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NOTA CLÍNICA

Primary spinal epitheliod angiosarcoma

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KEYWORDS
Angiosarcoma;
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Abstract
Primary bone angiosarcoma is rare, and is only seen in 1%of all sarcomas. These tumours are more often found in the long bones. Primary spinal involvement has been reported in 10%of angiosarcomas.
As a result of its rarity, its pathological, clinical and therapeutic characterists are largely unknown.
We presnt the case of a patient with a primary vertebral epithelioid angiosarcoma in D8 which initially presented as dorsal pain and paraparesis, with the aim of contributing to a better knowlwedge of this rare process.

PALABRAS CLAVE
Angiosarcoma;
Epitelioide sarcoma;
Quimioterapia

Angiosarcoma epitelioide vertebral primario

Resumen
El angiosarcoma óseo primario es una rara eventualidad que representa, tan solo, el 1% de todos los angiosarcomas. Estos tumores asientan con mayor frecuencia en huesos largos. La afectación vertebral primaria se ha comunicado en un 10%de angiosarcomas.
Como resultado de esta rareza se desconocen, en gran medida, sus características patológicas, clínicas y terapéuticas.
Presentamos el caso de una paciente con angiosarcoma epitelioide vertebral primario en D8 que debutó con dolor dorsal y paraparesia con el objetivo de intentar contribuir a un mejor conocimiento de este raro proceso.

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Introduction

Bone and soft tissue sarcomas represent a highly heterogeneous group of tumours insofar as their anatomical location, histological subtypes, and clinical behaviour are concerned.[1]

Angiosarcomas comprise less than 1%of all sarcomas and bone angiosarcomas are a rare disease and account for fewer than 1%of all angiosarcomas. They are preferentially located in the long bones of the limbs, although, in 10%of the cases, they may be found in the vertebrae.[2]

In general and due to the local extension of the process, surgical removal is impracticable or incomplete; hence, it entails a poor prognosis.

We present the case of a primary vertebral epithelial sarcoma with the aim of contributing to a better understanding of the disease.

Case Report

Fifty-six year old female with a personal history of surgery for a herniated lumbar disc. She was admitted to hospital in April, 2008, after referral from another centre for evaluation by the Spinal Surgery Unit at the Traumatology Department.

She reported that for the past two weeks she had been suffering dorsal pain without prior trauma that was accompanied by paraparesis in the lower limbs.

The physical examination revealed the following: normal cranial pairs, diminished strength in the iliac psoas, quadriceps, and tibial muscles; plantar reflexes in flexion.

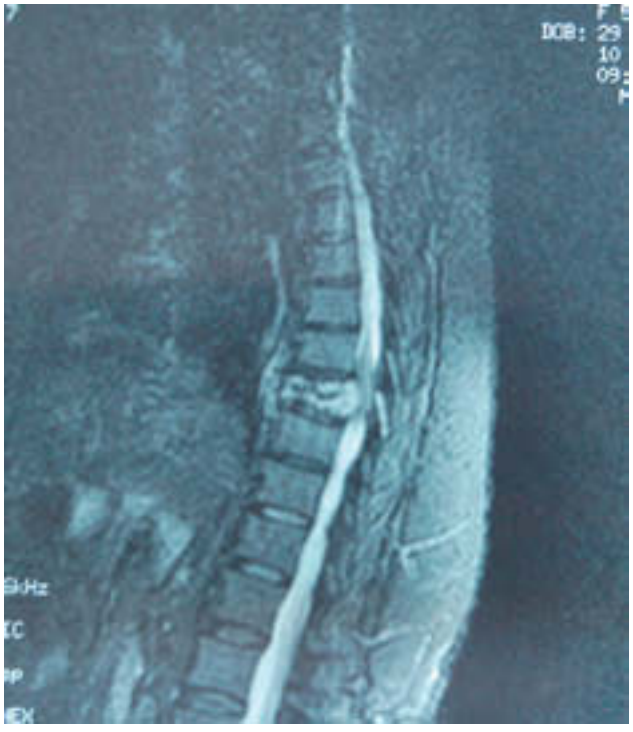

Figure 1 Proliferation of cells endothelial epithelial with large, eosinophil cytoplasm. H&E 20×.

