CASE STUDY

Myoepithelioma of the Nose

Mioepithelioma nasal

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A 64-year old female, with no other history of interest, presented with anosmia and nasal obstruction which had evolved over several months. Diplopia had also appeared during the last month.

On physical examination left-sided exophthalmos was observed. An endoscopy disclosed a tumour which completely obstructed the left nasal fossa. A biopsy of the lesion was performed, which caused moderate epistaxis that required anterior packing.

The CT scan showed a large, extensive tumour in the left nasal fossa, spreading into the nasopharynx, which had reshaped the adjacent bone structures and caused ipsilateral pansinusobital obstruction (Fig. 1A). The MRI disclosed a solid tumour, with internal cystic areas, well-defined contours, heterogeneous in appearance and with no signs of intracranial invasion (Fig. 1B).

Biopsy findings were of myoepithelioma with fusocellular components of salivary gland type. The non-aggressive nature of the tumour facilitated endoscopic resection and there were no complications.

Histologically, the tumour specimen confirmed the presence of mixed stroma myoepithelial cells (Fig. 2A and B), tested positive for cytokeratin, vimentin and S-100 protein (Fig. 2C and D). Since the patient’s progress was good, it was decided to refer check-ups to the outpatient department.

Discussion

Nasal cavity and paranasal sinus tumours are very infrequent, accounting for under 1% of head and neck tumours.1 Nasal mucosa contains mucous glands, minor salivary glands, melanocytes and olfactory neuroepithelial cells.2 Myoepithelioma is a benign slow-growing tumour of the salivary glands described by Sheldon in 1943.3

Its location in the nasal cavity means that it is asymptomatic up to advanced stages, with the result that when it does exist and is diagnosed, it has spread locally and surgery may sometimes be complicated due to the proximity of vital structures such as the skull base, brain, eye sockets, and carotid artery.

Presentation symptoms are nasal obstruction, epistaxis, pain and episodes of sinusitis.2

Macroscopically they are encapsulated tumours with the appearance of normal nasal mucosa. Microscopically the tumours are composed of myoepithelial cells (fusocellular components, plasmacytoid like cells, clear cells and epithelioids) in different histological patterns (solid, mixed or reticular).4,5

In the 1940s it was believed to be a variant of pleomorphic adenoma, with similar components and occasional tendency towards malignancy, but it was acknowledged as an individual disease in recent WHO classification.6 One difference with adenomas is that myoepithelioma may synthesize extracellular components such as proteoglycans, basal lamina, collagen and elastic fibres.7

Malignant myoepithelioma must also be differentiated. This is a medium to high grade malignant tumour which
**Figure 1**  (A) CT imaging in axial and coronal planes divided into soft tissue and bone, which show an expansive left nostril lesion. (B) Enlarged resonance imaging in T2 in sagittal and axial planes, and T1 with gadolinium in coronal plane.

**Figure 2**  Myoepithelioma cells, fusocellular in appearance, (A) haematoxylin-eosin at 100×; (B) haematoxylin-eosin at 400×; (C) positive cytokeratin, (D) positive vimentin.
in 65% of cases is located in the parotid gland, where myoepithelial cells with atypical cells and neural infiltrating of nearby structures are observed.\(^5\)

Finally, a differential diagnosis with other soft tissue tumours is recommended, such as with leiomyomas, leiomyosarcomas, nerve tissue tumours and synovial sarcoma.\(^3\) Immunohistochemical staining is conclusive here, as it tests positive for cytokeratins, actins, vimentin and S-100 proteins.\(^3,5\)

Despite its most frequent location in the salivary glands, other infrequent locations have been described, such as the outer ear canal, the eye socket, and the nasal cavity. Only 4 cases to date have been published in the latter location.\(^7\)

Subsequent to anatomopathological confirmation from biopsy findings, we decided to perform en bloc endoscopic resection as the tumour was benign. Due to the scarcity of described cases no suitable surgical procedure has yet been established for this condition. In several publications it was treated as a stage T1 on the Krause scale for inverted papilloma.\(^3\) One case was resolved with partial maxillectomy via a lateral rhinotomy approach, on suspected malignancy, and another case via a sublabial approach due to large size of the tumour.\(^9,10\)

Of the 5 published nasal myoepithelioma cases, 3 are female and 2 male, with a mean age of 66 at diagnosis (age range: 64–72), 3 were of Asian origin and one American. Ours was the only European case published to date. The symptom common was nasal obstruction, with cases of recidivant epistaxis and sinusitis. In 2 cases the origin of the tumour identified during surgery was the root of the middle nasal concha. In another case it was the ethmoids/superior wall, and in another the anterior septum. In our case the origin of the tumour appeared to stem from the posterior segment septal mucosa.

Myoepitheliomas have a tendency to recur, and for this reason access to them should be made through the route where total resection can be ensured. Furthermore, there is the probability of malignant degeneration of the tumour, which increases over time with and with plasmocytoid variants, where cases of salivary gland malignant myoepithelioma have been described.\(^3\)

To conclude, we would like to reiterate that to date there have only been 5 cases published on myoepithelioma of the nose. Literature is even scarce on its more frequent location in the parotid gland. We therefore consider long-term follow-up as necessary in order that a more precise prediction as to its evolution and prognosis may be made.

References