Case study

A unique case of sclerosing polycystic adenosis of the sinonasal tract☆

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Summary Sclerosing polycystic adenosis is an extremely uncommon, recently described, sclerosing lesion of the salivary glands that appears histologically similar to fibrocystic changes of the breast. The key histopathologic features of sclerosing polycystic adenosis include lobular proliferation of ductal and acinar elements, cystically dilated ducts exhibiting frequent apocrine and sebaceous metaplasia, eosinophilic intracytoplasmic granules within some acinar-type cells, intraductal epithelial hyperplasia, and dense fibrosis. Most described cases have occurred in the major salivary glands, particularly the parotid gland. Although most authorities consider sclerosing polycystic adenosis to be a pseudoneoplastic process, the occurrence of dysplasia and carcinoma in situ of ductal epithelium reported recurrence rates of up to 30%, and recent evidence of clonality suggests a possible neoplastic etiology. However, there have been no cases of metastasis. Herein, we report the first case of sclerosing polycystic adenosis of the sinonasal tract in a 79-year-old woman presenting with a sinonasal mass.

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1. Introduction

Sclerosing polycystic adenosis (SPCA) is a rare, sclerosing process of the major and minor salivary glands that was initially described by Smith et al [1] in 1996. The key histologic features of SPCA are lobular proliferation of ductal and acinar elements; cystic ducts with frequent apocrine-like and sebaceous-like cells; eosinophilic cytoplasmic granules within some acinar-type cells; intraductal epithelial hyperplasia with occasional collagenous spherulosis; and dense, frequently nodular fibrosis [1]. These histologic features are reminiscent of fibrocystic changes of the breast. Most of the reported cases have occurred in the major salivary glands, most commonly the parotid gland [1–11]. Only rare cases have been reported in the minor salivary glands of the oral cavity [12–14]. Herein, we report a case of SPCA involving the sinonasal tract, a novel location for this entity.

2. Case report

A 79-year-old, otherwise healthy woman presented with a several-month history of an enlarging and palpable left sinonasal mass. Besides a complaint of left-sided epiphora, the patient denied the presence of nasal airway obstruction, epistaxis, drainage, or decreased sensation along the distribution of the left intraorbital nerve. A preoperative computed tomographic scan of the orbits and paranasal sinuses performed at an outside institution in April 2009
showed a 3.0 × 2.0 × 5.7 cm, heterogeneously enhancing, soft tissue mass in the left nasal cavity with associated remodeling of the left lateral nasal wall and the bony septum. The bony middle and anterior turbinates appeared severely thinned or remodeled. The left osteomeatal complex was obstructed by the mass at the left middle meatus and the hiatus semilunaris. There was no involvement of the left orbit; however, the mass was obstructing the left nasal lacrimal duct at the distal opening of the left inferior meatus of the nasal cavity. In July 2009, the patient underwent an endoscopic nasal cavity exploration, which showed a large, smooth-appearing mass filling approximately 80% of the left nasal cavity. It obliterated and appeared to arise from the left inferior turbinate without involvement of the middle turbinate. A biopsy obtained from the mass was initially interpreted as a respiratory epithelial adenomatoid hamartoma without evidence of malignancy. In September 2009, an endoscopic resection of the mass was performed. At the time of surgery, the mass was noted to be very firm and accommodated the dimensions of the nasal cavity. The mass was excised intact and submitted for pathologic examination.

Macroscopically, the specimen consisted of 3 separate pieces of pink-tan, firm, glistening, and rubbery tissue weighing 15.5 g in aggregate and ranging in size from 1.0 × 0.5 × 0.3 to 5.8 × 2.5 × 1.8 cm. There were small fragments of translucent and membranous-appearing bone attached to the largest piece, the surface of which was focally hyperemic. The cut surfaces of each fragment appeared to have variably sized, smooth-walled, fluid-filled cysts surrounded by dense fibrous tissue.

Histologic examination revealed an exuberant proliferation of both epithelial and stromal elements. The epithelial component consisted of a lobular proliferation of mostly ductal and scattered tubuloacinar structures within a fibrous stroma (Fig. A). These small, closely packed ductal structures were lined by cuboidal epithelial cells with moderately abundant, pale eosinophilic cytoplasm and relatively bland-appearing, round nuclei containing inconspicuous nucleoli (Fig. B). The acinar-type cells contained brightly eosinophilic, intracytoplasmic, modified zymogen granules (Fig. C). No significant mitotic activity, necrosis, or dysplastic features were observed. In some areas, there were prominent cystically dilated ducts that exhibited dense, periductal fibrosis and contained eosinophilic secretions (Fig. D). These ectatic ducts were lined by a markedly attenuated epithelium and embedded within a relatively cellular, bland-appearing fibroelastic stroma (Fig. E).

Immunohistochemical staining was performed using standard immunoperoxidase techniques. The epithelial component was uniformly positive for cytokeratin 7 and negative for cytokeratin 20. Immunostains for cytokeratin 5/6, p63, calponin, and smooth muscle actin (SMA) (Fig. F) highlighted the abluminal, myoepithelial cells surrounding each of the ductal structures. In contrast, carcinoembryonic antigen and c-kit were positive in the luminal cells. Vimentin was positive in the fibroelastic stromal component. No detectable staining was seen for glial fibrillary acidic protein.

