Teaching NeuroImages: Chordoma

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Figure 1 Imaging features of a chordoma

(A) Axial CT image demonstrates a central skull base lesion destroying the clivus (arrow) and left sphenopalatine fossa (arrowhead). (B) The lesion appears hyperintense on T2-weighted imaging (arrow) extending into the left sphenopalatine fossa (arrowhead). (C) Postcontrast coronal T1-weighted imaging demonstrates heterogeneous enhancement (arrow).

Figure 2 The thumb sign

(A, B) Sagittal MRI of the brain demonstrates a T2-weighted hyperintense skull base lesion (arrow, A) centered in the body of the sphenoid bone. The lesion causes osseous destruction and extends posteriorly into the prepontine and interpeduncular cisterns. Indentation of the ventral pons (arrowhead, A) results in the characteristic thumb sign (B).

A 30-year-old man presented with recurrent headaches. CT head revealed a clival chordoma (figure 1A). Chordomas originate from the embryonic remnants of the notochord and account for 2%–4%

of all malignant bone tumors. They have a predilection for the axial skeleton, with 35% affecting the spheno-occipital region. The incidence peaks at ages 20–40 years. Male patients are affected twice

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as commonly as female patients.¹ Clinical symptoms often result from local mass effect. On imaging, the tumor appears as a midline lesion and can contain heterogeneous calcifications. MRI demonstrates high signal on T2-weighted sequences and heterogeneous enhancement with a honeycomb appearance (figure 1).² Indenting of the pons results in the characteristic thumb sign (figure 2). Differential diagnosis based on imaging appearance includes chondrosarcoma and metastasis.

AUTHOR CONTRIBUTIONS

Dr. Azzopardi was responsible for conducting the literature review and drafting the manuscript and accepts responsibility for conduct of research, final approval, and acquisition of data. Dr. Grech accepts responsibility for conduct of research, final approval, acquisition of data, and study supervision and made the radiologic diagnosis.

Dr. Mizzi accepts responsibility for conduct of research, final approval, and study supervision.

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