Journal of Current Research in Otolaryngology
ISSN: 3065-6974

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Case Report *Volume 6 Issue 1*

Myoepithelial Carcinoma of the Nasal Septum- Case Report of a Rare Entity

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Received Date: December 27, 2024; **Published Date:** February 10, 2025

Abstract

Myoepithelial carcinoma are malignant tumours primarily arising in the salivary gland; however, these tumours can arise in the nasal cavity rarely. We report a case of 56-year-old male clinically presenting as polypoidal growth in nasal septum. A whole-body PET scan showed an FDG avid enhancing soft tissue density lesion in left nasal cavity. Histopathology examination showed CK 7, SOX-10, S-100 protein and SMA positivity, eventually, diagnosed with nasal septum myoepithelial carcinoma.

Keywords: Myoepithelial Carcinoma; Nasal Cavity

Abbreviations

IHC: Immunohistochemistry; SMA: Smooth Muscle Actin.

Introduction

Myoepithelial neoplasms first described in 1898 by Zimmerman are rare tumours of the salivary glands composed almost exclusively of cells with myoepithelial differentiation showing infiltrative growth with potential for metastasis [1]. Morphologically these tumours are defined by their cytological differentiation (basaloid, clear cell, plasmacytoid), extracellular matrix production and architectural patterns (myxoid, solid, microcystic, reticular) of neoplastic myoepithelial cells. Such complex morphology poses diagnostic difficulties and use of immunohistochemistry (IHC) can be and effective approach to identifying these tumours [2]. Identification of myoepithelial cells includes specific immunohistochemistry like vimentin, calponin, S100, p63, glial fibrillary acidic protein, smooth muscle actin (SMA), and smooth muscle heavy chains [3]. These tumours are mostly seen in the major salivary glands which include parotid gland (50%), sublingual gland (33%) and submandibular gland (13%). The other common sites include nasopharynx, larynx, lung, retroperitoneum, skin, and soft tissue [4]. We present a case of myoepithelial carcinoma of nasal cavity presenting as a nasal polyp.

Case Report

A 56-year-old male patient presented with history of bleeding from nasal cavity for 8 months associated with nasal obstruction. On examination a pink polypoidal mass filling entire left nasal cavity with mucopurulent discharge and clots was seen. Computed tomography showed a 2 X 1.1 X 2.1 cm well defined heterogeneously and mildly

enhancing homogenous soft tissue density in the region of nasal vestibule with deviated nasal septum to right. Minimal mucosal thickening was seen in the bilateral inferior turbinates, ethmoid and maxillary sinuses. Few discrete bilateral lymph nodes were seen in the level IB, II and III neck lymph nodes. A whole-body PET scan using 10 mCi of 18F FDG intravenously showed an FDG avid enhancing soft tissue density lesion in left nasal cavity measuring 16x10mm (SUV max 6.6). No other evidence of metabolically active disease in present whole-body scan (Figure 1).

Informed consent was taken, and patient underwent endoscopic craniofacial resection (wide excision of the left anterior nasal septal lesion with septal cartilage excision) and growth attached to the anterior septum in the left nasal cavity was excised and sent for histopathology. The cut section shows a polypoidal grey-white lesion. The haematoxylin and eosin sections show polyp lined by mucosa with submucosa showing an invasive neoplasm arranged as multiple nodules separated by fibrohyaline septa. The tumours cells were round with moderate cytoplasm, vesicular nucleus and inconspicuous nucleoli showing brisk mitosis (Figure 2). The immunohistochemical profile showed positivity for epithelial marker cytokeratin 7 and myoepithelial markers such as SOX10, S100 and SMA (Figure 3). The immunohistochemistry antibodies were obtained from Cell Marque™, Germany; BioGenex, USA or BioCare Medical, USA and were performed using Ventana BenchMark XT, Ventana Medical Systems, Hoffmann-La Roche Ltd, Tucson, AZ, USA, or BOND-III, Leica biosystem.

