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Rare primary vertebral epithelioid angiosarcoma: a case report

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Background
Primary malignant vascular tumors of bone are very rare. The epithelioid angiosarcoma is a high-grade sarcoma that is extremely rare in spine location and presents unpredictable clinical course. Frequently it is delayed or misdiagnosis, and presents a high metastatic rate and poor prognosis. In literature, metastatic bone angiosarcoma is invariably fatal.
The diagnosis is histological and immunohistochemical confirmation is important.
There are only few cases described of vertebral angiosarcoma. The main treatment is surgical wide en-bloc resection with or without adjuvants.

Case Report
A 68-years-old male presented insidious low back pain in association to motor and sensitivity deficits, neurological gait claudication and weight loss.
An osteolytic lesion at the third lumbar vertebral body was analysed and characterized.
The PET scan showed an intense uptake at L3 and a slight uptake at the joint of the sixth dorsal vertebra with the rib that presented no tumor characteristics at the MRI.
A Tru-cut needle biopsy CT-guided revealed inflammatory tissue at the sixth dorsal vertebra and was compatible with primary epithelioid angiosarcoma of L3.
It was a type 5 of Tomita spine bone tumors classification, with paravertebral extension.
Tumor resection with total vertebrectomy of L3 was performed posteriorly. The surgical reconstruction consisted in posterior pedicle screw instrumentation of L1-L2-L4-L5 and interposition of a titanium expandable cage filled with iliac bone allograft and added a cross-link.
The histopathology and immunohistochemistry of the resected tumor confirmed a high-grade epithelioid angiosarcoma.
The prognostic was extremely poor due to the paravertebral extension of this malignant aggressive tumor. He recovered sensitivity deficits but persisted paraparesia. Then, he presented a Pseudomonas Aeruginosa nosocomial sepsis with urinary tract starting point. The patient died within 1 month postoperatively.

Discussion
In literature, few bone epithelioid angiosarcomas are described and in spine location are extremely rare. They are a diagnostic and surgical challenge.
Although the total vertebrectomy performed the en-bloc resection was not possible due to paravertebral tumor extension. It showed an aggressive behaviour in association to morbidity and mortality.
Survival rate of these tumors is unknown but is essential an early diagnosis to allow a successful surgical wide resection.

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