Epithelioid hemangioendothelioma of the craniocervical junction; case report and review

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Abstract
Epithelioid hemangioendotheliomas are uncommon vascular neoplasms and their spinal location is even rarer. We report clinical course of a 31-year-old man with an epithelioid hemangioendothelioma at the craniocervical junction. A cervical magnetic resonance imaging revealed tumor that caused posterior cervical cord compression. C1,2,3 total laminectomy and surgical excision of the tumor was performed. Postoperative external beam radiation was performed on the surgical field especially around the right vertebral artery. At 2-year follow-up there was no neurological deficit and no tumor recurrence.

Introduction
Epithelioid hemangioendotheliomas (EHEs) are one of the angiocentric vascular neoplasms with inherent metastatic potential. EHEs are one of the least common diagnosed but comparably more malignant of the vascular neoplasms that arise from the lungs, liver, and other organs.1 In recent years, there are not many reported cases of EHEs in the spinal region; therefore because of the rare incident, our knowledge about this disease is more limited.2–6 Malignancy behavior of EHEs vary between hemangiomas and conventional angiosarcomas.7 There is no standard widely accepted procedure for the treatment of this tumor. Surgical excision with negative resection margins followed by external beam irradiation is commonly used as first option. There have been no reported cases in the literature of EHEs in the cranio-cervical junction. We report here the unusual case of an EHE seen in the cranio-cervical region with the single symptom of pain in the neck, which is treated by surgical resection followed by external beam radiation in cranio-cervical region.

Case report
31-year-old man presented with neck pain and restriction of neck movement due to pain present ongoing for a year which got worsened in the last 6 months. Physical examination showed a solid fixed mass at the cranio-cervical junction extending to C4 spinous process. The patient was neurologically intact with no radicular pain or muscle weakness. Blood runs showed no abnormalities. MR imaging revealed low signal intensity lesions on both the T1 and T2-weighted images (Fig. 1). Lesion showed little enhancement after Gadolinium injection (Figs. 2 and 3). Cervical computed tomography (CT) showed no destruction or invasion of the bone structures (Fig. 4). EHE was verified histopathologically after CT guided biopsy from the right quadrant that has been performed under local anesthesia.

Operation
Surgery was planned for the total excision of the lesion under general anesthesia. The patient was prone position with the head fixed in place the Mayfield headholder. Intraoperative MEP (Motor Evoked Potential) and EMG (electromyography) neuromonitoring had been used to avoid possible neurological deficits that may develop during the operation. Midline vertical incision from linea nuchae of the occipital bone to the level of C7 corpus was performed. Anterior part of the tumoral mass was adherent to all of the laminar processes of the C1,2,3 as well as it was extended from the C1–2 and C2–3 interlaminar spaces to the epidural space. C1,2 and C3 total laminectomy was performed and the tumor was resected with the attached part of the dura. A residue tumor attached to the vertebral artery was remained. Then using polyaxial screw/rod...
instrumentation posterior C0–C4 fusion stabilization was performed. Defect of the dura mater was sutured water-proof by using synthetic dural graft. Continuous lumbar drainage catheter had been applied preoperatively to the patient in order to prevent possible CSF fistula postoperatively. One week after the operation, the surgical wound was healed with no evidence of CSF leakage through the sutured wound edges and the lumbar CSF drainage was terminated. After 10 days of hospitalization, the patient was discharged sans any complications. Pathological examination of the surgical excision material confirmed the diagnosis as EHE (Fig. 8). Patolojik After the wound healing, patient was consulted with the radiation oncology department and a total of 5000 rads of irradiation therapy was planned and applied.