Analyses failed to reveal any data of interest.

An x-ray was taken of the dorsal spine: compression and complete destruction of D8. The nuclear magnetic resonance (NMR) image revealed collapse and compression of D8 with paravertebral mass invading the spinal canal and shifting the spinal cord, contrast-enhanced (fig. 1).

The bone gammagraphy only showed enhanced uptake in D8. The CT scan of the abdomen with contrast was normal. The chest CT showed: extensive lytic lesion in the vertebral body and left pedicle of D8, with a soft tissue component at the level of the left paraspinal and toward the spinal canal.

With the diagnosis of pathological fracture of tumour origin and secondary spinal compromise, the treatment chosen was surgery consisting of decompression with complete laminectomy of D8, widened to the lower half of D7 and upper half of D9 in addition to stabilization by means of a D5 to D11 transpedicular system.

The pathology reported the following: endothelial epithelial cell proliferation with eosinophil cytoplasm and large vesicular nuclei with a solid, vessel-forming pattern.

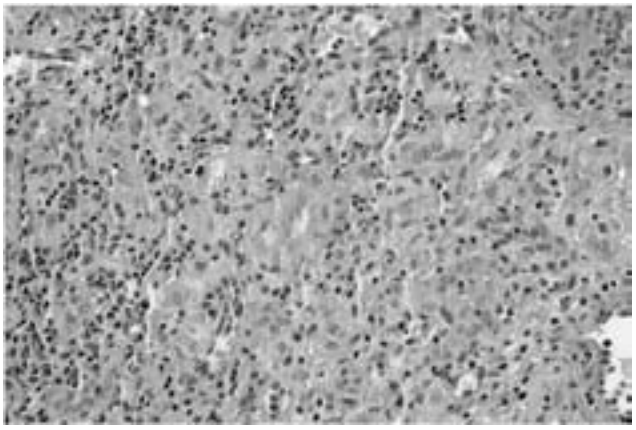

Figure 2 Proliferation of endothelial epithelial cells with large, eosinophil cytoplasm. Vesicular nuclei with eosinophilic nucleoles. H&E 20×.

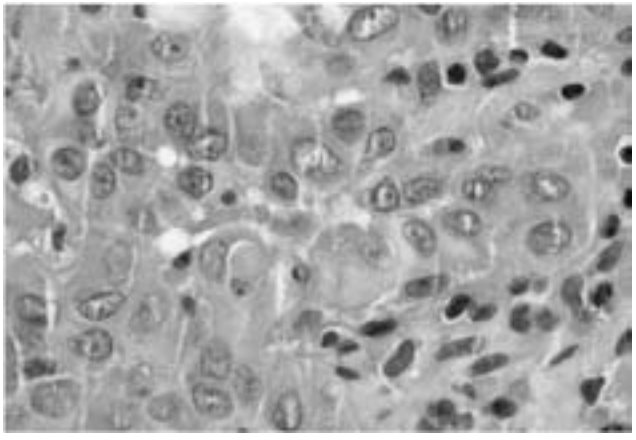

Figure 3 Detailed view of large vesicular nuclei with eosinophilic nucleoles. H&E 60×.

Areas of necrosis and bleeding (figs 2 and 3). The tumour cells expressed vimentin, CD 31, CD 34, and, to a lesser extent, factor VIII. The proliferation index with Ki-67 was moderate (30%) and the diagnosis reached was grade II epithelioid angiosarcoma.

During the immediate post-operative period, the patient presented neurological deterioration and paraplegia.

Given the broad decompression performed, as well as the appropriate functioning of the drains, the clinical picture was not attributed to a problem of compression; hence, it was decided that a conservative approach would be used.

