Dedifferentiated skull base chordoma

Makise et al. identified a novel group of dedifferentiated chordoma of the skull base that lost H3K27me3/me2 only in the dedifferentiated component, which was associated with EED homozygous deletion and malignant peripheral nerve sheath tumor (MPNST)-like histology. This data suggest a distinct "polycomb-type" dedifferentiation pathway in chordoma, similar to a recently described dedifferentiated chondrosarcoma with H3K27me3 loss [1)].

A case of "dedifferentiated" skull base chordoma occurred in a 31-year-old man.Morphologically, the tumor was characterized by a biphasic pattern (classical chordoma associates with sarcoma-like areas), and by coexpression of epithelial and stromal markers. Because of these traits, we believe this case shows features superimposable to those seen in sarcomatoid carcinoma [2)].

A case of dedifferentiated chordoma arising from the skull base region of an 11-year-old boy, with tumor recurrence within one year. This tumor showed features of pleomorphic cell sarcoma with areas more typical of chordoma. Most tumor cells expressed cytokeratin, epithelial membrane antigen, vimentin, and S-100 protein, thus confirming the diagnosis of dedifferentiated chordoma [3)].

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