CASE STUDY

Sclerosing Polycystic Adenosis of the Submandibular Gland

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Abstract Sclerosing polycystic adenosis (SPA) is a rare, newly reported lesion of the salivary glands. Most cases occurred within major salivary glands and were frequently confused with other salivary gland carcinomas such as acinar cell carcinoma. We report a case of sclerosing polycystic adenosis within the submandibular gland in a 63-year-old woman.

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PALABRAS CLAVE
Adenosis poliquística esclerosante; Glándula submaxilar; Tumores glándulas salivales

Adenosis poliquística esclerosante de la glándula submaxilar

Resumen La Adenosis Poliquística Esclerosante (APE) es una entidad poco frecuente descrita hace poco tiempo que afecta a las glándulas salivales, principalmente mayores, y que frecuentemente se ha confundido con otros carcinomas de glándula salival como el carcinoma de células acinares. Presentamos un caso de adenosis poliquística esclerosante en la glándula submaxilar de una mujer de 63 años.

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Introduction

Sclerosing polycystic adenosis (SPA) is a rare entity which was first described in 1996. Its anatomicopathological appearance is similar to that of fibrocystic breast disease. A total of 51 cases have been described in the literature. Its aetiology is unknown, although it is clonal and, therefore, neoplastic nature has been reported. Most cases appear in the major salivary glands, especially in the parotid gland.

It is important to carry out the differential diagnosis with other salivary gland tumours, mainly acinar cell carcinoma.

Case Report

We present the case of a 63-year-old female who attended consultation due to a left cervical tumour of a few months evolution. The patient was a smoker.

Examination revealed a normal oropharynx, with reddening of the drainage orifice of the left Wharton duct. Fibroscopy was normal, whilst bimanual and cervical palpation noticed an enlarged, left submandibular gland, with a
Sclerosing Polycystic Adenosis of the Submandibular Gland

Figure 1  Sclerosing polycystic adenosis (x2) (H-E staining). Circumscribed, partially encapsulated lesion consisting of a proliferation of ducts and acini, with a lobar pattern and without atypia. By comparison, the adjacent gland is not affected and presents no fibrosis, inflammatory infiltrates or atrophy.

posterior adenopathy to a mobile and rolling gland of about 2 cm in diameter.

A cervical computed tomography (CT) scan revealed a solid nodule of 16 mm in the left submandibular gland, with an adenopathy of 6 mm adjacent to the submaxillary gland.

Ultrasound-guided fine needle aspiration reported a well-differentiated acinar cell carcinoma (low grade).

Given these findings, we performed a left submaxillectomy with neck dissection of group I.

The pathological anatomy of the surgical specimen reported an irregular, whitish tumour of firm consistency, measuring 1.7 cm in its greatest dimension, occupying 30% of the gland, without extension into the rest of the gland and without vascular or perineural invasion.

Microscopically, we observed dilated ducts with intraluminal, epithelial hyperplasia and cellular atypia, surrounded by a stroma with abundant sclerosis. The lymph nodes were negative for malignancy. The final diagnosis was SPA (Figs. 1 and 2).

Two years after surgery, the patient remains asymptomatic and has not suffered recurrence or distant metastasis.

Discussion

SPA was first described in 1996 by Smith et al.1 At present, there are about 50 cases reported in the literature.

It mainly affects the major salivary glands, especially the parotid gland, although it may exceptionally affect the minor salivary glands.2,3 It appears in middle-aged subjects (40–50 years) with no clear gender prevalence. The tumours measure between 0.5 and 7 cm in diameter.

It is characterised by a slow growth, which is usually asymptomatic, nodular and similar in many respects to fibrocystic breast disease.4

The pathological anatomy reveals circumscribed, but not encapsulated nodules, with dilated ducts and epithelial hyperplasia, surrounded by a very fibrous stroma and with chronic inflammation.5

The aetiology of this condition is currently unknown, although some recent articles have reported conducting trials with human androgen-receptor gene (HUMARA),6 and demonstrated its clonal and, therefore, neoplastic nature. Furthermore, immunohistochemistry techniques have detected positivity for oestrogen and progesterone receptors which may play a role in its pathogenesis. Moreover, some studies have pointed to the role of Epstein–Barr virus (EBV) in SPA.7

Its treatment is surgery with disease-free margins, given that 30% of cases have reported recurrences. There have been no reports of distant metastases or mortality in this condition.

In many cases, fine-needle aspiration (FNA) techniques8 may confuse the tumours with other types of salivary gland tumours, such as acinar cell carcinoma or mucoepidermoid carcinoma. Therefore, we must always consider this entity when conducting the differential diagnosis, since, while still remaining a neoplasm, treatment and prognosis of this disease are different from those of other malignant tumours of the salivary glands.

Conflict of Interests

The authors have no conflict of interests to declare.

References