Case Report / Olgu Sunumu

Myoepithelial neoplasm of nasal cavity: an uncommon tumor presenting with an unusual clinical presentation

Burun boşluğunun miyoepitelyal neoplazmı: Olağan dışı klinik tablo ile seyreden nadir bir tümör

Anirban Ghosh, M.D.,[1] Somnath Saha, M.D.,[2] Sudipta Pal, M.D.[2]

[1]*Department of Otolaryngology, MGM Medical College, Kishanganj, India*
[2]*Department of Otolaryngology, RG Kar Medical College, Kolkata, India*

Myoepithelial tumors are rare malignant tumors of salivary glands with uncertain biological behaviors and clinical course. Parotid gland is the common involvement site. In this article, we present a rare case of myoepithelial neoplasm of nasal cavity in an 11-year-old boy in the light of literature data.

Key Words: Immunohistochemistry; myoepithelial neoplasm; nasal cavity.

Miyoepitelyal tümörler, biyolojik davranışları ve klinik seyri kesin olarak bilinmeyen, tükürük bezinin nadir malign tümörleridir. Parotis bezi, en sık tutulum yeridir. Bu yazıda 11 yaşında bir erkek çocukta burun boşluğunda görülen nadir bir miyoepitelyal neoplazm olgusu literatür verileri eşliğinde sunuldu.

Anahtar Sözcükler: İmmünhistokimya; miyoepitelyal neoplazm; burun boşluğu.

Myoepithelial neoplasm is an uncommon, low-grade, malignant epithelial cancer composed of variable proportions of ductular cells and large, clear staining myoepithelial cells arranged around the periphery of the ducts. About 120 cases have been documented in the world literature,[1] most of which were located in salivary glands; except for a few cases occurring in such unusual locations as breast, lacrimal gland, lungs, nose and paranasal sinuses, and trachea. Here we describe a rare case of myoepithelial neoplasm of the nasal cavity.

CASE REPORT

An 11-year-old boy presented with a history of recurrent epistaxis and left nasal blockage over the last three years. On anterior rhinoscopy a polypoidal reddish mass covered with slough was noted in the left nasal cavity (Figure 1). Probing suggested that the mass arose from the medial nasal wall and bled on touch. Posterior rhinoscopy also revealed the same mass obscuring the posterior nasal space. Computed tomography (CT) scan revealed a homogenous

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Correspondence / İletişim adresi: Anirban Ghosh, M.D. Rambagan, Po- Searsol-Rajbari, Dt- Burdwan, West Bengal, India, Pin-713358 Raniganj, India.
Tel: +919 836 469 324 e-mail *(e-posta):* way2anirban@gmail.com

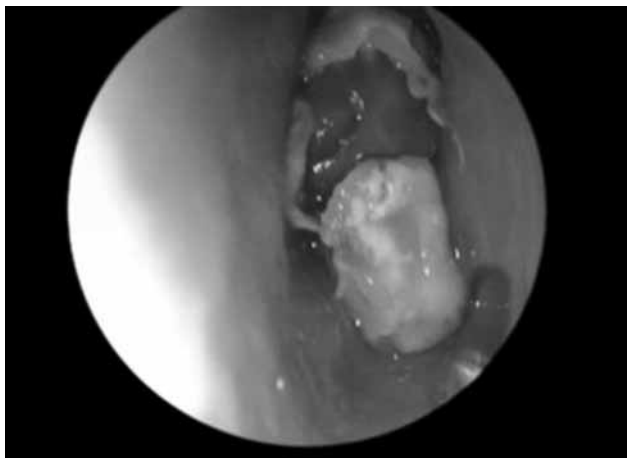

Figure 1. *Endoscopic photograph showing left sided polypiodal mass covered with slough.*

