EJMO

DOI: 10.14744/ejmo.2018.0035
EJMO 2019;3(1):73-75

Case Report

Hemangiopericytoma at the Craniovertebral Junction: A Case Report

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Abstract

Intradural extramedullary hemangiopericytomas (HPCs) are extremely rare and a hemangiopericytoma located at the craniovertebral junction might present radiologic features similar to those of meningioma or schwannoma. To the best of our knowledge, this report is among a few reported cases of HPC at the craniovertebral junction. Although they are very rare, HPCs should kept in mind in the differential diagnosis of the intradural extramedullary lesion due to differences in treatment such as adjuvant radiotherapy.

Keywords: Craniovertebral junction, hemangiopericytoma, histopathology

Cite This Article: *Delen E, Akinci A, Turkkan G, Yalta T, Simsek O. Hemangiopericytoma at the Craniovertebral Junction: A Case Report. EJMO 2019;3(1):73-75.*

Hemangiopericytomas (HPCs), which are premalignant lesions, most commonly occur in subcutaneous tissues and skeletal muscles. They are rarely involved in central nervous system and account for only <1% of all primary central nervous system tumors.[1] Most of these tumors are located in the cranium, and an intradural extramedullary location of HPC is extremely rare. Radiological findings of HPC may be similar, even identical to a meningioma or a schwannoma. As the main treatment, strategy is similar for both tumors and consists of surgical resection that radiological similarity may not create a management problem at the first stage. On the other hand, a histological diagnosis of HPC requires a sure total resection and close follow-up, due to the high recurrence rates. Post-operative radiotherapy is suggested for patients with histopathologically diagnosed anaplasia (the World Health Organization Grade III tumors), subtotal resection, or adjacent surgical margins. We are presenting HPC case at the craniovertebral junction, which was treated with a total resection and adjuvant radiotherapy.

Case Report

A 30-year-old woman was admitted to the neurosurgery department with suboccipital headache whose course was over 3 months. We learned from her history that she had undergone an excision of osteochondroma from the tibia 10 years ago. On physical examination, she had no abnormalities. The neurologic examination was also intact. A magnetic resonance imaging (MRI) revealed an intradural extramedullary tumor compressing the spinal cord at the craniovertebral junction. The lesion was isointense on T1-weighted sequences and highly enhancing homogeneously (Fig. 1). With a presumed diagnosis of meningioma or schwannoma, a surgical total resection was planned. Perioperative period was uneventful. Post-operative MRI confirmed a total resection of the tumor (Fig. 2). The histopathological examination showed high proliferation, positive staining for cluster of differentiation 34, and negative staining for glial fibrillary acidic protein (Fig. 3). These findings were consistent with the World Health Organiza-

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Submitted Date: August 15, 2018 **Accepted Date:** November 08, 2018 **Available Online Date:** January 02, 2019

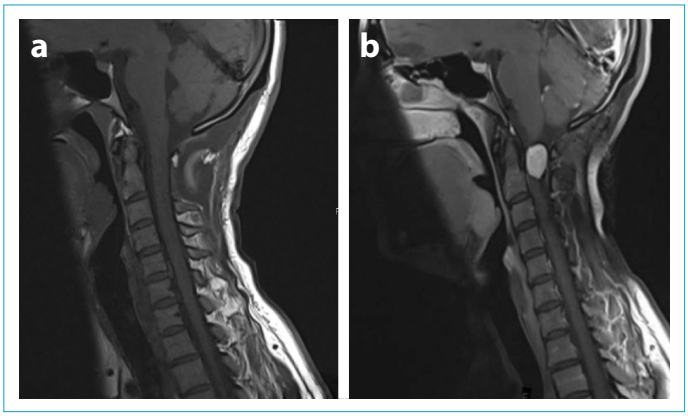

Figure 1 (a, b). Sagittal T1 **(a)** and contrast-enhanced T1 weighted, **(b)** magnetic resonance imaging showed an intradural extramedullary tumor at the craniovertebral junction.

