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Cervical spine epithelioid hemangioendothelioma: case report

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Abstract. – Epithelioid Hemangioendothelioma (EHE) is a rare aggressive vascular tumor and can occur in almost all locations. Spine location is rare. There are just a few cases published in the literature and the longer series included only five cases.

CASE REPORT: We report a case of a 58-year-old woman who presented with anemia and cervical pain without neurological deficit. Radiological diagnosis revealed a C2-C3 mass with vertebral artery involvement and liver metastases. Partial resection and arthrodesis followed by radiotherapy and chemotherapy were performed. Local control of the diseases was achieved but distance metastasis appears two years after surgery.

CONCLUSIONS: EHE of the spine is extremely rare in clinic as a primary aggressive vascular tumor. Metastatic disease is the most important factor to predict prognosis. It is not clear in the literature which is the best surgical approach in this patients and it mainly depends of the location and systemic illness. En bloc resection or expanded resection supported with adjunct radiation therapy may present with acceptable results of local control of the tumor. Systemic disease control must be achieved with chemotherapy.

Key Words:
Epithelioid hemangioendothelioma, Cervical spine tumor.

Introduction

Epithelioid Hemangioendothelioma (EHE), previously known as angiolymphoid hyperplasia with eosinophilia or histiocytoid hemangioma, is a rare vascular tumor that has been described to have an intermediate biological behavior between hemangioma and angiosarcoma. It has a reported local recurrence rate of 11% and metastasizes in 2.7% of cases[1,2]. It can occur in almost all location but is mainly described in soft tissues.

Primary bone presentation accounts for less than 1% of malignant bone tumors and is mainly described in long bones of the lower extremities with approximately 50% of them being multifocal[1,3-6]. Spinal involvement is even more unusual and few cases have been reported, making it unclear which is the ideal surgical treatment and the role adjuvant radio- and/or chemotherapy may have. Here, we report a case of a cervical EHE that underwent surgical treatment with subsequent radio- and chemotherapy.

Case report

A 58-year old woman was referred to emergency room for ongoing headache with partial response to analgesics and a recent onset of constitutional symptoms (asthenia and weight loss). In light of an iron deficiency anemia detected on the blood analysis an endoscopy and colonoscopy were performed with no findings. Since neither the anemia nor asthenia responded to medical treatment, further studies were deemed necessary.

An abdominal CT scan showed several small masses in the liver's parenchyma suggestive of metastatic lesions. A bone scintigraphy was then performed with no new findings. PET-CT described a hypermetabolic osteolytic mass at the first second and third cervical vertebrae.

Cervical CT scan showed an important lytic lesion affecting C1 anterior arch, C2 and C3 vertebral bodies with posterior wall displacement occupying the spinal canal and left lateral masses/facet processes of C2 and C3. No pathological fractures were identified (Figure 1). MRI showed an expansive infiltrative lesion with moderate compression of the neural root of C2-C3 and spinal cord. Angiography showed displacement and moderate stenosis of the left vertebrae artery at the cervical region.

Liver and C2 vertebrae biopsies were performed and the pathological analysis reported the presence of cells with elongated cytoplasm and occasional atypia compatible with a metastatic epithelioid hemangioendothelioma.

Clinically the patient was neurologically intact. Due to such findings and the lack of medullar

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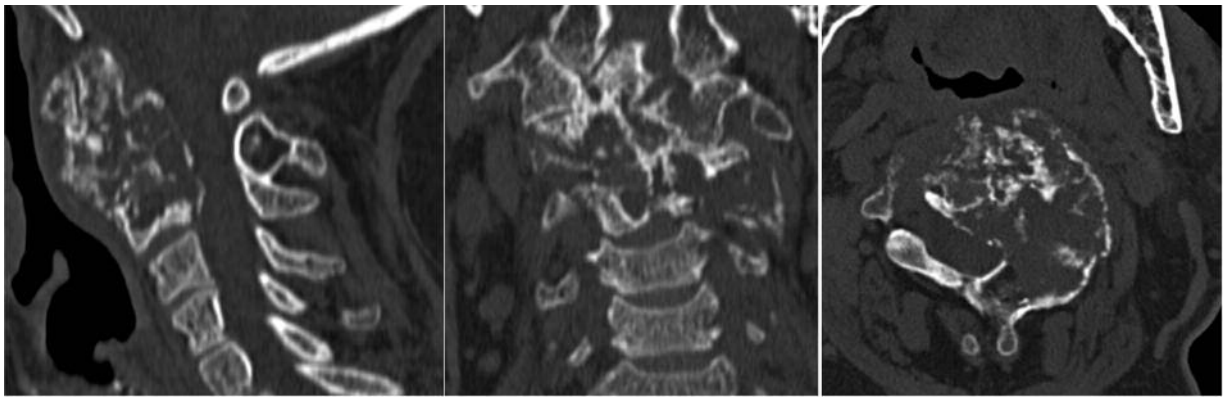