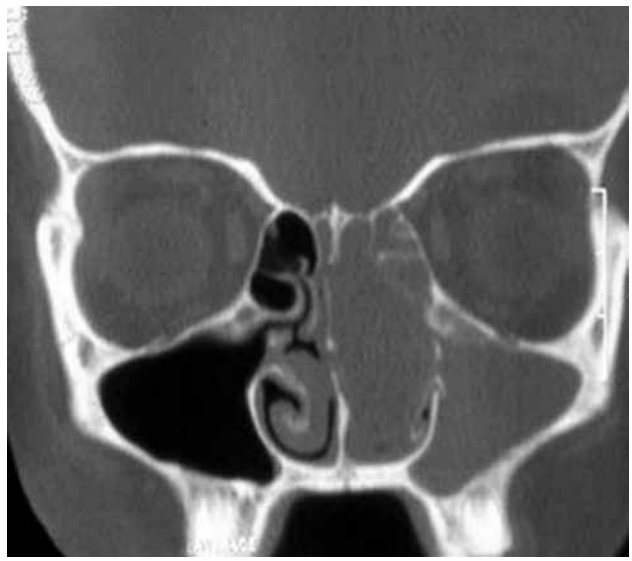

Figure 2. *Coronal computed tomography scan showing homogenous mass in nasal cavity and maxillary sinus without bony destruction.*

mass in the nasal cavity with an homogenous opacity in the ipsilateral maxillary sinus with no significant dilatation of the maxillary ostium or bony destruction (Figure 2). Endoscopic sinus surgery with microdebrider was performed. A stalk was found attached to the medial lamella of the left side. Middle meatal antrostomy was done only to yield retained mucosal secretions. Proper hemostasis was achieved. Histological examination revealed that the tumor consisted of solid proliferation of clear cells and in some places elongated duct structures. The arrangement was in double layer with inner cuboidal cell-layer and outer clear cell layer. Dual differentiation towards myoepithelial and ductal cells was confirmed immunohistochemically. This tumor was positive for cytokeratin (Figure 3), epithelial membrane antigen (EMA) (focal), P63 and cytokeratin (CK)-5/6 and immunonegative for synaptophysin, chromogranin A and calponin.

The patient was under strict endoscopic follow-up for seven months showing no recurrence (Figure 4).

DISCUSSION

Myoepithelial carcinoma is a rare tumor of salivary glands with an incidence of <1%. Myoepithelial tumors of soft tissue affect any age group

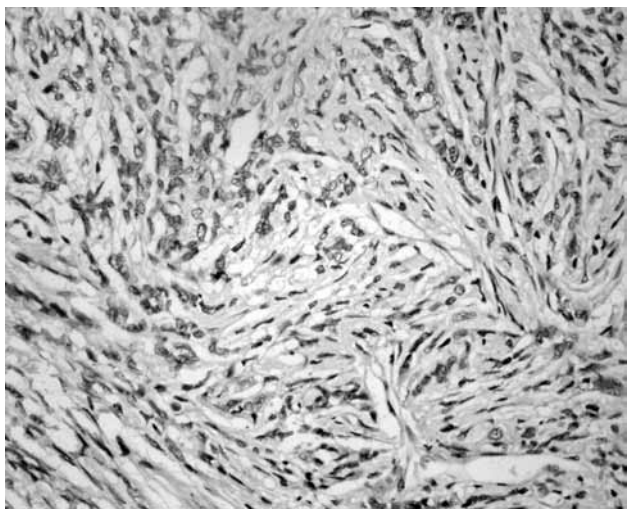

Figure 3. *Microphotograph showing positive immunohistochemical staining for Pan-cytokeratin (Pan-CK x 400).*