The NMR image revealed an expansive lesion of the posterior wall of D8, in addition to the post-surgical changes. A low T1 signal collection and intermediate T2 signal were observed between the posterior slope of the canal and the surgical field, without contrast uptake, with collapse of the spinal cord at this level between the aforementioned collection and the expansive lesion dependent on the vertebral body.

The radiotherapy department decided to abstain from treatment as they believed paraplegia to be established.

Insofar as our department was concerned and after obtaining informed consent, we administered 3 cycles of palliative monochemotherapy with adriamycin without any kind of response; hence, the patient was sent to the palliative care unit for on-going care. Her last visit to this unit took place in August, 2008, four months following surgery.

Discussion

Angiosarcoma is a vascular tumour made up of mesenchymal cells that tend toward angioblastic differentiation with the formation of new vessels; aggressive, in general, when located in bone, they account for less than 1% of all primitive bone tumours.[2] Up until 2004, only 16 cases of vertebral angiosarcoma had been reported.

Its aetiology is unknown, involving both genetic, as well as environmental factors, without any relevant factors having been detected in the present case.

The median age at diagnosis is between 60 and 71 years, although our patient was somewhat younger. There was a slightly greater proportion of males versus females in the largest series reported (112 cases).[2]

It can present in two different forms: multifocal or, as in the case we present, as a single, rapidly growing lesion with a tendency to produce distant metastasis.

From a pathological point of view, the epithelioid variety presents poor differentiation and aggressive biology. The first report of this variety located in bone was made by Hasegawa et al.[3] in 1997. It entails a worse prognosis than other vascular tumours and may be associated with bleeding diathesis by producing fibrinolytic compounds, a complication to be taken into account during post-operative periods.[4] In our case, in addition to the local extension of the tumour, it is possible that there had been post-operative haematoma and that this may have contributed to the irreversible spinal compression.

The diagnosis of the disease is established by ruling out other primary tumours, given the presence of the tumour mass in the vertebra and by the pathology or immunohistochemical analysis. In the case we report here, we excluded the presence of another primary tumour by bone gammagraphy and CT; the NMR image shed light on the diagnosis, as did the typical histology of the epithelioid variety, and the characteristic immunohistochemical findings.

On the other hand, the involvement of the epithelioid variety can look very much like carcinoma metastatic involvement, requiring immunohistochemical analysis for differential diagnosis. As in the case that currently concerns us, it was positive for endothelial markers, with CD-31 being the most sensitive.[5]

The following prognostic factors have been described: size of 5 cm or more, high grade, positive surgical margins, locations hindering complete removal, and advanced age; several of these factors were present in our case.

Treatment in localized disease consists of surgical resection, often followed by radiotherapy, which may be successful.[6] In the present case, the patient debuted with symptomatology of compressive spinal injury and at the time of surgery, the disease was ASIA stage D. The ensuing paraplegia was not attributed to compressive origin due to the major decompression performed, and the proper functioning of the surgical drains. The oncological radiology department did not feel it wise to proceed with radiotherapy as they were of the opinion that the symptomatology was irreversible.

Had the patient not presented with such an acute neurological picture, the attitude might have been different and an angiography might have been performed to embolize the tumour prior to surgery and application of the Tokuhashi score following the extension study.

On the other hand, adjuvant chemotherapy (ACT) is recommended fairly frequently; the most widely used agents are adriamycin and iphosphamide, alone or in combination.[7]

For advanced disease, these same drugs and others, such as paclitaxel or liposomal doxorubicin, achieve good response rates and improved progression-free survival (PFS) with acceptable toxicities.[8] More recently, anti-angiogenic drugs alone or combined with traditional cytostatic drugs are being incorporated into the treatment arsenal.[9]

In the case presented, we opted to use adriamycin in monotherapy, as it is considered to be the standard first-line drug, obtaining a 25% response rate.[10] Although combining adriamycin with iphosphamide increases response rates, it fails to improve survival and is more toxic; as a result we chose to use this cytotoxic. After three cycles without achieving any objective response, we suspended its use.

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