mast cell in 100x and, D: 400x

Regarding the extension of the lesion and histopathologic evaluation hemimandibulectomy was performed. The gross examination demonstrated a section of mandible measuring 8.5 ×4×2.5cm with irregular surface and boldging in some areas. (Fig. 1)

Microscopic examination showed myxomatous tissue consist of fine and scarce collagen fibers together with proliferation of small, spindle shape cells with wavy nuclei similar to Schwann cells .

Immunohistochemical examination of markers including vimentin, S-100, and neurofilament confirmed the diagnosis of nerve sheath myxoma (Fig. 4).

Intense positivity for vimentin, scattered positivity for S-100 and very scant positivity of neurofilament specially in the area of nerve bundles was present. (Fig. 4). Patient follow up had been done a year after and she was free of any recurrences.

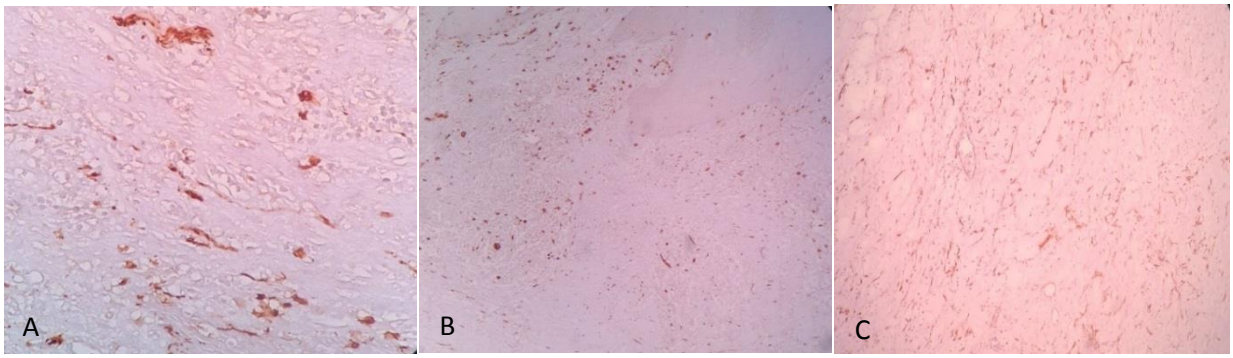

Figure 4: Photomicrograph showing A,B: S-100 imunoreactivity A:400x, B:100x, C: Vimentine immunoreactivity, 100x.

Discussion

Nerve sheath myxoma is a rare neural tumor arising from Schwann cells. Rodriguez and el-Naggar reported that the mean age of patients with oral nerve sheath myxoma is 33 years (5).

Oral involvement of this tumor is extremely rare. Tongue is the most common site which has been reported by several authors (6-8). Involvement of retromolar area (9), palate (10-11) and buccul mucosa (1, 5, 9) have been also reported. Intra-osseous nerve sheath myxoma of the jaw is very rare. kaltman et al. reported a cellular varient of mandibular NSM and it was S-100 negetive (12).

Our case is myxomatous varient and is the first case which is centrally located in mandible. In our case

the neoplastic elements demonstrated S-100 immunoreactivity which according to argenyi et al. the myxomatous variant is S-100 positive (13). Positive reactivity for S-100 has been also reported by several authors (3,8,11).

Histopathological differential diagnosis for NSM are myxoid neurothekeoma, neurilemmoma, myxoid neurofibroma and oral myxoma (14). In myxoid neurothekoma the myxoid area is located at the periphery of tumor but in our case it is more of central location and myxoid neurothekoma is S100 negative.(3) Both of NSM and neurilemmoma arise from the nerve sheath and may show positive reactivity for S100. Myxoid appearance is rarely seen in neurilemmoma.

Although areas of increased cellularity may be seen in NSM similar to neurilemmoma, verocay body formation is not observed in NSM (14).

The histopathologic feature of myxoid neurofibroma is a circumscribed and unencapsulated tumor with slender, closely aggregated fascicles of Schwann cells. The myxoid matrix is confined within fascicles in NSM, whereas in neurofibroma it usually involves both fascicles and surrounding tissue (8). It should be noted that lobular pattern and S100 immunoreactivity are not seen in oral myxomas (5).

The treatment of choice for NSM is local excision (8). Regarding the extension of the lesion and histopathologic evaluation hemimandibulectomy was performed for the present case.

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Corresponding Author:
Mehdi Shahabinejad,
Oral and maxillofacial pathology,
mashhad dental school,mashhad university of medical science, mashahad, iran
Tell: 05138829511
Email: shahabinejadm@mums.as.ir