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EXTRA-NEURAL PERINEURIOMA OF MANDIBLE : A RARE PRESENTATION

Pathology

Dr Sumanta Das — Resident , Department of Pathology, Vardhman Mahavir Medical College and Safdarjung Hospital

Dr Charanjeet Ahluwalia* — Professor, Department of Pathology, Vardhman Mahavir Medical College and Safdarjung Hospital *Corresponding Author

DR Rashmi Arora — Professor and Head, Department of Pathology, Vardhman Mahavir Medical College and Safdarjung Hospital

ABSTRACT

Perineurioma is a soft tissue tumor composed of cells resembling normal perineurium.Lazarus and Trombetta first described it in 1978 on the basis of ultrastructural findings. All perineuriomas express antigens that are identical to normal perineurium of nerve. Soft tissue perineurioma is most commonly seen in extremities and trunk. It has been very rarely described in gingiva, mandible, lips, retrotonsillar mucosa and maxillary vestibule. We present a case of soft tissue type perineurioma arising in mandible, an extremely rare site.

KEYWORDS

INTRODUCTION

Perineurioma is a soft tissue tumor derived from perineurium and resembles normal perineurium.[1] It was forst described by Lazarus nd Trombetta in 1978.[2]Perineural cells are mesodermal origin cells sharing an immunophenotype with the cells of the pia-arachnoid (S-100 negative; EMA, GLUT-1 and Claudin-1 positive).[3]

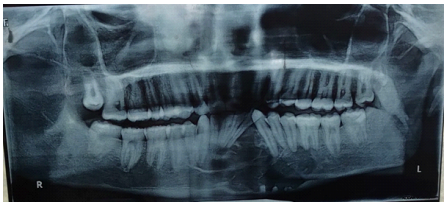
Ultrastructurally perineural cells form close junctions with each other and have basal lamina along the endoneurial and perineurial aspects of the cell which are not present in schwann cells and fibroblasts).

Perineuriomas are benign peripheral nerve sheath neoplasms composed of perineural cells with characteristic immunohis tochem ical and ultrastructural features. There are 4 types of perineurioma : intraneural, extraneural (soft tissue), sclerosing and reticular. Soft tissue perineurioma is the most common among these but it is still uncommon.[4-6] It affects primarily adults, equally in both sexes. Superficial soft tissue of extremity and trunk are the most common sites involved but 30% develop in deep soft tissue and rarely in visceral organs.[5]

CASE REPORT

A 16 year old adolescent male presented with swelling in left side of chin since 9 months. There was no history of trauma. The swelling was initially small in size and the size gradually increased over time to attain the current size. On examination the swelling was 3*3 cm in size on left side of symphysis menti. It was hard and non-mobile.

X-ray showed radiolucent expansile lesion causing divergence of root. CECT showed well-defined lytic expansile lesion in the body of the mandible crossing the midline upto approximately 1cm on right side with enhancing soft tissue components suggestive of a benign lesion likely to be ameloblastoma or giant cell granuloma.

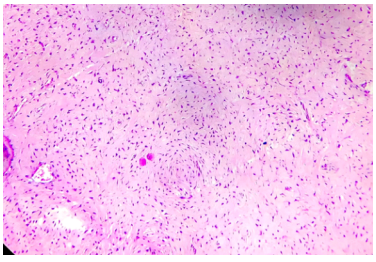


Xray showing radiolucent expansile lesion with tooth divergence

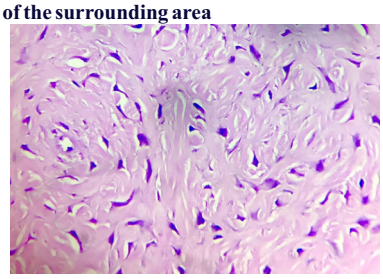
We received multiple soft tissue fragments along with part of mandible and teeth together measuring 5.5x5x2 cm. A grey white tumor was identified measuring 3x3x3 cm.

Microscopically showed a spindle cell lesion composed of slender fibroblast like cells with long cell process arranged in anastomosing pattern. They were also forming whorls. Individual cells had long eosinophilic cytoplasmic process and spindle wavy nuclei with minimal atypia and pleomorphism. Tumor cells were seen dissecting in between collagen bundles. No infiltrative border was present. Based on histomorphology, perineurioma was considered first impression. On immunohistochemistry the tumor cells were positive for EMA and they were negative for S-100, CD-34, SMA,Desmin,ER and PR. So the final diagnosis of soft tissue type perineurioma was made.