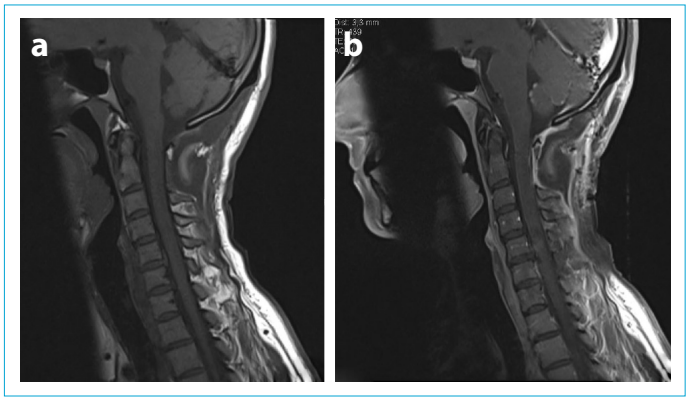

Figure 2 (a, b). (a) Sagittal T1 and contrast-enhanced T1 weighted, **(b)** magnetic resonance imaging confirmed a total resection.

tion Grade II hemangiopericytoma. After she had definitive diagnosis through histopathological examination, a positron emission tomography was performed. Results showed no metastasis. Fluorodeoxyglucose uptakes on lymph nodes were commented to be reactive. Due to the adjacent surgical margin, adjuvant radiotherapy was suggested to the patient to improve local control. A total dose of 50.4 Gray (1.8 Gray/fraction daily and five fractions per week) of radiotherapy was postoperatively administered to the tumor bed. At the follow-up after 6 months, she had no neurological deficits.

Discussion

In the past, hemangiopericytoma had been called hemangioendothelioma as a variant of meningioma. Subsequently, Stout and Murray termed them as hemangiopericytoma.[2] 9 years later, Stout reported that these tumors should be named solitary fibrous tumor.[3] In the following years, the debate on whether these tumors should have been termed as HPC or solitary fibrous tumors has continued. Eventually, the World Health Organization reported that these tumors were similar, should be termed as solitary fibrous tumors/HPC, and should be classified as one entity in 2016.[4]

These tumors are uncommon vascular neoplasms originating from pericapillary cells also known as the pericytes of Zimmermann.[5] They are most commonly located in lower extremities, retroperitoneum, head, or neck.[6] They only account for <1 % of all primary central nervous system tumors. When the spinal cord is involved, they are frequently reported in the thoracic spine.[7]

Before and after intravenous administration of gadolinium-based contrast material, MRI is the most useful tool in

