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Accessory parotid gland carcinoma ex pleomorphic adenoma. Case study diagnosed by fine needle aspiration

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SUMMARY

Objective. The accessory parotid gland is salivary tissue separated from the main parotid gland and lying on masseter muscle. It has secondary duct empting into the Stensen's duct. The accessory parotid gland exists in 21-61% of individuals. However, the appearance of an accessory parotid tumor is rare, with a reported frequency of 1-7.7% of all parotid gland tumors. Carcinoma ex pleomorphic adenoma arises from a pre-existing benign mixed tumor.

Most of these tumors will have malignant epithelial component, but not malignant stromal component. Reports of Fine Needle Aspiration Cytological (FNAC) diagnosis of malignant mixed tumor are uncommon and have been limited to cases arising in the parotid. We report a case of carcinoma ex pleomorphic adenoma of the accessory lobe of the parotid, and address the cytopathology features and pitfalls of this condition.

Case. A 73 aged female presented with a right nontender midcheek mass.

The lesion had been present 18 months, with a recent increase in size. FNA was performed and the smears demonstrated features indicative of pleomorphic adenoma admixed with findings indicative of a poorly differentiated carcinoma.

Conclusion. FNAC can accurately diagnose carcinoma ex pleomorphic adenoma when strongly fixed requirements are implemented.

Key words: accessory parotid gland carcinoma, pleomorphic adenoma, fine needle aspiration cytological diagnostics.

INTRODUCTION

Accessory parotid gland tumors are defined as masses within salivary gland tissue located adjacent to Stensen's duct, but separate from the main body of the parotid gland. A mass arising in the mid-cheek region may often be overlooked as a rare accessory lobe parotid neoplasm. Accessory parotid gland tissue has been described as salivary tissue adjacent to Stenson's duct that is separate from the main body of the parotid gland. Accessory parotid glands are present in approximately 21% of the population [1]. These small flat structures were once considered to be mere extensions of the main parotid gland, but it is now known that they are independent glandular units with respect to their function and anatomic location. Accessory parotid glands are associated with a higher rate of malignant tumors (26 to 50%) than are the main parotid glands (18 to 20%) [2]. It has been postulated that this higher rate of malignancy is attributable to the histology of the accessory parotid gland. In contrast to the predominant serous composition of the main parotid gland, the accessory parotid gland is made up of a fairly equal percentage of mucinous and serous acinar units, as is the submandibular gland [3]. The lack of anatomic barriers to tumor extension predisposes these tumors to significant soft-tissue infiltration.

We herein describe a case of accessory parotid gland tumor. The diagnosis was carcinoma ex pleomorphic adenoma.

CASE STUDY

A 73 year old female presented to the Ear-Nose-Throat Surgery clinic with a right nontender midcheek mass. The lesion had been present 18 months, with a recent increase in size. Physical examination revealed a 2 cm, ill defined mass and computed tomography confirmed a solitary mass separating from the main parotid gland (Fig. 1).

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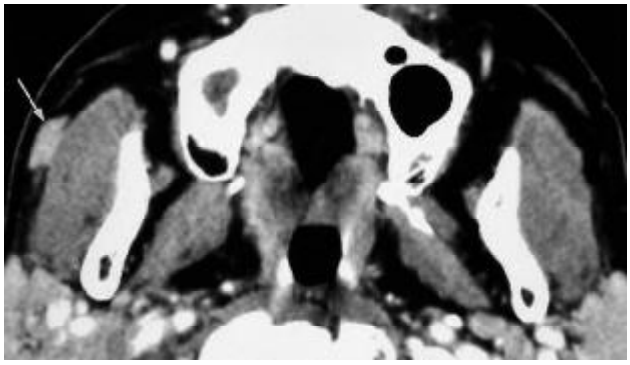

Fig. 1. CT shows a subcutaneous mass (arrow) outside the masseter muscle and separated from the right main parotid gland

