CASE REPORT

Myoepithelioma of hard palate: A case report
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Abstract
Myoepitheliomas (ME) are benign neoplasm originating from myoepithelial cells of seromucinous glands. Majority of these tumours arise in parotid gland followed by minor salivary glands of palate. The tumor does not show any unique clinicopathologic features that is different from other benign salivary gland tumors but is believed to have more aggressive behaviour. The histopathological patterns vary to very great extent and usually show overlapping features with other benign and malignant tumours. We report a case of 38-year-old male patient with a solitary swelling present on the right side of the hard palate. The histopathologic picture of the lesion showed spindle, plasmacytoid and epitheliod cells. The connective stroma showed areas of hyalinization and keratin pearl formation making it difficult to differentiate from pleomorphic adenoma as well as its malignant counterpart. An Immunohistochemical analysis was done using S100, smooth muscle actin, Ki-67 and a definitive diagnosis of ME was made.

Keywords: Benign myoepithelial tumor, cellular pleomorphic adenoma, myoepithelial adenoma, myoepithelial carcinoma, myoepithelioma

Introduction
Myoepithelioma (ME) is said to be a rare kind of tumor composed of an ectodermally derived cell having special contractile property like smooth muscle cells and therefore called myoepithelial cells, the term first used by Renault.⁰¹² They are usually seen in exocrine glands like salivary, lacrimal, mammary and sweat glands.⁰²³ ME are also known as myoepithelial adenoma and benign myoepithelial tumor.⁰¹ The tumor was first described and classified by Sheldon in 1943 as a different entity which was till then considered as a variant of pleomorphic adenoma (PA).⁴⁵ Burke et al. in 1995 were the first to report primary ME of soft tissue from retroperitoneal region.⁶⁷ ME of salivary glands together constitute <1% of all salivary glands tumors. Majority of the cases are reported in parotid gland and the minor salivary glands especially the soft palate.⁸⁹ The tumour is also recently reported in skin, soft tissue and bone.⁶⁷ The tumour is now considered as a separate pathological entity with biological behaviour distinct from PA and not a variant of PA. The ME although benign are understood to be more aggressive than PA.⁴

Case Report
A 38-year-old male patient visited the outpatient department of our institution with a complaint of swelling in the upper right posterior palatal region since 7 years, which showed rapid increase in size from 3 months associated with mild pain. The pain was localized, intermittent in nature, aggravated on food intake and subsided subsequently. Patient also gave a history of fever, difficulty in eating and speaking. Patient had visited a general physician for the same 3 months before, who did fine needle aspiration cytology (FNAC), the reports of which are not available and was on medication. Pain subsided, but ulceration was seen on the site of FNAC after few days. On intraoral inspection a well defined solitary swelling was seen on the hard and soft palate region measuring about 3 × 4 cm, extending from distal to 16 and crossing the midline and extending towards 26. Posteriorly the swelling extended towards the soft palate region and involved palatoglossal arch. Blanching of the surface epithelium and telangiectatic vessels were noted. The ulcer at the site of FNAC, showed everted edges and erythematous margins [Figure 1]. On intraoral palpation a well-defined solitary swelling which was soft to firm in consistency and attached to
underlying structures was felt. Swelling was tender on palpation, non fluctuant and non compressible. The computerised tomographic examination showed an ill defined heterogeneously enhancing soft tissue mass with cystic areas in the soft palate displacing the uvula [Figure 2]. The patient was referred to oral and maxillofacial surgery clinic, at the same institution, for an incisional biopsy and the histopathology report was given as PA. One week later the lesion was excised. Macroscopically, the excisional biopsy specimen was whitish grey in colour, soft to firm in consistency with an irregular surface texture measuring about 5×3×2 cm. The specimen was fixed in 10% buffer formalin, processed and sections of 4 micron thickness were made.

Microscopically, the section showed orthokeratinised surface epithelium with pseudoepitheliomatous hyperplasia. Underlying lesional mass was well circumscribed and was separated by thick connective tissue capsule from normal palatine salivary gland [Figure 3].

The tumor cells appeared to be spindle, epitheloid and plasmacytoid shaped [Figure 4]. The tumor cells were arranged in sheets and nests separated by thin connective tissue septae and in some places showed alveolar and whorl pattern. Areas of hyalinization were seen with few keratin pearls, along with very few ducts with mucoid secretions [Figure 5]. Presence of plenty of dilated vessels and extravasated RBCs were found throughout the sections. There was no cellular or nuclear pleomorphism, necrosis or increased mitotic figures. Areas of fibrinous exudates and necrosis were seen that corresponded to previous sites of incisional biopsy. Immunohistochemical analysis was positive for smooth muscle actin (SMA) and S-100 and negative for Ki-67 [Figure 6]. Final diagnosis of ME was given.