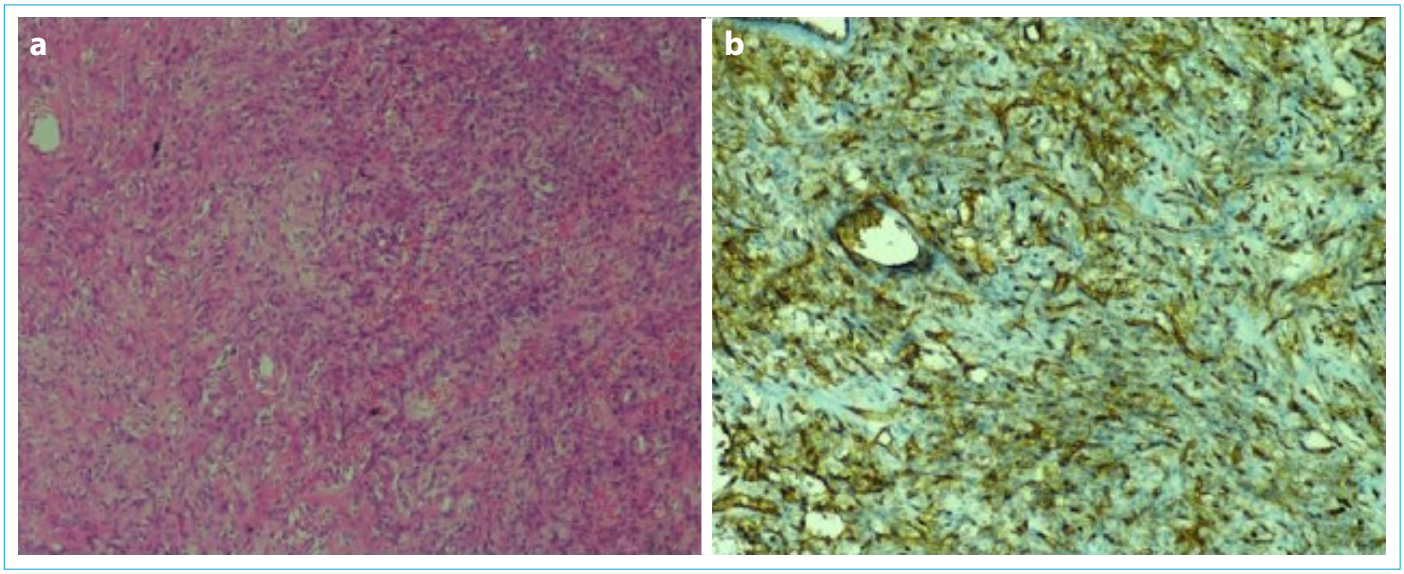

Figure 3 (a, b). Hematoxylin and eosin stained. **(a)** Section showed moderate cellular proliferation (H and E, ×200), **(b)** showing a weak staining for CD34.

the diagnosis of these tumors. HPCs usually have homogeneous, low-to-intermediate signal intensity relative to muscles on both T1-weighted imaging and T2-weighted imaging, as well as a very intense enhancement. The radiological appearance of our case was quiet similar to meningioma. It was an isointense lesion on T1-weighted sequences, and it was highly enhancing homogeneously after gadolinium injection. Therefore, the differential diagnosis of spinal hemangiopericytoma includes tumors such as meningiomas, schwannomas, neuroblastomas, and neurofibromas.[8]

Surgery is the mainstay treatment for central nervous system HPCs. Adjuvant radiotherapy might be an effective supplement postoperatively to reduce recurrences, but chemotherapy is disappointing.[9] In the presence of subtotal resection, adjacent surgical margin or histopathologically diagnosed anaplasia (the World Health Organization Grade III tumors) post-operative radiotherapy has been suggested. A dose of 50–60 Gray of radiotherapy is suggested to improve local control. In our case, 50.4 Gray of radiotherapy was administered to the tumor bed by taking into consideration the maximal tolerance radiation dose of the spinal cord.

Conclusion

In this report, we discussed a rare case of primary spinal HPC for contribution to literature. To the best of our knowledge, this report is among a few reported cases of HPC at the craniovertebral junction.[10] Although they are very rare, HPCs should kept in mind in the differential diagnosis of the intradural extramedullary lesion at the craniovertebral junction. A short interval follow-up is crucial due to the high incidence of local recurrences and to the possibility of metastasis.

Disclosures

This research did not receive any specific grant from funding agencies in the public, commercial, or not-for-profit sectors.

This case is presented at the "Joint Meeting of Istanbul Spine Masters and ISMISS Turkey 2017" as a "poster Presentation."

The authors declare that they have no relevant or material financial interests that relate to the research described in this paper.

Informed consent: Written informed consent was obtained from the patient for the publication of the case report and the accompanying images.

Peer-review: Externally peer-reviewed.

Conflict of Interest: None declared.

Authorship Contributions: Concept – E.D., A.T.A., G.T., T.D.Y., O.S.; Design – E.D., A.T.A., G.T., T.D.Y., O.S.; Supervision – T.D.Y., O.S.; Materials – E.D., A.T.A., G.T., T.D.Y.; Data collection &/or processing – E.D., A.T.A., G.T.; Analysis and/or interpretation – E.D., A.T.A., G.T., T.D.Y., O.S.; Literature search – E.D., A.T.A.; Writing – E.D., A.T.A., G.T., T.D.Y., O.S.; Critical review – E.D., A.T.A., G.T., T.D.Y., O.S.

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