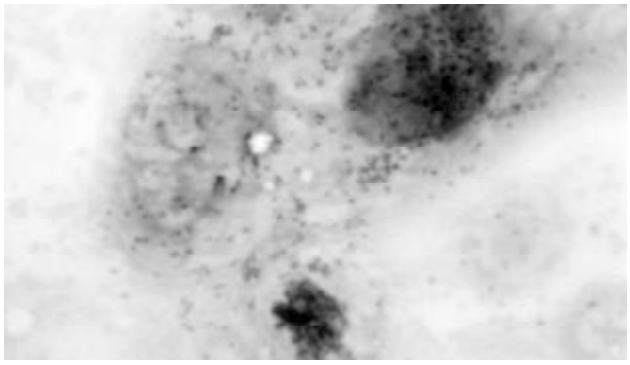

Fig. 2. Benign component demonstrated features of a mixed tumor with abundant metachromatic and fibrillary stroma and small round or ovoid bland cells in clusters. Direct smear. Papanicolaou stain X 200

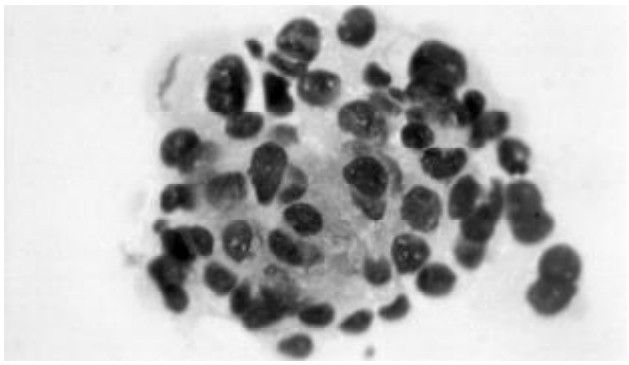

Fig. 3. Malignant component showing large pleomorphic cells occurring in non cohesive groups with anisonucleosis and overlapping. Direct smear. Papanicolaou stain X 200

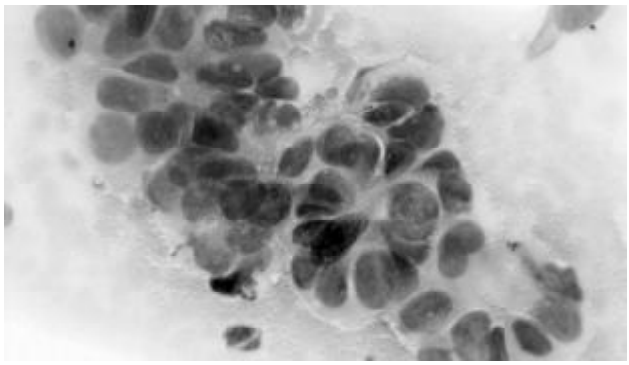

Fig. 4. Neoplastic cells in glandular or pseudoacinar structures. Direct smear. Papanicolaou stain X 200

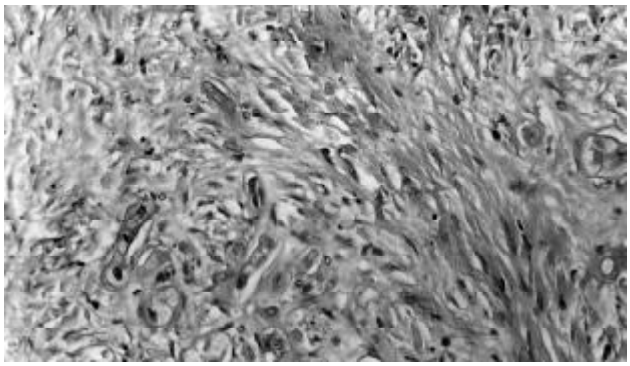

Fig. 5. Poorly differentiated adenocarcinoma. Cell block preparation. Hematoxylene-Eosin stain X 200

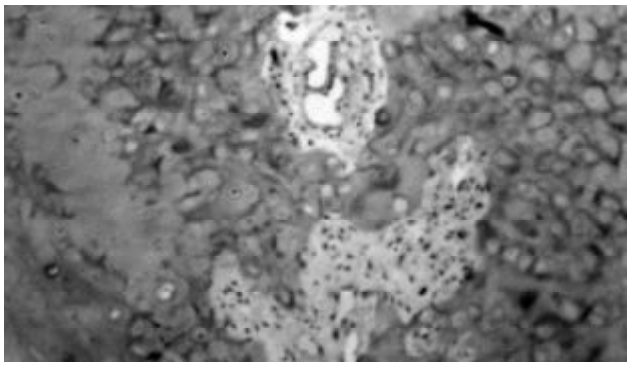

Fig. 6. Benign areas showing large portions of acellular, blue-gray chondromyxoid stroma and mature cartilage with intermixed glandular epithelium. Tissue section. Hematoxylene-Eosin stain X 200

