

Sinonasal Spindle-Cell Myoepithelioma

P Boscolo-Rizzo, M Da Mosto, C Marchiori

Citation

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Abstract

Myoepithelioma is a rare benign neoplasm of salivary gland derivation. Pure myoepithelioma accounts for less than 1% of all salivary gland tumors. Only three cases of sinonasal myoepithelioma have been reported. Diagnosis of myoepithelioma is not easy by light microscopy alone and immunocytochemistry is indispensable. The treatment of myoepithelioma should be complete surgical excision

A rare case of sinonasal spindle-cells myoepithelioma is reported in a 68-year-old woman.

CASE REPORT

An otherwise healthy 68 year-old woman presented with a 6 months history of headache, progressive right nasal obstruction accompanied by episodes of epistaxis. At admission a large polypoid tumor occupying the right nasal cavity was noted. CT and MR scan (Figure 1 and Figure 2) showed a 3.0 x 4.0-cm mass arising from the mucosa of the right ethmoid region; some areas of necrosis were present; the surrounding bony structure was intact but its growth expanded nasal septum and lamina papyracea.

An excisional biopsy was performed and a significant bleeding occurred requiring intravenous fluid and transfusion although several nasal packings were performed.

Figure 1

Figures 1 and 2: MR shows a 3.0 x 4.0-cm mass arising from the mucosa of the right ethmoid region with some areas of necrosis; the surrounding bony structure is intact but its growth expands nasal septum and lamina papyracea

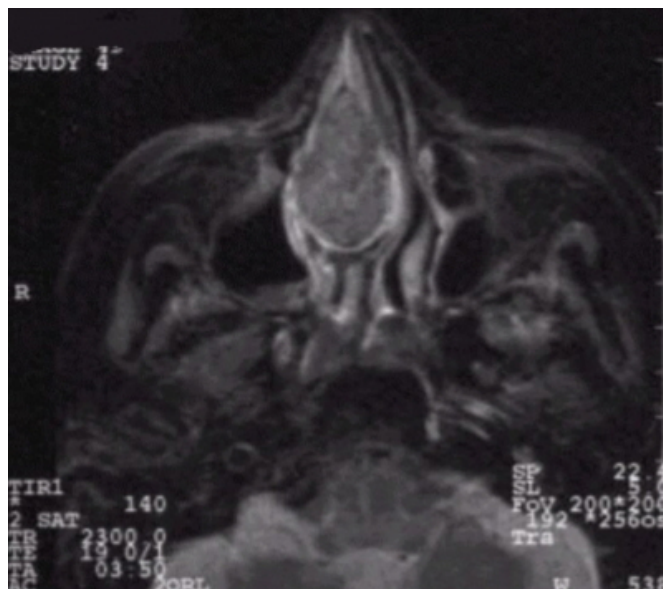
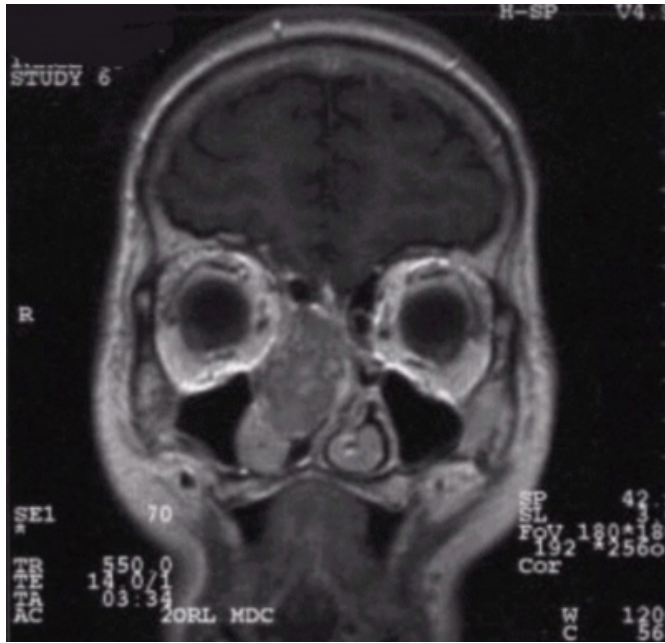


Figure 2



The surgical treatment was done via a lateral rhinotomy approach. The sessile tumor was located on the middle-upper portion of ethmoid, close to sphenopalatine artery; it appeared encapsulated, moderately firm, brown with multiple haemorrhagic areas. It was resected en-bloc with ethmoid labyrinth.

On the 2nd day an important epistaxis occurred; it was controlled and after 1 week the patient was discharged.