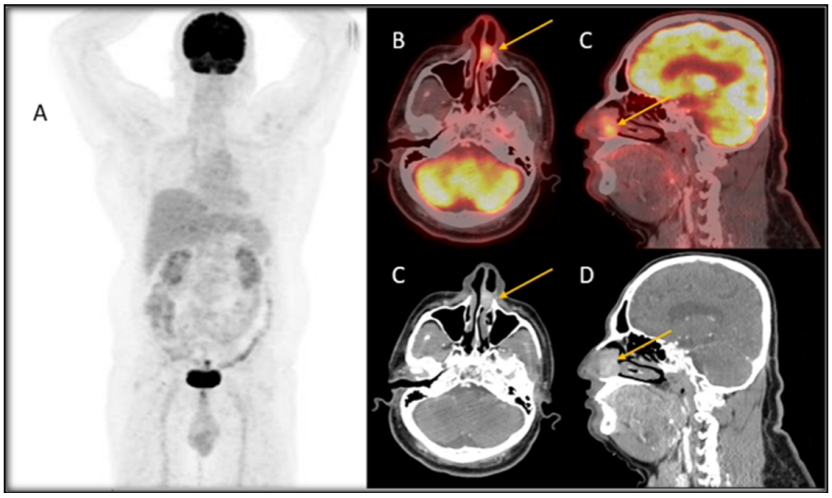

Figure 1: F-18 FDG PET/CT maximum intensity projection (MIP) (A) and axial, sagittal CT (C,D), fused PET/CT images(D,E) of the patient showing FDG avid soft tissue density lesion in left nasal cavity.

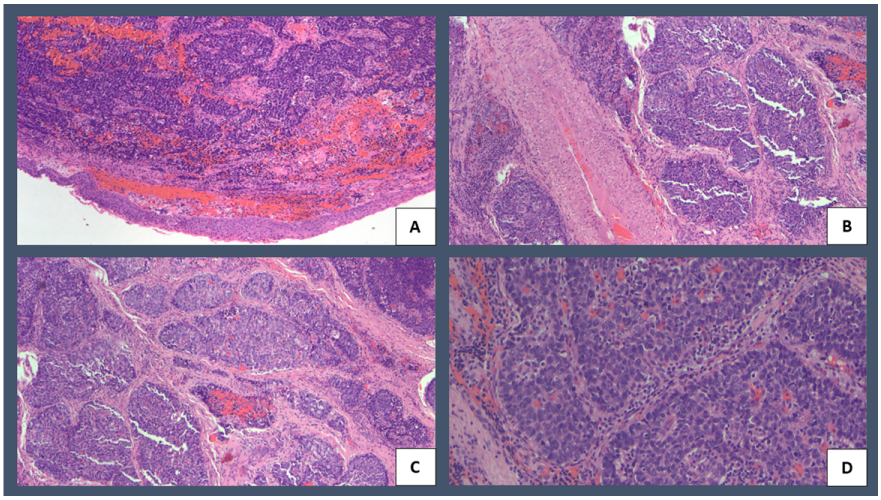

Figure 2: Photomicrographs of the tumor a), b), c) Haematoxylin and eosin showing mucosa with tumour showing multinodular architecture divided by fibrous bands. d) The 100X H&E images showing tumor cells with round vesicular nucleus.

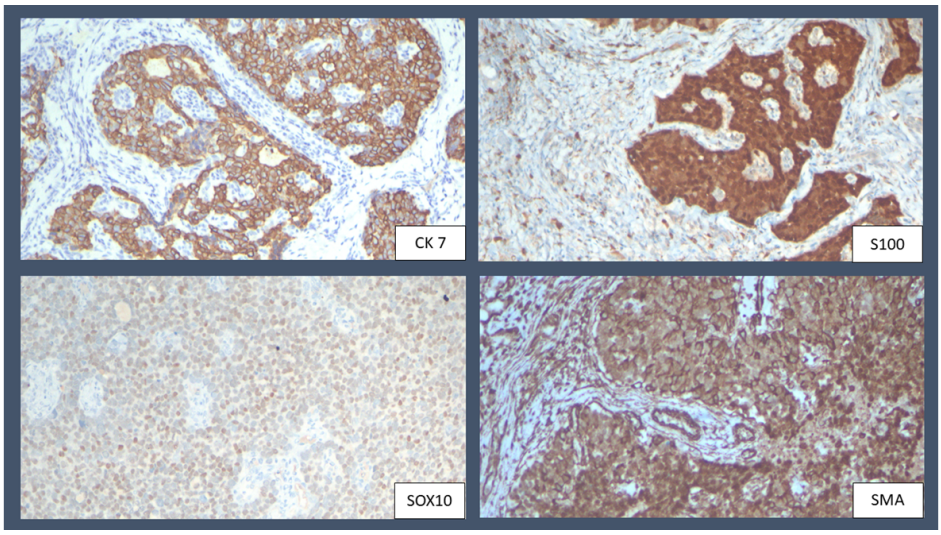

Figure 3: Immunohistochemical staining, 100x for CK7, S100, SOX10, SMA shows positivity.

