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Case Report

Sarcomatoid Carcinoma of the Parotid Gland: A Case Report and Literature Review

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Abstract

Despite the benign features that constitute the majority of pleomorphic adenomas, there are rare instances where malignant transformations have been noted to occur. The most common of these transformations is carcinoma ex-pleomorphic adenomas. This subset can be further classified by unique histological variations that represent their malignant component. We present an unusual case of a rapidly enlarging parotid mass discovered to be a sarcomatoid carcinoma ex pleomorphic adenoma. This case will address the different categories of malignant mixed tumors, discuss the prevalence and clinical relevance of their histological sub classifications, and highlight different treatment modalities.

Introduction

Carcinoma ex pleomorphic adenoma (CXPA) is defined as a carcinoma arising *de novo* or from a recurrent pleomorphic adenoma. Histologically, it can be sub-classified by upwards of eight different variations (e.g., myoepithelial carcinoma, salivary duct carcinoma, adenoid cystic carcinoma sarcomatoid carcinoma) [1]. They are rare entities, constituting only 3.5% of all salivary gland neoplasm's with an estimated prevalence rate of 5.6 cases per 100,000 malignant neoplasm's [2]. Patients frequently present in the 7th decade of life with facial nerve paralysis, paresthesia, or pain subsequent to invasion into the facial nerve [2]. Survival outcomes depend heavily on factors such as advanced-stage disease, multiple lymph node metastases, perineural or vascular invasion, or state of recurrence [3]. However, only few reports have discussed the clinical relevance that histologic subtype has on prognostic decisions.

Case Report

A 70 year-old male with a 40 pack-year smoking presented to clinic with a one-month history of a left perimandibular mass discovered while shaving. It had grown substantially over several days with accompanying discomfort at the site, mild trismus, and left lower lip weakness. He had three left sided temporomandibular joint surgeries, significant sun exposure and no history of skin cancers. He quit smoking 20 years ago and consumed three alcoholic beverages daily. Physical exam revealed a three-centimeter mobile, mass overlying the angle of the mandible that was tender to palpation. Overlying skin was mobile without erythema. There was mild left lower lip weakness, though no other facial asymmetry was observed. No cervical adenopathy was present.

Computed tomography showed a 2.6 cm left parotid mass of the superficial lobe with irregular borders and intralesional calcification (Figure 1). FNA showed "highly pleomorphic cells present with rare mitotic figures consistent with a malignant neoplasm." A radical parotidectomy with cable nerve graft reconstruction of the buccal branch of facial nerve, neck dissection (levels I-V), and wide local excision of cheek skin was performed. The defect was closed with advancement flaps.

Gross examination of the specimen revealed a 3 cm well-circumscribed tan-white stellate nodule with areas of hemorrhage and necrosis. Histology revealed two distinct morphologies present within the lesion (Figures 2-4). Immunohistochemistry showed the malignant components were negative for pan-cytokeratin, CAM5.2, and S-100 and positive for vimentin, epithelial membrane antigen (EMA), smooth muscle antigen (SMA) and p63 (focally). These immunohistochemical findings confirmed the carcinomatous nature of the lesion and given the spindled morphology on H&E, a diagnosis of sarcomatoid carcinoma ex pleomorphic adenoma was rendered.

The patient's case was reviewed at the multidisciplinary tumor board. He was staged T2N0M0 and received postoperative radiation therapy (74.4Gy) based on the consensus recommendations. He was last seen 2 years 9 months post-op with no evidence of recurrence and mild corner of mouth facial weakness which has progressively improved.