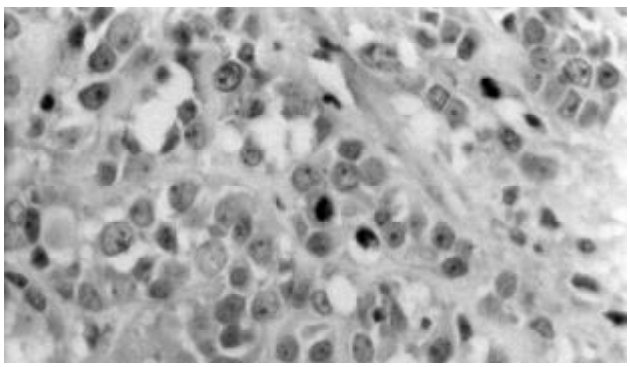

Fig. 7. Malignant epithelial structures described as poorly differentiated adenocarcinoma. Tissue section. Hematoxylene-Eosin stain X 200

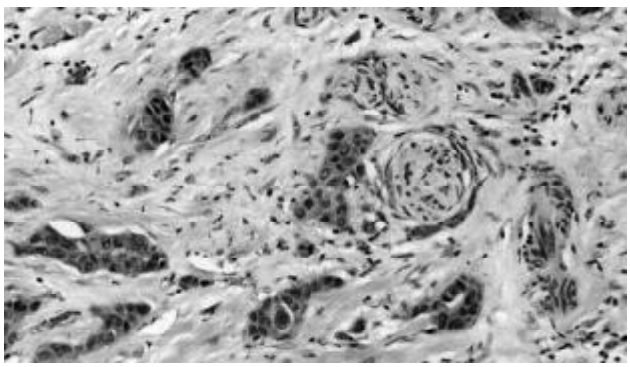

Fig. 8. Perineural involvement. Tissue section. Hematoxylene-Eosin stain X 100

FNA was performed using a cutaneous approach with a 22-gauge needle. Material from aspirates was divided between air dried Diff-Quik stained slides and ethanol fixed Papanicolaou stained smears. A paraffin embedded cell block was made from the needle rinses. Sections from the cell block were stained with the Hematoxylene-eosin technique.

Examination of the smears revealed a mixture of benign and malignant elements. The benign component of the smears demonstrated features of a mixed tumor with abundant metachromatic and fibrillary stroma (Fig. 2) and small round or ovoid bland cells occurring in clusters. The malignant component showed large pleomorphic cells occurring in non cohesive groups with anisonucleosis and overlapping (Fig. 3). Some of the neoplastic cells occurred in glandular or pseudoacinar structures (Fig. 4). The nuclei contained irregular membranes, and macronucleoli. They resembled as those cells derived from a poorly differentiated carcinoma of glandular origin

Cell block interpretation showed areas of clearly defined, mature cartilage along with a poorly differentiated adenocarcinoma (Fig. 5). Following cytologic diagnosis under general anesthesia a standard preauricular-submandibular incision with an anterior extension was made, and then dissection was carried forward in order to expose the anterior margin of the parotid, and the zygomatic and buccal branches of the facial nerve. The tumor existing over the masseter muscle without any connection with the parotid, was extirpated. A single accessory duct running from the tumor to Stensen duct was ligated and resected with the tumor. Grossly the tumor consisted of an apparently well circumscribed, encapsulated mass of homogeneous, white-tan, solid tissue measuring 2,8x1,5x1,5 cm. A total of eight adjacent lymph nodes were found in the limited neck dissection.

The tumor was composed of both benign and malignant elements. Benign areas included large portions of acellular, blue-gray chondromyxoid stroma and mature cartilage with intermixed glandular epithelium (Fig. 6). The epithelial component displayed trabecular and tubular features. Individual cells were medium sized with moderate amounts of cytoplasm and rounded angulated nuclei with infrequent nucleoli. Malignant epithelial structures were best described as poorly differentiated adenocarcinoma (Fig. 7**)**. The cells showed an increased nuclear/cytoplasmic ratio, pleomorphic nuclei with irregular contours, nucleoli and mitotic figures. Capsular invasion was focally identified, with malignant cells penetrating 3 mm beyond the pseudocapsule. Surgical margins were tumor free; vascular involvement was absent but perineural involvement was found (Fig. 8). None of the nodes had evidence of metastasis.

