Pathology – Research and Practice 210 (2014) 342–345

Contents lists available at ScienceDirect

Pathology – Research and Practice

journal homepage: www.elsevier.com/locate/prp

Original Article

Sclerosing polycystic adenosis of parotid gland: A unique report of two cases occurring in two sisters☆

Spomenka Manojlović[a,*], Mišo Virag[b], Aleksandar Milenović[b], Luka Manojlović[c], Zrinko Šalek[c], Alena Skálová[d]

[a] University of Zagreb School of Medicine, Department of Pathology, University Hospital Dubrava, Zagreb, Croatia
[b] University of Zagreb School of Medicine, Department of Maxillofacial Surgery, University Hospital Dubrava, Zagreb, Croatia
[c] University of Zagreb School of Medicine, Croatia
[d] Department of Pathology, Charles University in Prague, Faculty of Medicine in Plzen, Plzen, Czech Republic

ARTICLE INFO

Article history:
Received 3 October 2013
Received in revised form
18 December 2013
Accepted 13 January 2014

Keywords:
Sclerosing polycystic adenosis
Salivary gland
Familial occurrence
Recurrence

ABSTRACT

Sclerosing polycystic adenosis (SPA) of salivary glands is a tumorous lesion of salivary glands, with clinical presentation of a slow-growing mass characterized by a combination of histological features, some of which are reminiscent of mammary fibrocystic disease. SPA is mostly unifocal, but rarely may be multifocal and/or bilateral. Recurrences have been reported in up to 19% of cases. Although originally considered pseudoneoplastic, the occurrence of "dysplasia" and carcinoma in situ of ductal epithelium, and recent evidence of clonality suggest a possible neoplastic nature. Herein we describe, for the first time, two cases of SPA in two sisters (7 and 33 years old). The younger patient experienced multiple recurrences. This is the first report of familial occurrence of SPA, suggesting a possible genetic background.

Introduction

Sclerosing polycystic adenosis (SPA) is a rare salivary gland lesion, first described in 1996 by Smith et al. [24]. Since then, about 60 cases have been published mostly in small series or as case reports. Most reported cases have arisen in the major salivary glands, particularly in parotid gland [2,4,5,7,9,11,14,18,21,22]. Only rare cases have been reported in the submandibular gland [8,16] and minor salivary glands of the oral cavity [17,18]. Recently, a unique case of SPA in the sinonasal mucosa has been published [25]. Reportedly, the age range at presentation is 9–84 years [4,21]. Gnepp [8] reported an average age of 44.5 years with a female predominance. Most patients present with a slow-growing mass with occasional pain and tenderness. The lesion is usually well-circumscribed and may or may not have a (pseudo-)capsule. Histologically, SPA is characterized by a combination of histological features some of which are reminiscent of mammary fibrocystic disease. SPA is mostly unifocal, but rarely may be multifocal and/or bilateral. Recurrences have been reported in up to 19% of cases.

The etiology and pathogenesis of SPA is still unknown. The nature of this lesion was initially believed to be pseudoneoplastic and inflammatory [1,3,24]. Recently, using the HUMARA analysis, we have demonstrated monoclonality in six out of six informative (female) cases of SPA, thus providing evidence that SPA is likely a neoplasm [23]. Moreover, ductal epithelial atypia ranging from mild dysplasia to in situ carcinoma can be found in 40–75% of cases [5,7,10,14,20,22]. Invasive carcinoma arising in SPA has been recently presented at the poster session of the 25th ESP Congress in Lisbon [15]. However, to date, no case of invasive carcinoma in SPA either with or without metastatic disease has been documented in the literature.

Herein we present, for the first time, two cases of SPA in a familial setting affecting two sisters aged 7 and 33 years, respectively. The younger patient suffered multiple recurrences.