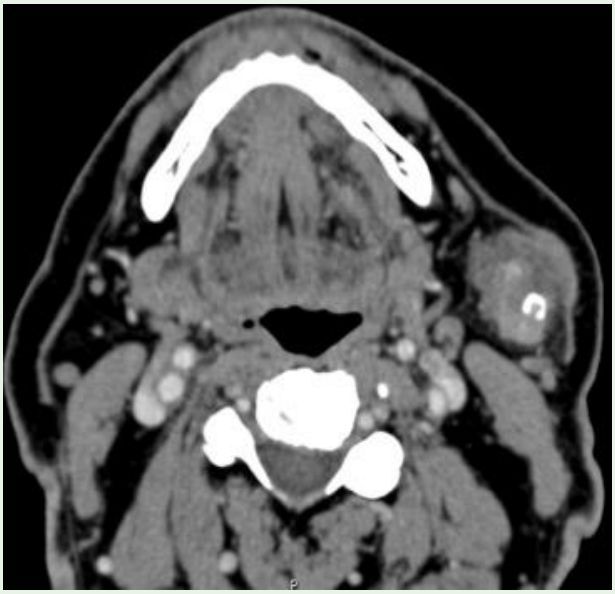

Figure 1: Axial CT scan showing left tail of parotid mass with invasion of the overlying musculoaponeurotic system and intralesional calcification.

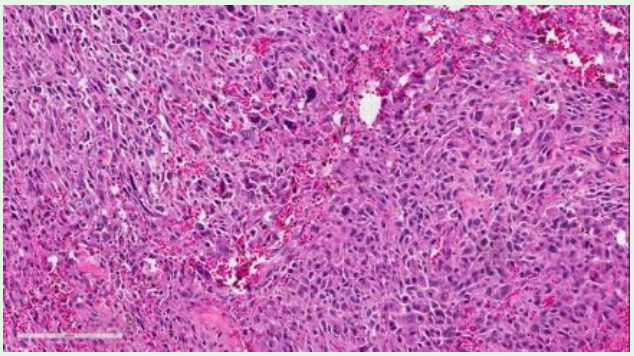

Figure 2: (200X, H&E): This figure demonstrates spindle to epithelioid cells with severe atypia and numerous mitotic figures including an atypical one.

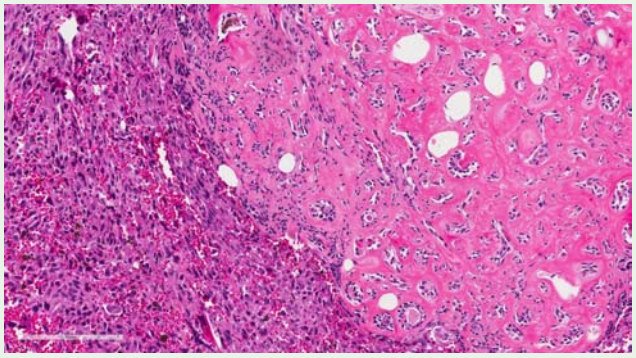

Figure 3: (200X, H&E): This figure demonstrates the interface between bland tubular structures in a hyalinized background and severely atypical cells.

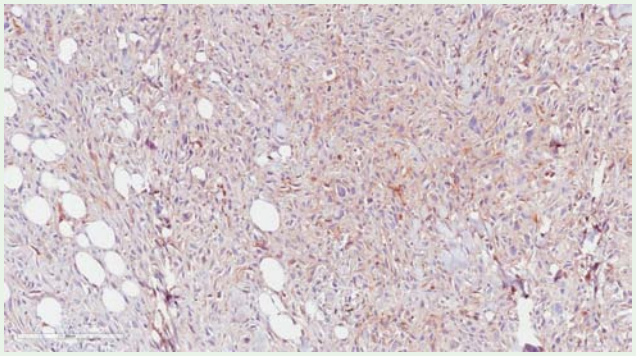

Figure 4: (200X, EMA IHC): The severely atypical cells demonstrate cytoplasmic staining with epithelial membrane antigen (EMA).