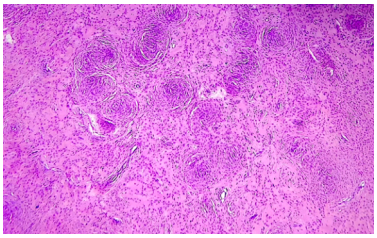
Part of the mandible along with multiple tissue fragments showing a grey-white tumor



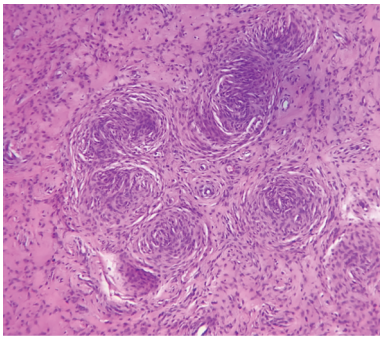
H&E (4X) : Spindle cell tumor with well-defined margin with no infiltration of the surrounding area



H&E (40X): Tumor cells are spindle shaped with long cytoplasmic process seen in between collagen



H&E (4X): Spindle cell tumor cells arranged in whorls



H&E (10X): Spindle shaped tumor cells arranged in whorls

DISCUSSION

Peripheral nerves consist of 3 layers with differing characteristics: the endoneurium, perineurium, and epineurium. The perineurium represents a continuum with the pia-arachnoid from the central nervous system and extends distally with the sheath of capsular cells of peripheral sensorial organs and propioceptive receptors. It is made of layers of flattened cells surrounded by a basement membrane and collagen fibers, forming concentrically laminated structures around single nerve fascicles. Functionally, the perineurium modulates external stretching forces (that could be potentially harmful for nerve fibers), and along with endoneurial vessels, forms the bloodnerve barrier.

The perineurium was first described in 19th century by Friedrich Gustave Jacob Henle.[7] The perineurium along with endoneurial vessels maintains endoneurial pressure and Blood-Nerbe-Barrier.[8]

The perineurium forms a tubular sheath composed of specialized concentrically oriented layers of flattened (perineurial) cells, surrounded by a continuous basement membrane and separated by layers of collagen and extracellular matrix. The number of cell layers varies from nerve to nerve and also depends on the size of nerve fascicles and proximity to the meninges. As the number of fascicles increases within a nerve, the width of the perineurium decreases. The number of perineural cell layers also decreases close to nerve endings or as the nerve branches, and increases at the bifurcation of nerves to give additional protection to nerve fibers.[9,10]

Multiple pathologic conditions associated with the perineurium have been described. Perineurial invasion is considered an important prognostic factor in several malignant neoplasms. Perineuriomas are true benign infrequent perineurial cell neoplasms that have been divided in 2 categories: those with intraneural localization and a more common extraneural (soft tissue) group, including sclerosing and reticular variants.Sporadic cases of malignant perineuromas have been reported. Interestingly, neurofibromas and malignant peripheral nerve sheath tumors may also display perineurial cell differentiation.

Soft tissue perineuriomas usually affects superficial soft tissues of extremities and trunk. Very few cases have been described in oral cavity. It has been described in mandible,[11,12,13]upper lip[4],lower lip[14],gingiva[15,16],nasolabial fold[17] etc. Histologically they are characterized by spindle cell proliferation forming fascicles, whorls or they may be arranged in storiform pattern. Individual cells have elongated spindle shaped wavy nuclei and long slender bipolar eosinophilic process. Sometimes epithelioid morphology may be seen but pleomorphism and atypical mitosis are hardly seen. Tumor cells may be seen in between collagen and dissecting them. Loose myxoid stroma may be present.[1]

Immunohistochemically all the perineuriomas express EMA. Pattern of EMA staining is membranous which sometimes become difficult to interpret because of long cell process. Majority of perineuriomas also express claudin-1,[18] a tight junction associated protein and GLUT-1,[19] human erythrocyte glucose transporter.

Claudin-1 is easier to interpret because it shows diffuse and strongly positivity. The cells are negative for S-100, Neurofilament, Desmin, GFAP.[22-25] CD34 and SMA show positivity in 20-60% cases.[23] Electron microscopy shows slender non-tapered process containing large number of pinocytic vesicles and partial investment with basal lamina.

Important differential diagnosis include low grade fibromyxoid sarcoma (LGFMS), neurofibroma, dermatofibrosarcoma protuberans (DFSP), low grade malignant peripheral nerve sheath tumor (MPNST) etc.

LGFMS shows sharply demarcated zones of collagenized and myxoid areas and curvilinear blood vessels. They lack long cytoplasmic process typically seen in perineuriomas. Although EMA can be positive in both, MUC-4 is specific for LGFMS. DFSP can be excluded by poorly circumscribed margins with plump to spindle shaped cells which lack long cytoplasmic process and negative staining with CD-34. Neurofibroma and low grade MPNST show S-100 positivity.

Hornick and Fletcher had described in their case series of 81 cases of perineuriomas,[26] 14 cases had one or more atypical features,, including mitotic activity(13/30 hpf), occasional pleomorphic cells, hypercellular foci and infiltration of skeletal muscle. Only 2 cases recurred out of 81 cases and there was no metastasis with mean follow up for 41 months.

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