

Spinal Hemangiopericytoma with Liver Metastasis

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Abstract

Hemangiopericytoma is a very rare and aggressive tumor that originates from pericytes of Zimmermann. Spinal hemangiopericytoma is a rare neoplasm that behaves similar to intracranial hemangiopericytoma, with approximately 140 cases being reported in the literature. We report a case of recurrent hemangiopericytoma of dorsal spine in a 55-year-old women treated with surgery for 3 times, with liver metastasis. Gross total resection, if possible, is the first choice of treatment. Radiation therapy or chemotherapy may be indicated, especially in cases with high-grade lesions or tumors with unresectable.

Keywords: Spinal tumor; Hemangiopericytoma; Metastasis

Introduction

Hemangiopericytomas (HPC's) are rare vascular tumors. They arise from Zimmermann's pericytes, contractile spindle cells that surround capillaries and venules [1,2]. They correspond to 1% of all vascular and 15 to 25% of head tumors, being rarely found in the central nervous system [3-6]. The spinal HPC's are even rarer and may involve the structures of the vertebrae and the spinal canal, including epidural and intradural space. In a literature review the authors found about 140 cases previously reported.

Case Report

Patient referred to the neurosurgery service for evaluation, with historic of surgery for spinal intradural extramedullary tumor at T9-T10



Figure 1: Sagittal T1W MRI shows an intradural extramedullary lesion at T2 level with intense gadolinium enhancement.



Figure 2: Abdominal CT scan shows a liver metastasis of HPC affecting left and right lobes.

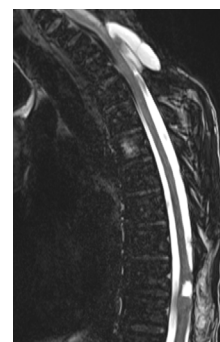


Figure 3: Sagittal T2W MRI shows an intradural extramedullary lesion at T2 and T9-T10 levels.

level, presenting paraparesis. She hadn't a histopathological diagnosis so far. Admission tests showed spinal intradural extramedullary tumor at T2 level (Figure 1). It was performed surgical treatment by posterior approach with bilateral laminectomies, achieving gross total resection. The histopathological analysis confirmed hemangiopericytoma grade III. Patient remains neurologically stable after the procedure. During hospitalization it was diagnosed an abdominal mass. CT scan showed liver mass in the left lobe, suggestive of giant hemangioma or hemangioendoneuroblastoma, being referred for surgery, on which was performed a left hepatectomy plus right segmentectomy (Figure 2). Analysis of the lesion confirmed metastasis of HPC. The oncological evaluation defined not be required complementary therapy. A year later, the patient return for evaluation with new recurrence of the tumor at the level of T2 and T9-T10, now with intramedullary extension of the lesion on T9-T10 level (Figure 3), being submitted to surgical resection in 2 times, achieving a gross total resection and no new neurological deficit. Latest review with staff, one year after the last surgery, patient

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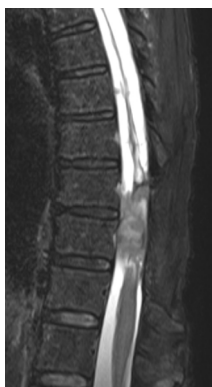


Figure 4: Sagittal T2W MRI shows an intradural extramedullary lesion at T9-T10 level.

continues with unchanged neurological deficits – paraparesis; but MRI revealed new recurrence at T9 level (Figure 4) and the patient is in surgical programming.

Discussion

Hemangiopericytomas were first described by Stout and Murray in 1942 [1]. Originated from vascular pericytes, these tumors may affect any part of the human body, but the most common sites are the extremities, pelvis, retroperitoneum and head and neck [4-8]. Spinal HPC's are very rare and mostly involve extradural bone structures. Primary intradural HPC was only reported in 10 cases, all occurring in extramedullary region [9].

The clinical presentation varies according to the tumor size and location, being the most common symptoms associated with pain and mass effect. These tumors can be locally aggressive, with high chance of local recurrence and spread. Local recurrence is a bad prognostic indicator and is associated with appearance of metastases, commonly to lungs and bones [10]. Thus, clinical findings, presence of metastasis, and histopathology should be evaluated simultaneously to predict the biological behavior of this tumor [11].

These tumors do not have specific radiological pattern, however their angiographic characteristics can help in the diagnosis. The appearances in arterial phase known as “spider-shaped” with a dense, well-defined lesion in venous phase are characteristic [11].

Surgical resection is the first choice of treatment in all cases when feasible, and a gross-total resection should be achieved in order to reduce recurrence ratios. Endovascular embolization is recommended

in preoperative time due to the pronounced tendency to bleeding during procedures. Radiation therapy or chemotherapy may be indicated, especially in cases of high-grade lesions, unresectable tumors or in cases of recurrence after surgery and radiotherapy [12].

The prognosis is poor, with high rates of local recurrence and metastasis, and low response to radiation and chemotherapy, which worsens the outcome [5-7].

Conclusion

Hemangiopericytomas are rare vascular tumors, with infrequent spinal cord involvement, being the primary intradural extramedullary subtype very rare. They are locally aggressive, with high incidence of local recurrence and metastasis; thus, the treatment may also be aggressive, with embolization and gross total resection in all cases whenever possible, despite the bad prognosis.

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