The sinonasal region has been the anatomical site for origin for tumours with diverse histological features. IHC was also performed to rule out squamous cell carcinoma, spindle cell neoplasm, sinonasal undifferentiated tumours and SMARCB1-deficient sinonasal carcinoma. A negative immunostaining for p40, desmin and a retained integrase interactor 1 ruled out the above differentials (Figure 4). With the above IHC profile a diagnosis of myoepithelial carcinoma was confirmed.

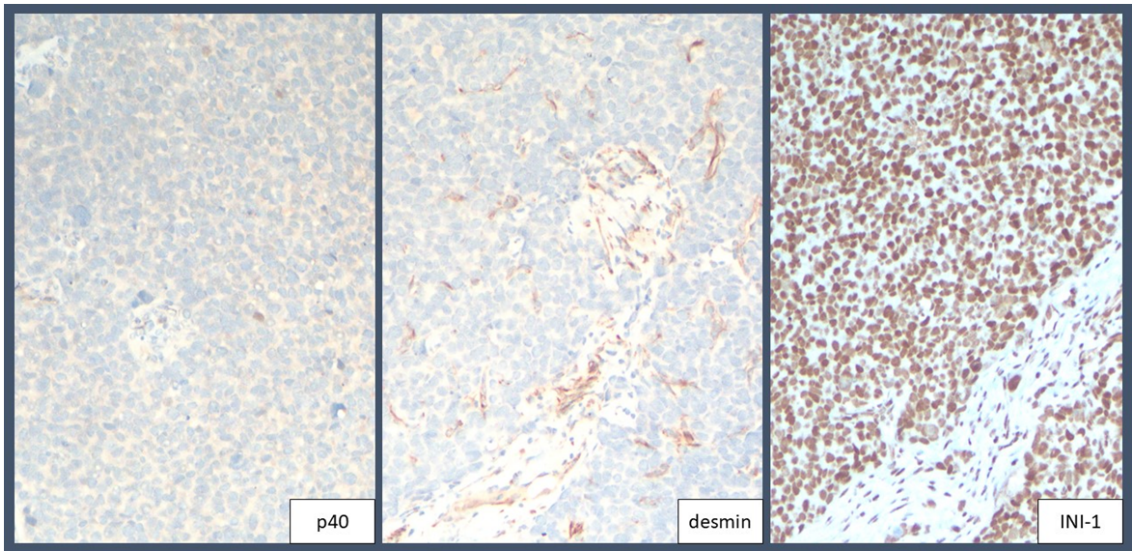

Figure 4: Immunohistochemical staining, 100x for p40, desmin, INI-1 shows negative staining for p40 and desmin while a retained INI-1.

Discussion

Nasal cavity and paranasal sinus tumours are very infrequent, accounting for less than 1% of all head and neck tumours [5]. We report an extremely rare tumor namely myoepithelial carcinoma of nasal cavity having aggressive clinical course with risks of local recurrence and distant metastasis, necessitating an accurate diagnosis for proper management. These tumours have been included in the World Health Organization classification since 1991 Morphologically diverse these tumours are characterized by a wide range of architectural and cytologic features [6]. They show multinodular or lobulated appearance variable growth patterns having solid sheet-like growths of tumor cells, with myxoid or collagenous, hyaline background [7]. Our case showed similar morphological features with tumor

cells arranged in sheets and multiple nodules separated by fibrohyaline septa.

Recognition of myoepithelial cell differentiation is not easy on routine H&E-stained sections and therefore often correct diagnosis requires IHC. This tumor typically shows co-expression of epithelial markers and S-100 which is frequently positive (72–100%) while SOX10 staining occurs less frequently in up to 30%. Myogenic markers show more variable staining and are overall of limited diagnostic value; calponin is most frequently positive (90%), followed by SMA (34–64%) and desmin (0–20%) [6,8]. In a study on 15 patients by Telugu, et al. the tumor cells were immunopositive for vimentin (100%), S-100 protein (100%), cytokeratin 7 (21%) and SMA (50%) [1]. The mainstay of management of localised soft tissue myoepithelial carcinoma is surgical resection with clear margins [9]. In our case all the resected margins re free of tumor. In the patients with extensive dissemination palliative surgery, adjuvant radiotherapy or chemotherapy might be beneficial.

Conclusion

In conclusion, myoepithelial tumours are rare tumours with aggressive behaviour and varied morphological patterns. Accurate diagnosis often requires comprehensive IHC panels to identify myoepithelial differentiation and ruling out other differentials. Early diagnosis and treatment can significantly reduce the rate of recurrence and metastasis thus improving the survival and prognosis of patients.

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