Case reports

Case 1

A 7-year-old, otherwise healthy girl presented with a firm nodule under her right ear. Cytological smears revealed a reactive lesion in a lymph node and bilateral tonsillectomy was performed. Six months later two firm, movable, elastic nodules, measuring 1 cm and 2.5 cm in diameter, were noted in her right parotid region and surgically excised. The lesion was initially misdiagnosed as a

☆ The Case 1 was presented as a platform presentation in the Slide Seminar at ESP Congress 2012, Prague, Czech Republic (SM).

* Corresponding author. Tel.: +385 12902740; fax: +385 12903141.
E-mail addresses: spoma@mef.hr, smanojlo@kbd.hr (S. Manojlović).

0344-0338/$ – see front matter © 2014 Elsevier GmbH. All rights reserved.
http://dx.doi.org/10.1016/j.prp.2014.01.006

multilocular pleomorphic adenoma. Eighteen months after that, at the age of 9 years, the patient presented with multiple nodules in the same region. Frozen section was judged to be consistent with the diagnosis of pleomorphic adenoma. Therefore, a conservative parotidectomy was performed. Histopathological examination revealed multiple nodules within both the superficial and deep lobe. All nodules were well-circumscribed and some of them were encapsulated. The lesions were described as "tumor-like" with a peculiar pattern, but a definitive diagnosis of pleomorphic adenoma was not rendered. Ten years later several nodules appeared within the scar. Cytological smears revealed normal cuboidal and cylindrical ductal epithelial cells. An additional firm nodule appeared two years after that. An FNA-examination of the additional firm nodule was judged to be consistent with a recurrent pleomorphic adenoma. An extended total parotidectomy with sacrifice of the facial nerve was performed. The extensive surgical treatment included a part of the sternocleidomastoid muscle, the masseter muscle, the external auditory canal, and the periost of the mandibular ramus followed by postoperative radiotherapy. The histopathological findings were again interpreted as a multilocular recurrent pleomorphic adenoma. The patient has been followed up for 13 years since the last treatment in 2000 with no further recurrences.

Case 2

The older sister of the former patient presented at the age of 33 with a firm, partially fixed lump in her right parotid region, which had been growing slowly for about eight years. The clinical impression was that of a benign salivary gland tumor. Superficial parotidectomy was performed. Gross examination revealed a well-circumscribed nodule, measuring 4 cm in greatest diameter, surrounded by normal parotid gland tissue. The diagnosis rendered after histopathological examination was "consistent with chronic sclerosing sialadenitis". The patient revealed no evidence of recurrences or metastasis during a follow-up period of four years.

Materials and methods

All available tissue material from both cases was collected and reviewed by two of us (SM and AS).

For conventional microscopy, the excised tissues were fixed in formalin, routinely processed, embedded in paraffin, cut, and stained with hematoxylin–eosin. In both cases, additional stains were also performed, including periodic acid-Schiff (PAS) with and without diastase, mucicarmine, and alcian blue at pH 2.5.

For immunohistochemical studies, 4-μm-thick sections were cut from paraffin blocks, mounted on slides coated with 3-aminopropyltriethoxy-silane (Sigma, St. Louis, USA), deparaffinized in xylene, and dehydrated in descending grades (100–70%) of ethanol. An immunohistochemical study with commercial antibodies using protocols according to the manufacturers' recommendation was employed. Antibodies to the following antigens were included: EMA, CK7, AE1/AE3, CEA, S-100 protein, vimentin, p63, calponin, estrogen receptor (ER), androgen receptor (AR), progesterone receptor (PR), HER2/neu (HercepTest), Ki-67 (MIB-1), and EBV. The source of all antibodies was DakoCytomation, Denmark. In situ hybridization using EBER PNA probe (DAKO, Y5200) and BCIP/NBT (DAKO, PNA ISH Detection Kit, K5201) was also performed according to the manufacturers' protocol.