Discussion

Carcinomatous transformation of a pleomorphic adenoma was first described by Beahrs et al. in 1957 [4]. Since then, this transformation has been shown to progress into three different ways: carcinosarcomas, carcinoma ex pleomorphic adenoma, and metastasizing pleomorphicadenomas [5]. The primary distinction behind the first two ultimately lies on the extent of malignancy within its tissue components. Whereas both stromal and epithelial cells have malignant properties in carcinosarcomas, this is often just limited to epithelial components in CXPA [5].

There is a paucity of literature describing the molecular differences between types of CXPA. In one major review of 73 cases of CXPA, Lewis et al found the most common classifications were adenocarcinoma not otherwise specified (NOS, 31 cases, 44%) and salivary duct carcinoma (24 cases, 34%). Least common were epithelial-myoepithelial carcinoma (1 case, 1%) and sarcomatoid carcinoma (1 case, 1%) [1]. Similar numbers were reported in another review of 36 cases by Mariano et al. with higher number of salivary duct carcinoma (15 cases, 42%) and just one case of sarcomatoid carcinoma (2.8%) [6]. Another recent review of 43 cases by Katabi et al. also showed higher frequency of salivary duct carcinoma (25 cases, 58%) and myoepithelial carcinomas (15 cases, 35%) without a single case of sarcomatoid carcinoma [7]. There have reports of sarcomatoid carcinomas (i.e. spindle cell tumors with positive EMA and vimentin markers)arising in different organs throughout the body [8]. However, this is the third reported case of a sarcomatoid carcinoma arising from an underlying pleomorphic adenoma of the parotid gland.

When the classifications of CXPA were further analyzed, conflicting results unraveled in regards to their clinical use. Lewis et al. felt the histologic subtype did not have a statistically significant relationship with outcome. Instead, they argued that the focus should remain on tumor staging, proportion of carcinoma, extent of invasion and proliferation index of the carcinoma [1]. This coincides in part with Mariano et al who found concluded that there were statistically significant differences in the final proliferative index of different histological subtypes.

Interestingly, the single case of sarcomatoid carcinoma in their study was found to have proliferative index predominately higher than the average, but lack of additional cases precluded any definitive

findings [6]. Additionally, the literature review of sarcomatoid carcinomas in other organs summarized similar conclusions to Lewis et al. [8].

When comparing the progression from residual (benign) pleomorphic adenoma to CXPA, Mariano et al. found notable results amongst the subtypes. Salivary duct carcinoma, adenocarcinoma NOS had statistically significant proliferation index progression and myoepithelial carcinoma had strong tendency for significance. This showed that there were certain subtypes of pleomorphic adenomas that may have a higher propensity to undergo transformation [6]. Furthermore, Katabi et al. found associations linking higher level of invasiveness (i.e. lower survival outcome) with specific histologic subtypes. From the 43 cases they described, 30 were found to be "wildly" invasive. Of those, 16 (53%) were salivary duct carcinoma and 13 (43%) were myoepithelial carcinoma. Therefore, almost all of their cases of myoepithelial carcinoma (87%) presented with a significantly poor prognosis [7]. In an earlier study, Tortoledo et al. also cited a 5-year survival rate of 62% for patients with ductal carcinoma subtypes, and 50% for myoepithelial carcinomas [9].

The current standard for all patients with high-grade parotid malignancies includes parotidectomy with neck dissection. Facial nerve preservation is recommended at sites free of tumor infiltration. Any facial paresis/paralysis encountered usually indicates tumor involvement, and affected branches of the facial nerve should be sacrificed. If needed, the great auricular nerve serves as a readily available option for cable nerve grafts as long as it is clear from the tumor margins [10]. In patients with poor prognostic factors, postoperative radiation therapy is generally recommended. One particular study by Chen et al. has shown a significantly improved 5 year local control rate overall, and higher survivals in patients without evidence of cervical lymph node metastasis [11]. However, there were no specific studies found connecting the histologic subtypes to treatment responses.

Conclusion

The subtype and histologic details of CXPA play a role in prognostication. Treatment plans should include aggressive wide local excision, neck dissection and high dose post-operative radiation therapy.

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