Discussion

ME of soft tissue when considered together, two thirds of them are reported on the extremities (lower extremity and upper

Figure 1: Intraoral picture of the lesion involving the palate

Figure 2: Computed tomography axial section (contrast) an ill-defined heterogeneously enhancing soft tissue mass with cystic area within the soft palate displacing the uvula

Figure 3: Microscopic view of encapsulated lesion with normal salivary gland tissue (H and E stain at ×10 magnification)

Figure 4: Microscopic view of plasmacytoid cells (H and E stain at ×10 magnification)
extremity); the rest one thirds involve the trunk, visceral soft tissues and head and neck region. In salivary gland, most ME affect the parotid and minor salivary glands of the palate. Majority of the cases are benign and constitute only 1/10th of salivary gland tumors.

Most of the authors consider ME as a one of the entity in the continuous histologic spectrum of salivary gland adenoma, one end being represented by canalicular adenoma where there is only luminal cell/epithelial cell proliferation with no myoepithelial component, followed by the basal cell adenoma with very few myoepithelial cells, continuing with PA where there is significant proportion of myoepithelial cells, then the myoepithelial cell rich PA and other extreme end represented by pure ME. At present the ME are considered as distinct pathological entity owing to its predominant myoepithelial cell proliferation and an aggressive biological behavior.

Most of the authors report that there is no gender predilection. The maximum incidence is noticed involving the age range of 30-50 years, if reported in children it is said to be more aggressive and malignant. ME show benign clinical course and present as painless, slowly growing, firm mass.

On gross examination, soft tissue ME is characteristically well-circumscribed except for tumors of minor salivary glands which may lack complete encapsulation. Gelatinous, myxoid areas are common and focal calcification or ossification may be seen. Necrosis is rare.

Microscopically ME is characterized by the proliferation of myoepithelial cells arranged in cords, nests or mantles. The morphology of myoepithelial cells can be diverse. Spindle, plasmacytoid, reticular, epithelioid, and clear, additionally mixed histological form are also described. Plasmacytoid cells are polygonal cells with eccentric nuclei and it has abundant eosinophilic, nongranular or hyaline cytoplasm. The epithelioid cells are round to polygonal cells with centrally located nuclei and variable amount of eosinophilic cytoplasm. Reticular pattern is hypocellular with abundant myxoid stroma present between the cells. The clear cell variant are polygonal cells with abundant and optically clear cytoplasm containing large amounts of the glycogen. The spindle cells are usually found to be arranged in interfacing fascicles. One of these cell types are present predominantly in a given ME but other variants can also be seen to some extent admixed with each other. These cells may be arranged in several architectural patterns, which include non-myxoid (solid), myxoid (PA-like), reticular (canalicular-like) and mixed. The present case showed predominantly plasmacytoid cells, along with spindle cells and epithelioid cells arranged in sheets, nests and focal areas in alveolar and whorl pattern.

The stroma of soft tissue ME is composed of fibro-hyalinised or myxoid connective tissue, similar to that seen in some PA and occasionally may show chondroid, osteoid, mature fat cells and extracellular collagen crystalloid structures. Various authors agree that such type of metaplastic changes are typical features of PA. This is one of the debatable features present in our case, where the focal areas presented, hyalinization along with few keratin pearl formation.

Besides, ductal/luminal differentiation is not normally expected in ME and when present, it constitutes <5% of the tumor parenchyma; which is considered to be the best diagnostic factor by many authors to distinguish it from PA. The present case showed similar findings.

The heterogeneous morphologic expression by myoepithelial cells and resultant overlapping of histopathologic features with other tumors necessitates an immunohistochemical analysis to confirm the diagnosis of ME in most of the cases. Immunohistochemical studies have revealed that myoepithelial neoplastic cells show variable immunoreactivity related to cell phenotype with positive immunoreactivity against S100 protein, cytokeratin, actin muscle specific, vimentin, and glial fibrillar glycoprotein. It is suggested that assessment of cell proliferative activity may be helpful in the differential diagnosis between benign and malignant ME. Ki-67 labelling index of more than 10% in ME is highly suggestive of malignant biological...
behaviour.\(^{5,8,9,12}\) The other clinical criteria for malignant ME are that the lesion will be painful, frequently exhibiting sudden and rapid growth. Histologically, the multinodular appearance of the lesion, presence of infiltrative growth, necrosis, increased and abnormal mitotic figures indicates malignancy.\(^{5,9,10,11,14}\) In the present case even though there is rapid growth reported since 3 months, which can be attributed to disturbance of the lesion caused by FNAC, none of the malignant clinical or histological criteria were met. The IHC analysis showed tumour cells positive for S100, SMA and negative for Ki67.

Peripheral nerve sheath tumors, fibrohistiocytoma, synovial sarcoma, nodular fasciitis, schwannoma, leiomyoma and hemangiopericytoma should be considered in the differential diagnosis of spindle cell variant of ME, whilst the clear cell ME should be correctly discriminated from the clear cell adenocarcinoma and mucoepidermoid carcinomas. Plasmacytoid variant must be differentiated from extra medullary plasmacytoma.\(^{1,3,4,6,9,14,15}\)

The ME have good prognosis without recurrence risk even after 10 years of surgery, provided complete surgical excision with margin of normal uninvolved tissue is done. Radiation therapy is used only in cases where surgical operation is not feasible.\(^{1,5}\) In the present case; surgical excision with periodic follow up of two year was done with no recurrence.

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References
