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Technical Note

Hemangiopericytoma of neck extending to craniovertebral junction treated by surgery, pre- and postoperative radiotherapy

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Abstract

Background: Hemangiopericytoma (HPC) is a rare tumor of uncertain malignant potential arising from mesenchymal cells with pericytic differentiation. It accounts for 3-5% of soft tissue sarcomas, and 1% of vascular tumors. The treatment of choice is a primary wide surgical resection with adjuvant radiotherapy (RT) reserved for cases of incomplete removal.

Case Description: We report a case of a 24-year-old female with a rapidly growing, highly vascular swelling in nape of the neck extending deep into the craniovertebral (CV) junction accompanied by extradural/intraspinal, and intracranial involvement. An incisional biopsy revealed a cellular, highly vascular tumor with HPC-like features. The patient received preoperative RT, which reduced both the size and vascularity of the lesion, facilitating subsequent near complete resection. Further postoperative RT resulted in a good clinical outcome, with no tumor recurrence observed at 2 postoperative years.

Conclusion: HPC of the soft tissues of neck accompanied by deep extension to the CV junction is uncommon. A high index of suspicion is required to diagnose these cases. which may be treated with preoperative RT (to reduce the lesion size/vascularity), aggressive surgical resection, followed by postoperative adjunctive radiation treatment as well.

Key Words: Craniovertebral junction, hemangiopericytoma, nape of the neck, preoperative radiotherapy, wide surgical resection

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INTRODUCTION

Hemangiopericytoma (HPC) is a rare tumor of uncertain malignant potential arising from mesenchymal cells that exhibits pericytic differentiation. Just above 300 cases of HPC have been reported since Stout and Murray described HPCs as "vascular tumors arising from Zimmerman's pericytes" in 1942.[5] Primary wide surgical resection is the treatment of choice and is typically accompanied by adjuvant postoperative radiotherapy (RT) if tumor removal has been incomplete. Here we present a case in which HPC was treated with subtotal resection accompanied by both preoperative and postoperative RT.

CASE REPORT

A 24-year-old female presented with a 1 year history of a painful, large, irregular, lobular mass rapidly growing at the

nape of the neck, accompanied by decreased sensation/numbness in the C1-C2 dermatomes [Figure 1]. Computed tomography (CT) scans of both the head and neck revealed a moderately enhancing 10-12 cm lesion (in greatest diameter) extending from skull base to the thyroid gland. It additionally involved the posterior cervical muscles, eroded through the posterior arch of atlas and occipital bone, where it exhibited both extradural/intraspinal, and intracranial extension [Figure 2]. An incisional biopsy documented a highly vascular lesion consistent with a cellular variant of a solitary fibrous tumor (SFT) with HPC-like features. Preoperative RT was administered to reduce the size and vascularity of the tumor, and was followed by aggressive subtotal resection of an encapsulated, soft, vascular tumor. Notably, radical resection was limited by the tumor's depth and vascularity. Further postoperative RT resulted in a good clinical outcome.

Histopathology revealed a vascular neoplasm characterized by sheets/groups of spindle/oval cells with a stag horn pattern consistent with HPC located in between plentiful of thin-walled vessels [Figure 3]. On immunohistochemistry, consistent with the diagnosis of HPC, the tumor cells were positive for both cluster of differentiation 34 (CD34) [Figure 4a] and vimentin [Figure 4b]. The postoperative CT scan documented only a minimal amount of residual tumor (near the atlanto-occipital junction) with near complete decompression of the craniovertebral (CV) junction [Figure 5]. Two years postoperatively, the patient remains asymptomatic and disease free (e.g., no recurrence or metastases).

DISCUSSION

Epidemiology

HPC is a rare, soft-tissue tumor of vascular origin, found in the head and neck in less than 20% of cases. In the neck, it typically affects the soft tissues, but rarely is accompanied by deep extension involving the posterior skull base, and eroding arch of the atlas or occipital bone.

Pathology

HPC is typically a well-circumscribed, brown, spongiform soft, firm, or friable lesion, that can be lobular or nodular surrounded by a pseudo-capsule, that is firmly attached to muscle or fascia.[2] Immunohistochemically, the tumor cells stain positively for CD34. Additionally, vimentin (type III intermediate filament protein that is expressed in mesenchymal cells) is the only marker expressed consistently in HPC.[2,3]

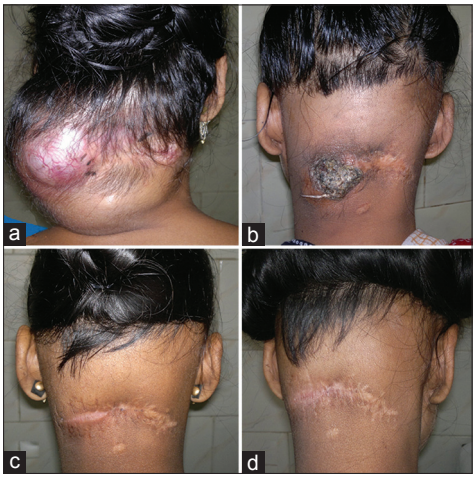

Figure 1: Comparative clinical photograph of the patient. At the time of presentation (a), after preoperative radiotherapy (b), after surgery (c), and after postoperative radiotherapy (d)

