Hemangiopericytoma of the Parotid: A Case Report

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ABSTRACT

Hemangiopericytoma of the Parotid is a rare soft tissue sarcoma of vascular origin derived from pericytes, which are small spindle cells surrounding capillaries. Hemangiopericytomas are rare neoplasms that can occur in any area of the body, and between 15 and 25% of hemangiopericytomas are known to occur in the head and neck region. However, primary presentation of hemangiopericytoma in the parotid is exceedingly rare.

INTRODUCTION

Hemangiopericytomas were first described in 1942 by Stout and Murray as “vascular tumors arising from Zimmerman’s pericytes.” Pericytes are the small spindle cells that surround capillaries to provide mechanical support and regulate the luminal diameter of capillaries.

PATHOLOGY

Gross examination revealed a tan-white circumscribed nodular mass measuring 2.0 x 2.0 x 1.5 cm.

Microscopically, sections showed a cellular spindle cell tumor with areas of fibrosis and hyalinized vessels, lined by endothelial cells, some with a stag horn appearance. There was 1 mitosis per 10 high power fields. Tumor cells stained strongly positive for CD34, FLI-1, focally positive calponin, and CD68, and stained negative for desmin, smooth muscle actin, S100, and keratin mix.

Based on the histopathologic features of the resected mass, the diagnosis of hemangiopericytoma was made. Considering the low number of mitotic figures and the small tumor size, it could be further characterized as a low grade tumor.

CLINICAL PRESENTATION

A 28 year old woman presented with a growing non-tender mass at the right parotid for one year following pregnancy. The patient was a smoker with no history of malignancy, radiation, chemotherapy, or TB. She denied facial pain, numbness, or paralysis. Examination revealed a soft to rubbery 2.5 cm right preauricular mass with the right 7th cranial nerve appearing intact.

CT SCAN

CT scan of the neck w/o contrast revealed a 26mm x 11mm x 25mm well-defined, lobulated, homogeneously enhancing mass in the most superior aspect of the right parotid gland at the level of the temporomandibular joint without evidence of calcifications. There was no evidence of bony destruction or remodeling, and no other masses were identified. Biopsy was recommended.

FNA BIOPSY

Local anesthesia was used to facilitate 10 ultrasound-guided fine needle aspiration. The tissue was difficult to aspirate yielding slightly bloody tissue. Both alcohol fixed and air-dried smears were prepared. Smears were mildly cellular showing few small moderately cellular aggregates of spindle to epitheloid cells with small and uniform cell nuclei. Scant dense hypocellular stroma was present. The aspirate did not appear fibrillar and contained scattered lymphoid cells in some areas. There were no giant cells or caseous necrosis. The cell button showed blood and a few groups of slightly plump, short spindle cells. A diagnosis of low-grade spindle cell lesion was made.

SURGICAL RESECTION

A right superficial parotidectomy was performed with the goal to preserve all branches of the facial nerve. The mass was resected successfully from the superficial parotid. The tumor was found to have fibrous attachments to the underlying tissue and to the TMJ which were addressed. All branches of the facial nerve were intact post-operatively except the frontal branch which showed a 2/6 nerve palsy.

FOLLOW UP

Following surgery, the patient was not treated with any adjunctive therapy.

At three month follow-up, all signs of facial nerve palsy in the frontal branch had resolved. A CT scan of the head/neck and chest showed no evidence of recurrence or metastasis. Currently, the patient is at 18 months following treatment and shows no sign of recurrence or metastasis at this time.

DISCUSSION

The clinical presentation of this case of hemangiopericytoma of the parotid was consistent with that described in the literature. Hemangiopericytoma is often a slow-growing, insidious tumor that is non-tender until late in the course of the disease when symptoms arise from pressure on adjacent structures. As a result, patients often wait years to seek treatment.

The literature reports the immunohistochemical profile of hemangiopericytoma can assist with a diagnosis of this neoplasm. Tumor cells typically stain positive for CD34 as was consistent with this case. Also, while normal pericytes stain positive for desmin and smooth muscle actin, hemangiopericytoma cells stain negative as was also demonstrated by this tumor.

While this case was diagnosed as a low grade neoplasm, it is often difficult to predict the biological behavior of this neoplasm based on its histologic features. The disease has been known to reoccur years following complete resection even in low grade tumors. As a result, long term follow-up is required with radioimaging of the head and neck to detect recurrence and the chest to detect metastasis, which commonly occurs to the lungs.

Surgical resection remains the standard treatment for hemangiopericytoma. Adjunctive therapy continues to be evaluated. Prior case reports have shown that use of radiation therapy in the case of high grade, invasive disease has successfully reduced rates of recurrence while chemotherapy continues to be evaluated in the case of adult hemangiopericytoma.

REFERENCES