Figure 1. Cervical CT Scan showing lytic lesion of C1, C2 and C3 vertebrae.

symptoms, the patient was immobilized with a Halo-Jacket to prevent pathological fracture while careful surgical planning was made. Due to the extension of the lesion, localization and the presence of distance metastasis we considered that en bloc resection was not possible to achieve and a debulking surgery due to posterior approach was performed after embolization of the mass. A C2 left laminectomy and partial mass excision and curettage was conducted through left lateral mass and pedicle of C2-C3. During the excision a small tear occurred on the left vertebrae artery which was successfully sealed with an hemosthatic matrix. Remaining cavity was filled with vertebroplastic cement. After that a posterior occipito-cervical stabilization with an occipital V-shaped plate and bilateral mass screws from C3 to C6 was implemented. There were no intraoperatory complications. Immobilization was maintained with Halo Jacket. Histopathological examination of the excised mass confirmed the diagnosis of epithelioid hemangioendothelioma.

During postoperative period, the patient referred paresthesia and functional motor deficit on left C5-C6 territory with no particular findings on CT and MRI with complete recovery six months after surgery.

3 months after surgery the Halo Jacket was removed. She then underwent chemotherapy with Paclitaxel and local radiotherapy which had to be suspended for a period of two months due to radiation induced mucositis. After this, 5 cycles of Adriamycin were administered.

On subsequent MRI controls remaining tumor mass on anterior arch of C1 and on vertebral body of C2 and C3 did not show neither progression nor spinal cord compression. Two years after surgery CT scan shows complete fusion of posterior arthrodesis and no local progression of tumor (Figures 2 and 3). MRI documented small

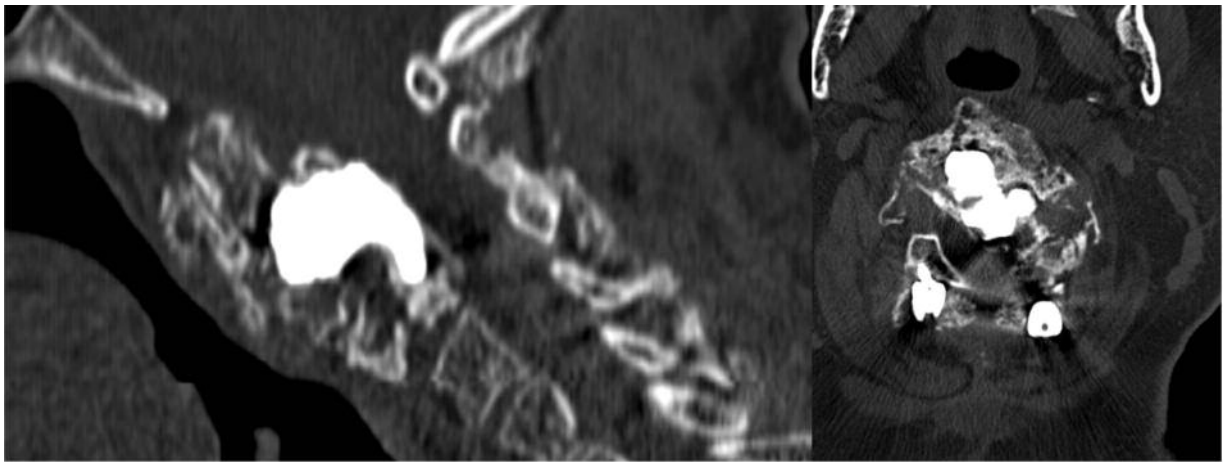

Figure 2. 2 year follow-up cervical CT scan. Partial resection of tumor with cement and posterior stabilization.