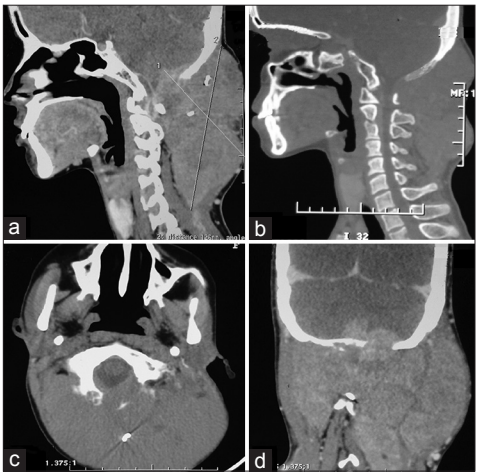

Figure 2: Computed tomography scan of head and neck sagittal (a), axial (c), and coronal (d) view showing moderately enhancing soft tissue lesion in the nape of neck, extending from skull base to thyroid gland level. Bone window (b) shows erosion of the posterior arch of atlas and occipital bone

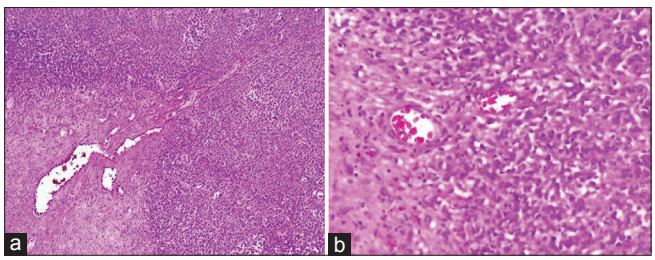

Figure 3: Photomicroscopy of the excised tumor showing vascular neoplasm consisted of sheets and groups of spindle and oval shaped cells (pericytes) in between thin wall blood vessels in some areas with stag horn pattern (a-low power, b-high power view)

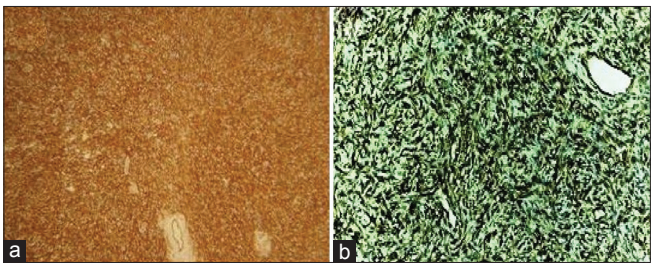

Figure 4: Immunohistochemistry demonstrating CD34 (a) and Vimentin (b) positive tumor cells

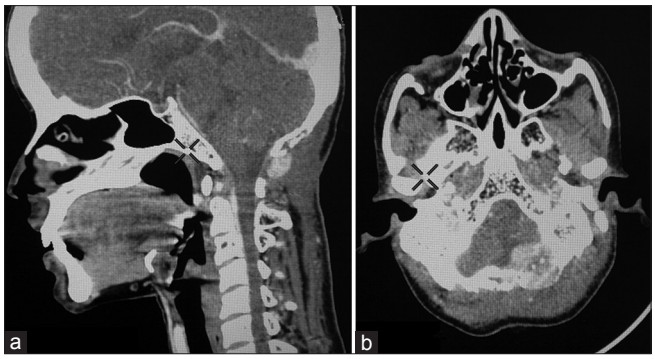

Figure 5: Computed tomography scan done at 2 month, sagittal (a) and axial (b) view demonstrating near total decompression of the craniovertebral junction with minimal residual tumor

Diagnosis and treatment

Preoperative evaluation of HPC should include both CT and magnetic resonance imaging (if available). Radiographically, the tumor is usually a well-circumscribed, radiopaque soft tissue, very vascular mass that displaces surrounding structures.[3] Angiography and preoperative embolization may be performed to address the larger, and more hypervascular lesions. Although the treatment of choice is typically primary complete resection (e.g., with a wide safety), where incomplete resection, local recurrence, metastasis, or an inoperable tumor are encountered, postoperative adjuvant RT is administered.[1,4] In this case, RT was utilized preoperatively to shrink/devascularize a massive lesion thereby facilitating subtotal resection, which additionally necessitated postoperative RT. Two years postoperatively, this aggressive management strategy successfully eradicated the tumor.

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