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CASE REPORT

Pleomorphic adenoma of the nasal septum



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KEYWORDS

Pleomorphic adenoma;
Nasal cavity;
Nasal septum;
Salivary glands;
Benign tumor

Summary

Introduction: Pleomorphic adenoma is the most common benign tumour of the salivary glands. The major salivary glands are most commonly involved, or more rarely accessory salivary glands, especially the oral cavity. Other locations, such as the nasal cavity, paranasal sinuses or upper aerodigestive tract, are exceptional.

Case report: A 26-year-old female presented with right-sided nasal obstruction. Radiology found a lesion involving the anterior third of the nasal septum. The patient underwent complete surgical resection of the tumour via an endonasal approach. Histological examination found a mixed cellular component (epithelial and myoepithelial) and mesenchymatous tissue with chondromyxoid stroma, enabling diagnosis of a typical pleomorphic adenoma.

Discussion/Conclusion: Pleomorphic adenoma is exceptional in the nasal cavity, with only a few cases reported in the literature. Although benign, the risk of local recurrence, malignant transformation and metastasis requires close long-term follow-up.

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Introduction

Pleomorphic adenoma (PA) is the most frequent benign tumour of the salivary glands. Sixty-five percent of cases involve the major salivary glands (MSG), mainly the parotid, and 35% the accessory salivary glands (ASG), mainly the oral cavity and especially the palate [1]. The nasal cavities, sinuses and upper aerodigestive tract are only exceptionally involved.

PA is a mixed (two-component) tumour: a cellular component comprising epithelial and myoepithelial cells, and mesenchymatous stroma (myxoid, hyaline, chondroid

and osteoid). Cross-sectionally, the aspect is of a well-circumscribed, round or lobular nodular tumour surrounded by a fibrous capsule of varying thickness, of a firm consistency or in some cases, soft and myxoid (gelatinous), and of homogeneous whitish hue.

We present a rare case of PA of the nasal septum.

Case report

A 26-year-old woman presented in January 2012 with unilateral right-sided nasal obstruction of six months' evolution.

Rhinoscopy found a mass of chondroid consistency entirely filling the right nasal cavity, pedunculated for about 1 cm at the septum.

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Figure 1 MRI finds a circumscribed tumor in the nasal septum, showing in high-intensity signal on T2-weighted sequences.

MRI found a well-circumscribed polypoid formation involving the anterior part of the nasal septum, showing in low-intensity signal on T1 and high-intensity signal on T2-weighted sequences (Fig. 1), heterogeneous, with generalized enhancement on gadolinium injection.

The tumour was completely resected, including a piece of cartilage, with associated septoplasty through an endonasal approach. Postoperative course was simple.

Pathologic examination found a well-circumscribed, homogeneous, firm $2.5 \times 1.2 \times 1$ cm grayish white tumour.

Histologic examination revealed two components: epithelial and mesenchymatous. The former comprised of basaloid epithelial and spindle-shaped myoepithelial cells in trabeculae, tubules and cribriform structures; the latter formed large chondroid lobules with spaces occupied by regular chondrocytes and loose myxoid zones. The two components were of varying proportion depending on the area (pleomorphic aspect): in some places, the cellular component predominated, without mitosis, atypia or necrotic sites. The tumour was well-circumscribed by a regular, non-invaded fibrous capsule, without perineural invasion (Fig. 2).

Histologic analysis thus diagnosed typical PA.

Simple surveillance was decided on, facilitated by the anterior location of the tumour. Over 10 months' follow-up, there were no signs of local recurrence.

Discussion

Pleomorphic adenoma of the nasal cavities is rare, although two large series have been reported: by Compagno and Wong, with 40 cases [2], and by Suzuki et al., with 41 cases

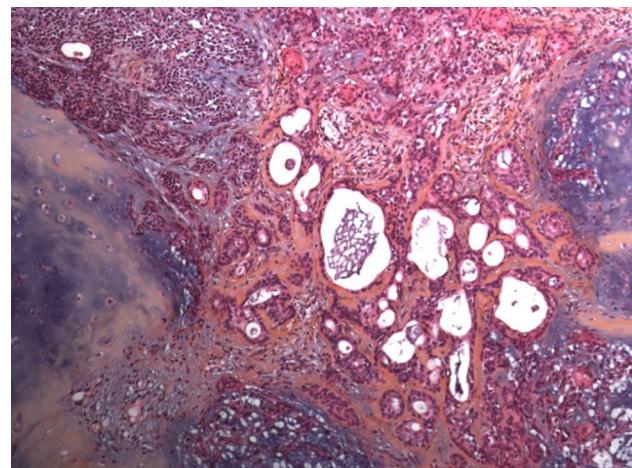


Figure 2 Pleomorphic adenoma with mesenchymatous component: chondroid (right), myxoid (below) and epithelial (center) zones forming tubules and trabeculae of cubic, basal or myoepithelial cells (HES staining).

[3]; incidence was predominantly female and in the 30–50 year age range [2].

PA grows silently, over several years, in the MSGs, but is often found early in the ASGs. The most common clinical symptoms are unilateral nasal obstruction (71% of cases), epistaxis (56%) and external deviation of the nasal pyramid [4].

The main risks are local recurrence [5], malignant transformation [6] and metastasis. Recurrence rates, after surgical resection, range from 0 to 8%, with multiple recurrence increasing the risk of cancerization [7]. The risk of malignant transformation is 6% [1] and is estimated at 1.5% within 5 years in the absence of resection [5].

For nasal PA, Compagno and Wong reported three cases of local recurrence among 40 patients (7.5%) at 3 years' follow-up [2].

Risk factors for recurrence are predominantly myxoid stroma, and an irregular or invaded capsule and multinodularity.

Many histological types of carcinoma are observed: usually undifferentiated carcinoma or adenocarcinoma, or less frequently squamous-cell carcinoma, cystic adenoid carcinoma or mucoepidermoid carcinoma.

Carcinoma included in an adenoma, said to be "non-invasive or enclosed" or "minimally invasive" (< 1.5 mm), is of very favourable prognosis. When, in contrast, the carcinoma greatly exceeds the adenoma (> 1.5 mm), becoming "invasive", prognosis is unfavourable, with 30% 5-year survival [6,8].

Treatment in "carcinoma on pleomorphic adenoma" is obviously not the same as in "simple" pleomorphic adenoma, and the two have to be distinguished if prognosis is to be assessed, especially as clinical presentation may be similar. Histology is the gold standard for differentiating between them and thereby guiding treatment.

The mechanisms of malignant transformation, however, remain poorly understood, and there is at present no consensus as to predictive histological criteria, although immunohistochemistry is beginning to shed some light.

Expression of the immunohistochemical marker MUC1 may be predictive of recurrence of salivary gland PA [9].

Röijer et al. suggested that amplification and overexpression of HMGIC and possibly MDM2 may be genetic factors for malignant transformation of PA [10].

Conclusion

Pleomorphic adenoma is not anodyne: it can affect locations other than the salivary glands and has a potential for malignant transformation that should not be underestimated. Prolonged follow-up is therefore mandatory.

Disclosure of interest

The authors declare that they have no conflicts of interest concerning this article.

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