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Case Report

Metastatic Malignant Perivascular Epithelioid Cell Tumor (PE-Coma) in Parotid Gland Diagnosed by Fine Needle Aspiration Biopsy- a Case Report and Literature Review

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Abstract

Perivascular Epithelioid Cell Tumors (PEComas) are exceedingly rare mesenchymal tumors in salivary glands; no case of metastatic malignant PEComas in salivary gland was described. Here we reported a case of a 74-year old female with a history of malignant PEComa of retro peritoneum, who underwent surgical resection 11 years ago. She had metastatic PEComas in her liver and right upper extremity 7 months prior to the current presentation, with a 0.6 cm soft mobile palpable nodule in the right parotid region without tenderness. Fine needle aspiration biopsy showed spindle-shaped cells with indistinct cell border, wispy acidophilic cytoplasm, and oval to cigar-shaped hyper chromatic nuclei with evenly distributed fine chromatin. No epithelioid form was identified. Immunohistochemistry study showed the neoplastic cells were positive for smooth muscle actin, h-caldesmon and negative for cytokeratin, S-100 and HMB-45. Diagnosis of metastatic malignant Perivascular epithelioid cell tumor was made and the patient was continued with chemotherapy. This case is the first report of metastatic malignant PEComa in parotid gland. The patient's known history of malignant Perivascular epithelioid cell tumor is important when cytopathologists make the differential diagnosis for the mesenchymal neoplasm of salivary gland.

Perivascular Epithelioid Cell Tumor (PEComa) is a family of mesenchymal neoplasms [1-2]. The most common PEComas include renal Angiomyolipoma (AML), pulmonary Lymphangioleiomyomatosis (LAM) and Clear Cell Sugar Tumor (CCST) of the lung. It is exceedingly rare in salivary glands, to our knowledge, only 3 cases of primary angiomyolipoma of parotid gland were reported [3]. No case of metastatic malignant PEComas in salivary gland was described. We report a case of metastatic malignant PEComa in parotid gland diagnosed by fine needle aspiration biopsy. The literatures on this rare mesenchymal neoplasm are reviewed.

Case Report

This was a 74-year old female with a history of malignant PEComa of retro peritoneum, who underwent surgical resection 11 years ago. She was followed up with no evidence of recurrence until7 months prior to the current presentation that a large liver mass was found on imaging and around the same time, a right upper extremity inner arm mass was noticed. These two masses were excised respectively and both pathology diagnoses were metastatic malignant PEComas. They are strongly and diffusely positive for Smooth Muscle Actin (SMA), Desmin, SMMS, H-Caldesmon, MiTF and HMB-45 on Immuno-histochemical studies. A palpable nodule was noticed in the right parotid region two months after abovementioned surgical procedures. Cytopathology service was requested by the otolaryngologist to perform a fine needle aspiration biopsy. The nodule was palpable, soft, mobile and measuring 1.5 cm. It appears on ultrasound exam as a hypo echoic, well-defined mass, along anterior margin of right parotid gland. FNA is performed by a cytopathologists using 22 gauge needles attached to disposable 20 ml syringes. Air-dried smears were stained with Diff-Quik staining and rehydrated smears were stained with Papanicolaou staining. Immediate cytological evaluation was performed by cytopathologists to

provide assessments for specimen adequacy. In addition, needles were rinsed in transport medium and cell blocks were prepared. She elected against surgical procedure and started Everolimus/ Afinitor as systemic management for her PEComa.

Cytological and Immuno-Histochemical Evaluations

The cytology smear showed multiple small groups of spindly shaped cells with cellular disorientation, indistinct cell border and wispy acidophilic cytoplasm. The nuclei feature oval to cigar shape and hyperchromasia. No epithelioid form was identified. The H&E stained cell block preparation showed small clusters of spindly shaped neoplastic cells with eosinophilic cytoplasm with occasional vacuolation and hyper chromatic nuclei. No necrosis or mitosis is present. Immunohistochemistry studies with adequate controls showed the neoplastic cells were positive for smooth muscle actin, h-caldesmon and negative for cytokeratin, S-100 and HMB-45.

Discussion

Perivascular Epithelioid Cell Tumors (PEComas) are exceedingly rare mesenchymal tumors in salivary glands, to our knowledge; only 3 cases of primary angiomyolipoma of parotid gland were reported. No case of metastatic malignant PEComas in salivary gland was described previously. The cytological features of PEComas is limited in the reports of a few pancreas and liver FNAs for primary or metastatic PEComas, in which small and large cohesive clusters of bland-appearing epithelioid to spindle cells with oval or elongated nuclei, indistinct nucleoli, and finely vacuolated cytoplasm are identified [1]. Among these, 5 out 11 pre-operative EUS guided FNA were diagnostic for PEComas. The remaining cases were misinterpreted as non-diagnostic, poorly differentiated carcinoma, clear cell carcinoma or neuroendocrine tumors [2]. Adequate tissue sampling and ancillary Immuno-histochemical studies are critical for propitiate interpretation in cytology. In this case, the patient's known history of malignant PEComa is important when cytopathologists make the differential diagnosis for the mesenchymal neoplasm of salivary gland.

The majority of PEComas are considered benign but some have malignant behavior and metastasize to distant organs. Poor prognostic factors include tumor size > 5 - 8 cm, infiltrative growth pattern, high nuclear grade, > 1 mitotic figure / 50 HPF or atypical mitotic figures, coagulative cell necrosis [2], which should be evaluated in surgical excised specimen. Excision is usually curative if tumors are benign. Everolimus/Afinitor® [4] is an inhibitor of Mammalian Target of Rapamycin (mTOR) which has shown significant clinical efficacy. Our patient elected against parotidectomy at this time because the significant decrease of the size of metastatic PEComa under Everolimus/Afinitor® treatment.

It is important to include PEComas in the differential diagnoses of spindle mesenchymal neoplasm of salivary gland. Schwannoma were the most common benign mesenchymal neoplasm in salivary gland. Plexiform neurofibroma, hemangioma, desmoid tumor, and solitary fibrous tumor have been reported in salivary gland. The malignant tumors consisted of dermatofibrosarcoma protuberans, synovial sarcoma, leiomyosarcoma, pleomorphic liposarcoma and desmoplastic small round cell tumor. But pre-operative imaging study and fine needle aspiration cytology had limitations in prediction of the mesenchymal nature of the tumors, due to either low index of suspicion, similarities to mixed tumors, or specimen inadequacy [5-8].

The differential diagnosis in cytological evaluation should also include several epithelioid neoplasm's, especially clear cell Neuroendocrinetumors (NETs), metastatic clear cell Renal Cell Carcinoma (RCC), melanoma, Alveolar Soft Tissue Sarcoma(ASPS), and metastatic epithelioid Gastrointestinal Stromal Tumors (GIST), although they are all extremely rare in salivary glands but case reports have been published in literatures. [9-13] Immunohistochemical studies are critical in these scenarios.

In conclusion, a preoperative diagnosis of PEComa on fine needle aspiration could be challenging, especially in uncommon sites such as salivary glands. Familiarity with the cytologic features and immunostaining profiles of PEComas may improve the diagnostic workup.

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