Microscopic findings

Since the specimen of the primary excision in Case 1 was not available, all recurrent lesions in Case 1 as well as the single nodule in Case 2 were analysed. Histological findings were virtually identical, showing well-circumscribed, partly encapsulated nodules surrounded by normal salivary gland tissue. The main characteristic of all nodules was a proliferation of ductal epithelial component in abundant sclerotic stroma focally infiltrated by lymphocytes (Fig. 1). Some of the ducts were cystically dilated and contained ariable amounts of intraluminal, eosinophilic material. Numerous other ducts exhibited intraductal epithelial proliferations with intraluminal papillary projections and cribriform or "roman bridge-like" structures (Fig. 2). Few dilated ducts exhibited florid epithelial hyperplasia with mild cytological atypia but not to suggest "high-grade" ductal carcinoma in situ. Many ducts showed large eosinophilic cells with abundant cytoplasm, and intraluminal projections with decapitation secretion suggestive of apocrine metaplasia (Fig. 3). Focally, nests of epithelial cells with vacuolated cytoplasm reminiscent of sebaceous differentiation or foam cells consistent with macrophages were seen. Adjacent salivary gland tissue exhibited large acinar cells with fine eosinophilic granules and dense globules within cytoplasm (Fig. 4).

Histological findings in all reviewed recurrent lesions of Case 1 as well as the lesion in Case 2 were consistent with diagnosis of sclerosing polycystic adenosis of salivary gland.

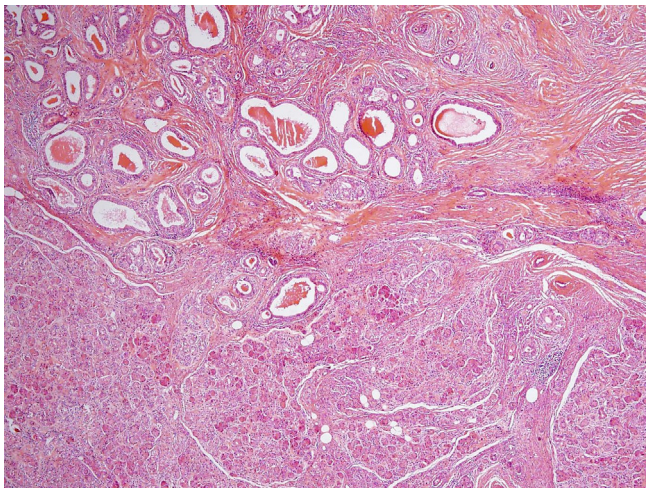

Fig. 1. Proliferation of ducts forming microcysts in abundant sclerotic stroma surrounded by unremarkable glandular structures (Case 1, H&E 100×).

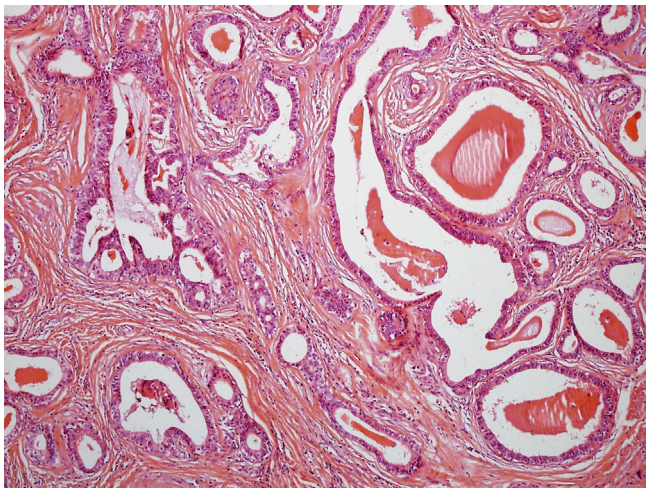

Fig. 2. Hyperplastic epithelium with intraluminal papillary projections and cribriform or "roman bridge-like" structures lining some of the ducts. (recurrent lesion of the Case 1, H&E 200×).