DISCUSSION

There are three types of malignant pleomorphic adenoma. The first type is a benign pleomorphic adenoma that metastasizes as benign pleomorphic adenoma. The second type is a carcinosarcoma. The third type, and perhaps the most common type, is carcinoma ex pleomorphic adenoma (CEPA). This condition is a carcinoma that develops in association with a benign primary or recurrent pleomorphic adenoma. Although CEPA are uncommon, these lesions account for most of the reported cases of malignant pleomorphic adenoma. The carcinomatous component is usually of high histologic grade. These clinically aggressive lesions often lead to metastasis and a tumor related death. CEPAs account for 3.6% of all salivary neoplasms and 11.7% of salivary malignancy [4].

It is an aggressive tumor with high overall mortality. Most of these lesions are found in major salivary glands. An asymptomatic mass is the most common symptoms. The tumors range in size in 1 to 17 cm with a mean of 3.9 cm. Tenderness and involvement of the facial nerve are seen in some cases. Nearly half of the patients noticed a painless mass in less than a year [5].

Some patients may experience rapid expansion of a pre-existing mass. When they occurs in the major salivary gland, CEPA are 6 times more frequently seen in the parotid gland than the submandibular gland [5]. As per one large series by Oslen and Lewis, the age range from 34 to 95 years with a mean of 61 years. There is a male predilection and a male:female ratio of 1.8:1. The ratio of primary to recurrent tumor is about 9:1. Survival is largely related to clinical staging. While the 5-year survival for stage I disease is over 80%, the overall 5-year survival is only 37% [5].

Malignant transformation in pleomorphic adenoma occurs most commonly in long-standing lesions or lesions with multiple local recurrences. The risk increases with the duration of the tumor. While only 1.6% of malignant transformation occurs in tumors less than 5 years, the risk for tumors over 15 years is 9.5% [6]. The most common carcinomatous components are adenocarcinoma not otherwise specified (NOS) (42.4%) and salivary duct carcinoma (32.8%). The less common entities include adenosquamous cell carcinoma, adenoid cystic carcinoma, undifferentiated carcinoma, myoepithelial carcinoma, epithelial-myoepithelial carcinoma, and sarcomatoid carcinoma [5]. The volume of the pleomorphic adenoma is often small and makes the diagnosis of CEPA difficult.

In our case, cytodiagnosis was both time and procedure sparing; malignancy was diagnosed by FNA at the patient initial clinical presentation. Furthermore, the

diagnosis of a primary malignancy, rather than a metastatic process, allowed the patient to be appropriately scheduled for a single operation without an intervening open biopsy.

Definite diagnosis of malignant mixed tumors on FNA have been infrequently reported [7,8]. Jacobs reported misdiagnosis of a low grade mucoepidermoid catcinoma arising in a pleomorphic adenoma (CEPA) from failure to identify malignant mucin-producing cells along with the typical biphasic pattern of benign epithelium and chondromyxoid stroma [9]. There have also been described false negative diagnoses of the malignant component of pleomorphic adenomas because of sampling error or misinterpretation [10]. In our case the cytopathologist aspirated the lesion four times even though the first pass was quite cellular and adequate to establish the diagnosis of a mixed tumor. The malignant component was not demonstrated until the next passes were performed. Thus it is recommended to sample the lesion at least three times. On the other hand attention must be paid to avoid overdiagnosis of CEPA since benign mixed tumors can be very cellular and show cytologic atypia; spontaneous infarction [11] and polyploidy [12] have been reported to cause atypia. Moreover, benign pleomorphic adenoma may demonstrate cylindromatous or cribriform pattern that masquerades adenoid cystic carcinoma and be misdiagnosed as malignant [13].

CONCLUSIONS

Benign and malignant epithelial parotid tumors can be diagnosed by their clinical presentations supplemented with FNAC [14].

Accuracy, sensitivity and specificity of FNAC on salivary gland pathology compare favorable to frozen section analysis [15]. Some could suggest that interpreting primary against metastatic tumor, rather than establishing a specific entity, is the aim of FNAC of salivary gland malignancies. However we have shown that CEPA can be accurately interpreted under intimate standards.

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