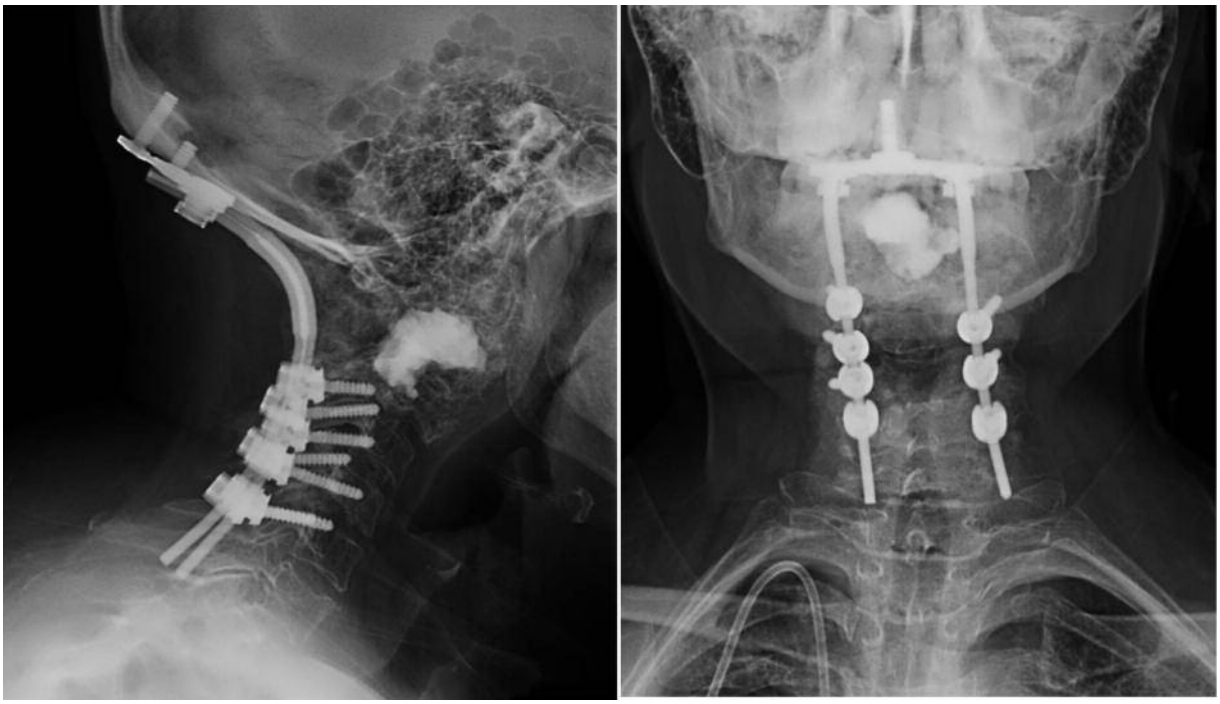

Figure 3. 2 year follow-up radiological control. Posterior instrumented stabilization and fusion.

cerebellum lesions, probably related to the tumor, causing instability during walking. No other neurological deficits were identified on clinical examination and some degree of neck pain well controlled with pain killers remains. At the final follow up (30 months) the patient is alive with local and systemin disease (AWD) and only palliative therapy are now possible.

Discussion

This rare vascular tumor was first described in 1982 by Weiss and Enzinger and is considered to have a highly variable outcome depending on its location and histological characteristics[7,8]. Malignancy is classifies into three categories depending on the degree of vasoformative activity, atypia of the endothelial cells and frequency of mitotic activity[3,8]. This tumor is usually composed of small chords of eosinophilic cells and irregular anastomosing vascular channels[7,8]. There seems to be no gender nor age predilection, even though some authors describe it as more common in males[1,5,6,8].

Clinical presentation varies depending on the organ of involvement. When arising in bone, local pain has been described as the most common presenting symptom[1,5]. Local mass compression and pathological fractures have also been described in approximately 10% of cases. When involving vertebrae, radicular symptoms or paraplegia may occur. In multifocal bone EHE, visceral involvement is deemed as the most important criteria in predicting prognosis[6,8].

Radiological findings are non-specific and differential diagnosis must include ostelytic sarcomas, lymphomas, giant cell tumor, bone cyst and metastasis. An expansive osteolytic lesion with distinctive soap-bubble matrix with a sclerotic margin and no periosteal reaction is how it is usually described. Lesions with marked loss of trabeculae and ill-defined margins are considered as more aggressive[1,9].

The CT-scan, though non diagnostic, may help differentiating from hemangioma and sketch the extent of bone destruction. MRI findings are also nonspecific, and as in most vascular tumors, T1 sequences show higher intensity than muscles but less than fat; in T2 sequences signal intensity is considerably higher than both muscle and fat[1]. EHE has been described as doughnut-like lesions in radionuclide imaging and it is believed that such shape is due to peripheral osteablastic activity with a central hemorrhagic or lytic region[3].

There is no consensus on the management of this tumor but it strictly depends on the lesion's aggressiveness grade and can include en bloc re-

section (if feasible) or debulking followed by radio and/or chemotherapy.

Radiotherapy seems beneficial in surgically inaccessible tumors and as adjuvant treatment after surgical resection. On the other hand, there is no precise indication for chemotherapy in such lesions, even though it is described as beneficial in patients with liver involvement[1,6,8,10].

Conclusions

Bone epithelioid hemangioendothelioma is a rare primary vascular bone tumor which is usually described in long bones. Spinal involvement is quite rare and very few cases are described in the literature. In the present case, diagnosis was made due to unspecific symptoms spawned by the primary's tumor metastasis in the liver. Cervical mass was not initially detected and once studied its extension made complete surgical excision impossible. Surgical stabilization, after tumor debulking was clearly needed due to the lesion's localization. Radiotherapy was administered for the residual tumor. Chemotherapy was also used, mainly due to the hepatic lesions. After a two-year follow-up, the patient presents neither new neurological symptoms nor objective growth of remaining lesions in imaging techniques. Sadly, prognosis is not too good due to the hepatic lesions which have not entirely responded to chemotherapy and the patient is currently receiving palliative treatment.

Conflict of Interest

The Authors declare that there are no conflicts of interest.

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