Histologic examination revealed a well circumscribed solid proliferation of spindle cells with formations of fascicles. Mitotic activity was absent. Immunohistochemical studies revealed diffuse positivity to cytokeratins (AE1, CAM 5.2), vimentin and S-100 protein. These histopathological features were coherent with spindle cells myoepithelioma (Figure 3 and Figure 4)

Figure 3

Figures 3 and 4: Spindle-shaped cells with marked pleomorphism showing a interlacing fascicular pattern of growth

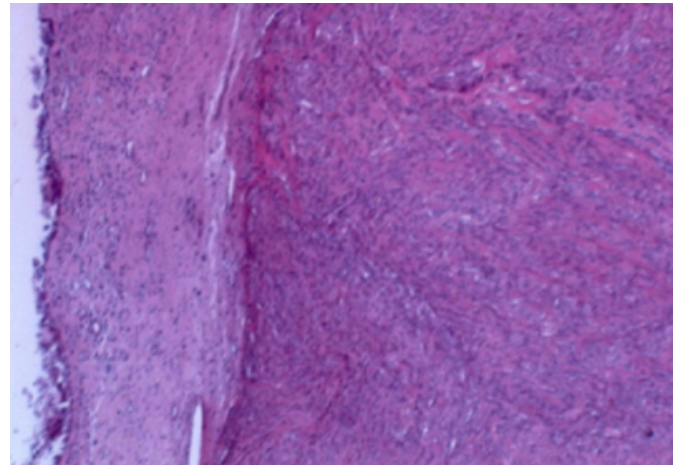
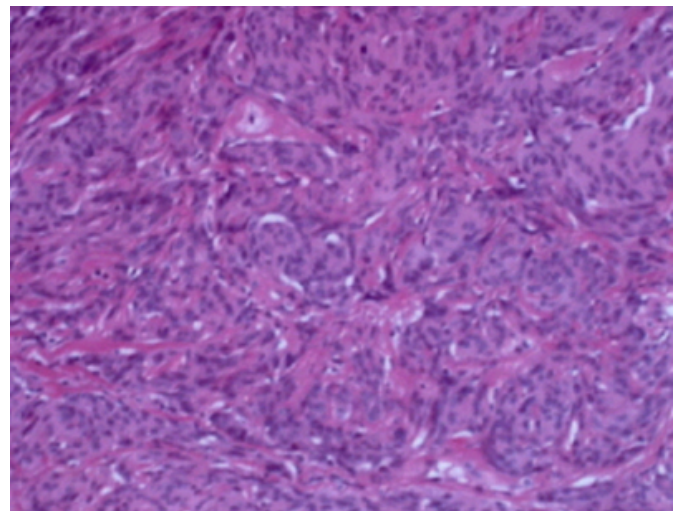


Figure 4



DISCUSSION

Myoepithelioma is a rare benign neoplasm of salivary gland derivation, defined as composed of myoepithelial cells and first described in 1943₁.

Myoepithelial cells exhibit a dual epithelial and smooth muscle phenotype, have contractile properties and are present in many secretory organs. In the salivary gland myoepithelial cells are located in the acini and intercalated ducts. These cells are an important element in many salivary gland neoplasms. However pure myoepithelioma accounts for less than 1% of all salivary gland tumors. This tumor occurs over a wide age range with a median of 53 years and affects both sexes equally. It usually presents as a painless, slow-growing mass of benign nature, but sometimes it may

be locally aggressive. The parotid gland is preferentially involved (40%); the intraoral minor salivary glands of the palate are less common sites of occurrence (21%)₂.

Only three cases of sinonasal myoepithelioma have been reported_{3,4,5}.

The imaging appearance of myoepitheliomas (a well-defined enhancing mass) is not specific₅.

Macroscopically benign myoepitheliomas are well circumscribed and encapsulated masses with a smooth external appearance and a white, tan or gray cut surface but there are no distinctive features.

Microscopically several growth patterns occur: solid, the most common, myxoid (pleomorphic adenoma-like), reticular and mixed. Cells can vary in histologic appearance being spindle-shaped, plasmocytoid, epithelioid; occasionally, epithelioid cells may have clear cytoplasm. The different cellular compositions have not correlation to prognosis₆.

Although myoepithelioma has been defined as a neoplasm exclusively composed of myoepithelial cells, many authors have introduced a less rigid definition to include tumors with a small number of ductal cells (less than 10% of surface area of the tumor). Probably myoepitheliomas constitute one end of a biologic spectrum that includes pleomorphic adenoma in the middle and non-membranous basal cell adenomas at the other end_{6,7}.

Due to their infrequency and variety of histopathologic features, diagnosis of myoepithelioma is not easy by light microscopy alone. Immunocytochemistry can aid in diagnosis. Almost all cases of myoepithelioma are strongly positive for S-100 protein and the tumor cells also display varying degrees of immunoreactivity for cytokeratin, GFAP, myosin, actin, vimentin and CEA. The most ultrastructural feature of myoepithelial cells is the presence of myofilaments they may appear in well-aligned bundles or as disordered arrays; desmosomes are commonly present₆.

The differential diagnosis of sinonasal myoepithelioma especially includes pleomorphic adenoma and mesenchymal neoplasms: nasal fibrosarcoma, fibromatosis of the sinonasal tract, neoplasms of smooth muscles, sinonasal mixoma, embryonal rhabdomyosarcoma.

Malignant myoepithelioma infiltrate the surrounding tissue and can metastasize; it can arise de novo or develop ex pleomorphic adenoma or ex benign myoepithelioma_{8,9}.

Myoepitheliomas are not more aggressive than benign mixed tumors as once thought. Instead, they are essentially the same benign tumor albeit at a different point along a histologic continuum_{6,10,11}.

The treatment of myoepithelioma should be complete surgical excision; in this case the prognosis appears to be good₁.

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Author Information

Paolo Boscolo-Rizzo, MD

ENT Department, University of Padua, Treviso Regional Hospital

Maria Cristina Da Mosto, MD

ENT Department, University of Padua, Treviso Regional Hospital

Carlo Marchiori, MD

ENT Department, University of Padua, Treviso Regional Hospital