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Extra-axial Chordoma of bone and soft tissues: report of two case.

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Chordoma is a rare low-to-intermediate grade malignant bone tumor usually affecting the axial spine of adult patients that shows a distinctive immunophenotype (cytokeratins, S100 and brachyury).
Very few cases of chordoma-like lesions have been described in extra-axial sites where, despite their rarity, they pose problems of differential diagnosis with primary or metastatic epithelioid tumors of soft tissue and bone, in particular with the mixed tumor/myepithelioma/parachordoma family of tumors.
Here we described two cases of extra-axial soft-tissue chordomas.
The first case was a 39 year old woman with a history of swelling and pain of the right arm that, at radiological examination, showed a juxta-osseous mass of 5 cm involving the humerus shaft and consequently underwent mid-humerus resection. The second case was a 58 year old woman with symptoms of knee joint monoarthritis and synovial hyperplasia at the imaging investigations which underwent diagnostic biopsy of the synovial membrane. On histological evaluation both cases showed an epithelioid morphology with mixo-chondroid pattern and a phenotype consistent with chordoma, in particular cytokeratin 19 and brachyury positive reactivity were observed.
The latter is the first case described in intrarticular localization within the synovial membrane to the best of our knowledge. Remarkably both cases were characterized by co-expression of CK 19 and brachyury which is considered highly specific of classic chordoma.
The diagnosis of extra-axial chordoma can be challenging, especially in biopsy specimen, because of the rarity of this entity and the immunomorphological features largely overlapping with other tumors (i.e. epithelioid features, Cytokeratins, S100). However, the recognition of extra-axial chordoma is important given its prognostic and therapeutic implications which are different from those of the other entities that could be mistaken for it.

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