EXTRA-AXIAL CHORDOMA IN POPLITEAL FOSSA

Chordoma is a rare malignant tumour, which arises typically in the axial bones from remnants of the embryonic notochord. Chordomas may develop outside the axial skeleton, but they are exceptional. We present an unusual case of extra-axial chordoma (EAC) in popliteal fossa.

CLINICAL CASE
Female 60 years-old, who complains about swelling, pain and growing mass in popliteal fossa of the left knee. The MRI reported the presence of encapsulated cystic lesion about 5cm near to popliteal vessels with small bone destruction of the lateral condyle surface.

We performed a core needle biopsy, and it revealed a tumour composed of large epithelioid or polygonal cells with clear or vacuolated cytoplasm and atypical vesicular nuclei. Immunostains for pan-keratin and epithelial membrane antigen (EMA) were diffusely positive but S-100 protein, inhibin, Melan-A and PAX-8 were negative. However, there was multifocal striking nuclear positivity for brachyury. In these circumstances, given the absence of any evident primary lesion elsewhere, these findings would fit best with an extra-axial chordoma (EAC).

The treatment was tumour resection and post-operative radiotherapy because of the marginal margin near the popliteal vessels. Eleven months after surgery, a 2.5cm soft tissue mass appeared on the posterior aspect of the external tibial plateau, close to external popliteal sciatic nerve. The imaging was highly suspicious for chordoma distal recurrence. The patient was operated and the pathological findings confirm the diagnosis of EAC. A CT scan showed no sign of systemic dissemination.

DISCUSSION AND CONCLUSIONS
A small number of tumors show histological resemblance to axial chordoma, but they arise from the bone or soft tissue outside the axial skeleton. Brachyury immunohistochemical staining is a sensitive and specific marker for notochordal origin, that allows more accuracy diagnosis for EAC distinguishing them from parachordoma, which resembles chordoma on histology. The distinction between EAC and parachordoma is clinically important because EAC confirmed by immunoreactivity for brachyury tends to grow and recur with local bone destruction.

The preferred treatment for patients with extra-axial chordoma is radical surgery, combined with radiotherapy in cases with marginal margins in tumour resection specimen. The role of chemotherapy is unclear. Survival, recurrence and rates of metastasis are unknown because of the rarity of this condition. However, it is known that the tumour may recur after many years (16% between 3-36 months after excision) and that it could metastasize (5%).

Keywords:

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