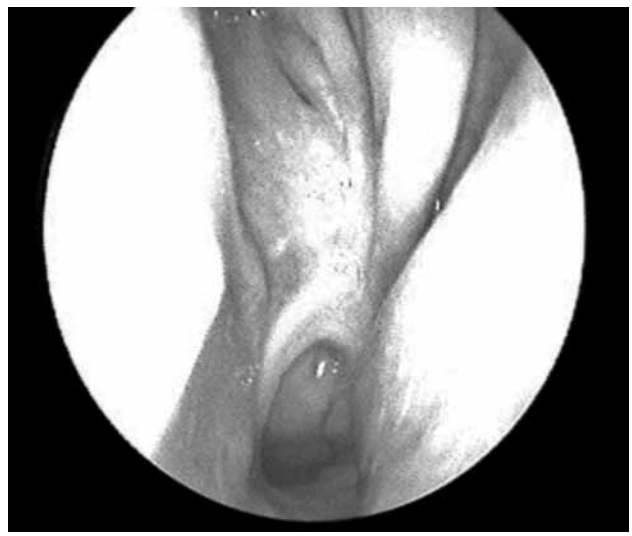

Figure 4. *Endoscopic picture showing no residual or recurrence of the tumor (three months after the surgery).*

(range, 3 to 83 years) with equal sex predilection. These carcinomas are disproportionately common in the pediatric age group (as in this case) and have an aggressive clinical course.[2] This is usually a tumor of parotid gland;[3] other sites are lung, bronchi,[4] breast, lacrimal gland, nose and paranasal sinuses.[1] The small number of reported cases of myoepithelial neoplasm in the nasal cavity and paranasal sinus makes the diagnosis and management more difficult. There are only five cases of histologically proven myoepithelial carcinoma of the nose and paranasal sinuses.[1,5-8] This, to the best of our knowledge, is the sixth case in the English literature.

As this tumor is slow growing, most patients present with long standing benign symptoms like nasal blockage and epistaxis for few months to years. In this case, the patient presented with nasal obstruction and few episodes of epistaxis for three years.

On CT scan, this neoplasm appears isodense to muscle and demonstrates moderate homogenous enhancement with contrast. On magnetic resonance imaging (MRI), it is hypointense on T_1 and displays dense, homogenous enhancement.[9]

The treatment for epithelial-myoepithelial neoplasm is complete excision.[10] Histologically this tumor is well circumscribed, so complete excision is possible. Sometimes incomplete excision leads to recurrence, which proves the aggressive biological behavior of this tumor.[9] This neoplasm is not radio or chemosensitive.

Immunohistochemistry and ultrastructural study have added important objective component to the diagnosis and classification of this tumor. Immunohistochemically, myoepithelial cells express S100 protein, smooth muscle myosin, calponin, high molecular weight CK-5/6.[10] In this case, the tumor was positive for CK-5/6, P63, CKs and immunonegative for calponin, chromogranin and synaptophysin. However immunoreactivity for S100 protein may be lacking possibly because of high differentiation. Forty-five percent of myoepithelial cells are strongly immunopositive for P63 gene product. P63 is also found in basal cells of respiratory epithelium and mucous glands. Other immunological markers that are positive for myoepithelial neoplasm are cytokeratin 7.14; vimentin, fibrillary acidic protein. Cellular localization of S100 protein allows identification of myoepithelial cells and helps in the diagnosis of this tumor. Histologically this tumor closely resembles malignant melanoma and malignant peripheral nerve sheath tumor. Malignant melanoma is positive for melanosome mouse monoclonal antibody (clone HMB-45).

Though recurrence is rare in case of benign myoepithelial neoplasm, 42% recurrence has been documented in its malignant counterpart with 32% metastases.[9] Recurrence and metastases are common in children with negative tumor margins.[2] In published cases, there are no recurrences or metastases noted in nasal and paranasal sinus sites.[6]

In conclusion, myoepithelial neoplasm is a rare low-grade neoplasm of parotid and submandibular glands. Myoepithelial tumors arising from the nasal cavity may originate from minor salivary glands present in the nasal cavity and paranasal sinus mucosa. Myoepithelial neoplasm is a challenging diagnosis. The different differential diagnosis includes pleomorphic adenoma, low-grade adenocarcinoma, and adenoid cystic carcinoma. Dual differentiation toward myoepithelial and ductal cells is confirmed by immunohistochemistry. Recurrence and metastases rates varied from 35% to 50% and 8.1% to 25% respectively in different reports. The present case had neither recurrence nor metastases seven months after surgery.

Declaration of conflicting interests

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