Excision of Sclerosing Polycystic Adenosis of the Parotid Gland

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Abstract

A 15-year old male presented to the hospital with a 2-month history of a right parotid mass. Magnetic Resonance Imaging revealed an encapsulated 3.7 cm mass. Fine-needle aspiration suggested a monomorphic adenoma. Total parotidectomy was performed with dissection and preservation of the facial nerve. Pathology identified the lesion as Sclerosing Polycystic Adenosis (SPA) after examining the histopathological and immunohistochemistry findings. Sclerosing polycystic adenosis is a rare lesion which leads to inflammatory changes in the salivary gland. The fibrocystic changes are morphologically similar to the histological developments of sclerosing adenosis of breast tissue.

Background

Sclerosing polycystic adenosis (SPA) is an extremely rare lesion of the salivary glands predominately found in the parotid that is reminiscent of histopathological changes that occur in the mammary gland. First described in 9 patients by Smith et al. in 1996, to date only 66 cases have been reported with the vast majority detailed in case reports and limited case series. Initially felt to be a pseudoneoplastic sclerosing and inflammatory process, more recently there have been reports of increased recurrence and one case of malignant transformation.

Case Description

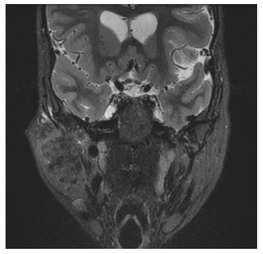
A 15-year old male presented with a 2-month history of right sided mass at angle of mandible. The area was initially tender but was asymptomatic at time of presentation. CT neck with contrast identified a 2.8 cm hyperdense mass in the right parotid gland and MRI showed a heterogenous, 3.7 cm mass predominantly in the superficial gland, with likely involvement of the deep lobe. T2/STIR images were hypointense with avid enhancement, and no signs of restriction diffusion. FNA suggested a monomorphic adenoma of the salivary gland versus a neoplasm of indeterminate malignant potential.

The patient underwent a total parotidectomy with dissection and preservation of facial nerve as the entire mass was found within the the deep lobe of the gland. Surgical pathology grossly showed a well-encapsulated, firm, white-yellow, lobular mass measuring 2.1 x 1.9 x 1.7 cm. Histology revealed dense sclerosis intervening between lobules of salivary gland ducts with occasional fibrocystic change. There were focal ductal hyperplasia with mild epithelial atypia, intraductal necrosis, foci of acinar cells with eosinophilic zymogen granules. Immunohistochemistry confirmed the benign and hyperplastic nature of the lesion

At six month follow up, patient was doing well with intact facial nerve function and no recurrence noted.

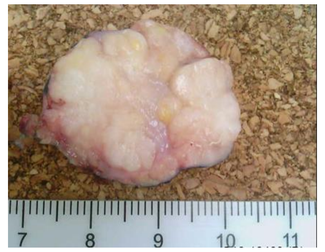
Results

MRI



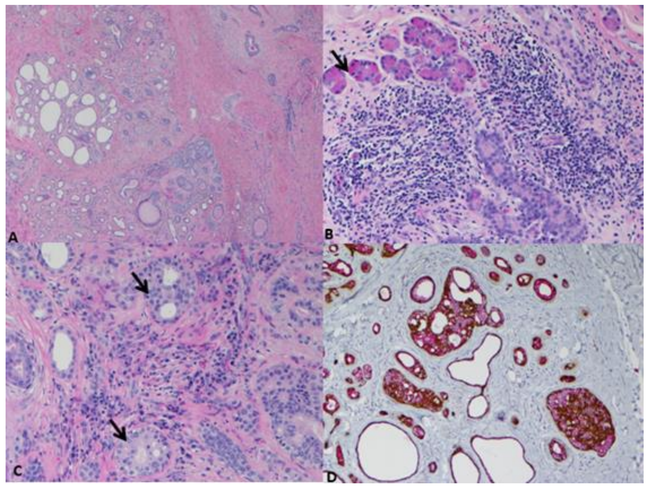
T2 weighted MRI with 3.7 cm, hypointense, heterogenous mass in right parotid

Gross Pathology



Cut surface shows a lobulated yellow-tan, pale, glistening lesion. Lesion lacks grossly visible areas of necrosis or hemorrhage

Histology



(A) Bands of sclerosis and fibrous tissue separating lobules of variably sized cystic glands,
(B) Acinar cells with eosinophilic zymogen granules
(C) Ductal epithelial cells with hyperplasia
(D) BNC-5 immunohistochemical stain confirming presence of myoepthelial cells and a benign hyperplastic ductal epithelium

Discussion

Sclerosing polycystic adenosis is a rare tumor of the salivary glands. About 80% of SPA cases occur in the parotid gland, but cases have been reported in the submandibular gland (7%), and minor salivary glands (12%) including the buccal mucosa, retromolar pad, and hard palate. Patients are often asymptomatic, CT reveals a heterogenous, well-circumscribed lesion with bright small cystic areas on T2-weighted MRI.

FNA can be challenging due to the ambiguous nature of the lesion with differential including Warthin tumor and pleomorphic adenoma, as well as possible malignant lesions such as mucoepidermoid carcinoma and low grade adenocarcinoma.

On pathology the nodular, sclerotic, and inflammatory lesion shares histological similarities to fibrocystic changes of the breast. The presence of acinar cells with abundant eosinophilic granules, similar in structure to zymogen granules, is highly characteristic for SPA.[1]

Immunohistochemical analysis shows the acinar and ductal components are positive for broad spectrum cytokeratins (AE1/3) and epithelial membrane antigen, and Gross Cystic Disease Fluid Protein (GSDFP) 15 is strongly expressed in the acinar component.[2] In the literature 25% - 30% of cases had low-grade dysplasia and ~ 10% of cases had high-grade dysplasia or low-grade carcinoma in situ.[3]

There has been a reported recurrence rate of 15 – 30% that is higher with enucleation (44 – 67%) vs superficial (7%) or total parotidectomy (10%).[4]

Due to rate of recurrence and high-grade dysplasia in close proximity to facial nerve, management is surgical.

Conclusion

SPA is a rare lesion of the salivary glands with one case of malignant transformation reported. Recurrence varies but is significantly less with superficial or total parotidectomy. Follow-up frequency of at least 5 years is required due to the rate of recurrence.

References

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[1] *Smith BC, Ellis GL, Slater LJ, et al. Sclerosing polycystic adenosis of major salivary glands. A clinicopathologic analysis of nine cases. Am J Surg Pathol. 1996;20:161–170*
[3] *Gnepp DR, Wang LJ, Brandwein-Gensler M, et al. Sclerosing polycystic adenosis of the salivary gland: a report of 16 cases. Am J Surg Pathol. 2006;30:154–164.*
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