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Chordoma dedifferentiated to osteosarcoma: two consecutive cases and literature review

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Introduction: Chordoma (CHO) is a rare, slowly growing, malignant neoplasm that arise from embryonic notochordal remnants. It accounts for 17.5% primary malignant tumors of the axial skeleton, arising from sacrococcygeal region (50%) and mobile spine (15%).
In less than 5% of cases, CHO contain highly malignant sarcomatous component. Little is known of their clinical feature and treatment options.
Methods: A retrospective review, out of a prospective database, found 2 patients with diagnosis of "CHO dedifferentiated to osteosarcoma (OGS)". Both the patients were submitted to exhaustive studies, and computed tomography CT-guided biopsy was done. Histopathological and immunohistochemical strains were done on the all specimens.
Results: Two cases of spinal "CHO dedifferentiated to OGS" are reported. Both patients had a first biopsy diagnostic for CHO, atypical features on MRI, showing two different areas within the lesion: a reduced signal area and a hyperintense component on T2-weighted images. Histopathological analysis on repeated CT-guided biopsy specimen (case 1) and on en bloc excised gross specimen (case 2) was diagnostic for "CHO dedifferentiated to OGS".
In one case, adjuvant chemotherapy was given, followed by en bloc resection and carbon-ion therapy (case 1). Both patients are not evidence of disease (NED) at more than 30 months of follow-up.
Conclusion: Dedifferentiated CHO is distinguished from "CHO with sarcomatous transformation" because of a sharp demarcation of the sarcomatous elements and a lack of transitional features between a conventional CHO and a high-grade OGS component.
A literature review found 2 cases of "CHO dedifferentiated to OGS", both located in the sacrum. One was diagnosed as it in the primary lesion and local recurrence and metastases were composed exclusively of conventional CHO, while the other was CHO that showed metastases (pulmonary and brain) on appearance of OGS. Local control of primary tumors is important for survival in "CHO dedifferentiated to OGS". Pathological analysis remains the gold standard for definite diagnosis, and MRI and FDG-PET guided biopsy can be useful to obtain an adequate sample in suspect of a dedifferentiated disease.
Treatment of "CHO dedifferentiated to OGS" must include chemotherapy first, followed by en bloc resection or carbon-ion treatment.