Discussion

The most common of the vascular tumors involving the spine are hemangiomas, which are benign in nature and most cases are not need to be treated. EHE is a rare disease of unknown etiology for which a standard systemic treatment has not been accepted. EHE is an angiocentric vascular tumor with metastatic potential. In the past, the concept of hemangioendothelioma included various kinds of benign, intermediate, and malignant vascular lesions. According to the new WHO classification of tumors of the soft tissue
and the bone, which was published in 2006, there has been some
effort to classify most hemangioendotheliomas into intermediate
malignant tumors, in which, a low-grade tumor was rarely associ-
ated with distant metastasis. Even the distant metastasis rate of
EHEs are not as frequent as the angiosarcomas. It is now under-
stood that 20–30% of EHEs presented distant metastases and
10–20% of these patients the EHEs were the primary cause of the
mortality. Such features correlate EHEs with an aggressive course.
Malignancy behavior of the EHEs are classified intermediate be-
tween hemangiomas and conventional angiosarcomas so it is
referred to as a low-grade epithelioid angiosarcoma.

Primary EHEs of the spine are rare. There were only a handful of
large series articles of the spinal EHEs, most probably due to the
rarity of this disease; hence, our knowledge of the disease is still
limited. To the best of our knowledge, it is not yet reported an EHE
in such localization. The objective of the current study was to
describe the clinical presentation and outcome of the surgical
treatment for this patient.

EHEs typically occur in the 20–40 age range with no gender
predilection, although the overall age range involved is much
broader. It has an indolent course, some have survived for decades
with multi-organ disease. In our case the patient was a 31 years
old male. Chronic, progressive pain in the neck or back was the
most consistent complaint. This atypical symptoms appearing before the diagnosis ranged from 3 months to 1 year, which averaged about half a year. Sometimes, spinal EHEs may cause severe symptoms due to cord compression.

Radiographic findings of the EHEs vary and are not specific to the disease. On the plain X-ray image, lytic process with solitary or multiple lesions has no diagnostic modality, due to the similarity with the other vascular tumors. Although computed tomography scanning or MRI scans detected bone destruction, the scale of the involvement and the evidence of the cord compression caused by the tumor, both had not enough diagnostic differentiation for the prebiopsy diagnosis. MRI scan is the gold standard for diagnosing spinal tumors. MRI imaging of the EHE in our report showed a mass with low signal intensity on T1-weighted images and high signal intensity on T2-weighted images. In our case there was no evidence of destruction of the bone structures on CT scan. MRI revealed low signal intensity on T1 and T2-weighted images and homogenous contrast enhancement was detected on contrast-enhanced T1-weighted images.

Due to inherently being a vascular tumor, intraoperative blood loss is a critical problem during the surgical treatments for the spinal EHEs. Therefore, preoperative angiographic evaluation and selective embolization of the involved vertebrae were suggested if possible. Sybert et al reported a case of spinal hemangioendothelioma which did not go through embolization preoperatively, and the operation had to be terminated due to perfuse bleeding. Although in our case preoperative embolization wasn’t applied and there was no severe bleeding problem.

Due to the rarity of the disease there is no accepted standard procedure for the treatment of this tumor. Resection is still the commonly preferred treatment for these tumors. According to some reports, the results of solitary external beam irradiation treatment were acceptable. However, we believe that as an aggressive tumor, the treatment of the spinal EHEs need a more aggressive approach including surgical excision with negative resection margins followed by external beam irradiation. The morbidity and outcome of the resection of a paraspinal soft tissue tumor mostly depends on the anatomic relationships of the neoplasm to the bone and neural structures. In our case, because of the existence of the tumor’s strict adherence to the right vertebral artery, we consulted the patient with radiation oncology and planned adjuvant radiotherapy. No instability of the spine and no complications presented in the patient. A significant relief of the neck pain was obtained following the removal of the lesion and decompression of the vertebral canal. We were able to achieve a radical resection but only leaving a small piece of residual tumor that attached to the vertebral artery. At the 2-year follow-up, there was no evidence of any tumor recurrence (Figs. 5–7).

Conclusion

We believe an en-bloc resection should be performed whenever possible in spinal EHEs. On the other hand, postoperative radiotherapy can be effective for the prevention of recurrence, in cases that could not be resected due to any strict adhesions or invasions to the adjacent neural and vascular structures. As in the presented case, valid surgical resection of the lesions with additional external beam irradiation may have presented acceptable results; however, our follow-up is too short to offer a viable outcome and the patient will need continued surveillance.

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