Taking together the unique histopathologic findings and the results of the immunohistochemical stains, a diagnosis of SPCA was rendered. Because the mass was completely resected, no further treatment was indicated. The patient has been monitored for recurrence via annual nasopharyngoscopies. Thirty-eight months after the resection of the sinonasal mass, the patient appears to be stable clinically with no evidence of recurrence or metastasis.

3. Discussion

Sclerosing polycystic adenosis is a rare, sclerosing lesion of the salivary glands that was first described by Smith et al [1] in 1996. The histologic features of SPCA are similar to fibrocystic changes of the breast and include lobular proliferation of ductal and acinar elements, cystically dilated ducts, and dense fibrosis [1]. Several cases of dysplasia and carcinoma in situ of the ducal epithelium have been reported [4,5,9,11]. The cytologic features and the associated cytologic diagnostic dilemmas of SPCA have also been described [3]. Thus far, all cases of SPCA reported in the literature have involved the salivary glands, most commonly the parotid gland [1–14].

Although SPCA is generally believed to be a reactive lesion, Skalova et al [10] demonstrated a pattern of X-chromosome inactivation in 6 of 12 cases of SPCA indicating a nonrandom, clonal process suggestive of a neoplastic etiology. Further evidence of a possible neoplastic etiology was provided by a study demonstrating the presence of Epstein-Barr virus in the lesional cells of SPCA by immunohistochemistry and real-time polymerase chain reaction [15]. However, reported cases of SPCA have been associated with a favorable outcome; recurrence is possible, but metastasis has not been described [1,2,4–6,8,12,14].

The sinonasal tract is lined by respiratory mucosa and contains seromucinous glands. Salivary gland tumors such as adenoid cystic carcinoma and mucoepidermoid carcinoma are known to occur in the sinonasal tract. Thus, it is not unreasonable to expect that other salivary gland lesions may occur in this location. The current case of SPCA diagnosed on a mass lesion arising from the left inferior turbinate is the first reported case of SPCA in the sinonasal tract.

If the histologic appearance of a sinonasal mass suggests a diagnosis of SPCA, the differential diagnostic considerations might include tumors with cystic or oncocytic features and benign lesions such as respiratory epithelial adenomatoid hamartoma, nasal seromucinous hamartoma (microglandular adenosis), and inflammatory polyp. One diagnostic consideration is mucoepidermoid carcinoma, which may resemble SPCA in that it exhibits cystic change, mucous and squamous cells, and desmoplastic stroma and inflammation. However, mucoepidermoid carcinoma may also contain
Sclerosing polycystic adenosis. A, Exuberant proliferation of ductal and tubuloacinar elements in a dense fibrous background (hematoxylin and eosin, original magnification ×40). B, Small, closely packed ductal structures lined by benign-appearing cuboidal epithelial cells (hematoxylin and eosin, original magnification ×200). C, Acinar-type cells containing brightly eosinophilic, intracytoplasmic, modified zymogen granules (hematoxylin and eosin, original magnification ×200). D, Prominent cystically dilated ducts with intraluminal eosinophilic secretions and dense, periductal fibrosis (hematoxylin and eosin, original magnification ×40). E, Ectatic ducts lined by markedly attenuated epithelium embedded within a relatively cellular, bland-appearing fibroblastic stroma (hematoxylin and eosin, original magnification ×200). F, Immunohistochemical stain for SMA highlighted the abluminal, myoepithelial cells surrounding each of the ductal structures (original magnification ×200).
features such as necrosis, infiltration, and increased mitotic activity and nuclear atypia, which should be absent or minimal in SPCA. Distinguishing between a low-grade adenocarcinoma and SPCA may present a diagnostic dilemma in light of the exuberant ductal proliferation but can be resolved by recognition of the lobular configuration of the latter. Furthermore, immunohistochemical stains for myoepithelial cells such as SMA, calponin, p63, and cytokeratin 5/6 will highlight the abluminal cells of SPCA and will be absent in low-grade adenocarcinoma. Sinonasal hamartomas such as respiratory epithelial adenomatoid hamartoma and seromucinous hamartoma may contain numerous glands and stroma with a variety of mesenchymal elements. The former contains glands that have a ciliated respiratory epithelium surrounded by a thick basement membrane. The latter exhibits seromucinous glands, which, in contrast to SPCA, lack a myoepithelial cell layer [16,17]. As such, immunostains for myoepithelial cells are a useful diagnostic adjunct in distinguishing between these 2 differential diagnostic considerations, particularly in small biopsy specimens. Inflammatory polyps have an edematous stroma with inflammation and may show prominent seromucinous glands. In contrast with the aforementioned benign lesions, the ducts in SPCA show frequent apocrine and sebaceous metaplasia. Moreover, the ducts and acini of SPCA generally have a lobular configuration.

As more cases of SPCA involving the sinonasal tract are reported along with follow-up data, more information regarding the behavior of SPCA in this location will be obtained. However, we speculate that SPCA in the sinonasal tract has a pathogenesis similar to SPCA in the salivary glands, and therefore, SPCA in the sinonasal tract may recur but should have a favorable outcome. The current case shows no evidence of recurrence or metastatic disease 38 months after resection. When more data become available, the exact behavior of SPCA arising in the sinonasal tract can be determined.

Since the first description by Smith et al [1] in 1996, SPCA has been increasingly diagnosed in salivary glands. In a similar fashion, as SPCA becomes more recognized in other locations such as the sinonasal tract, the diagnosis of SPCA may be made more often in the future. As more pathologists become familiar with SPCA, the diagnostic accuracy of lesions of the sinonasal tract will be enhanced.

References