
Neuroimaging Highlight

Editor: David Pelz

Chondrosarcoma of the Skull Base in Ollier's Disease

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Can J Neurol Sci. 2014; 41: 86-87

A 22-year-old female with Ollier's disease (OD) presented with a four-week history of diplopia on left gaze, attributable to a left sixth cranial nerve palsy. The remainder of the exam was normal. Her past surgical history was significant for many orthopedic procedures due to OD.

Brain magnetic resonance imaging (MRI) revealed a heterogeneously enhancing lesion within the left middle fossa, invading the cavernous sinus (Figure 1), with T2 signal hyperintensity. Computed tomography (CT) imaging of her chest, abdomen and pelvis demonstrated multiple enchondromas involving the pelvis (Figure 2).

Given her progressive neurological course and imaging findings, she underwent elective left pterional craniotomy. Intraoperative stereotactic image guidance and neurophysiology

monitoring methods were used. A subtotal resection was performed, given the intimacy between the lesion and neurovascular structures. The lesion was soft and gelatinous in consistency. She made a good postoperative recovery with moderate symptom improvement. On histology, the tumor was consistent with a WHO grade I chondrosarcoma. Radiation therapy will be arranged should there be a recurrence on future MRIs.

Ollier's disease, originally reported by Ollier in 1899, is a rare, sporadic, non-hereditary disorder of mesodermal dysplasia with an estimated prevalence of 1/1,000,000/year¹⁻². The classic features are multiple asymmetric, and unilateral enchondromas affecting the metaphyseal ends of long bones resulting in limb shortening³. Ollier's disease typically manifests between the ages

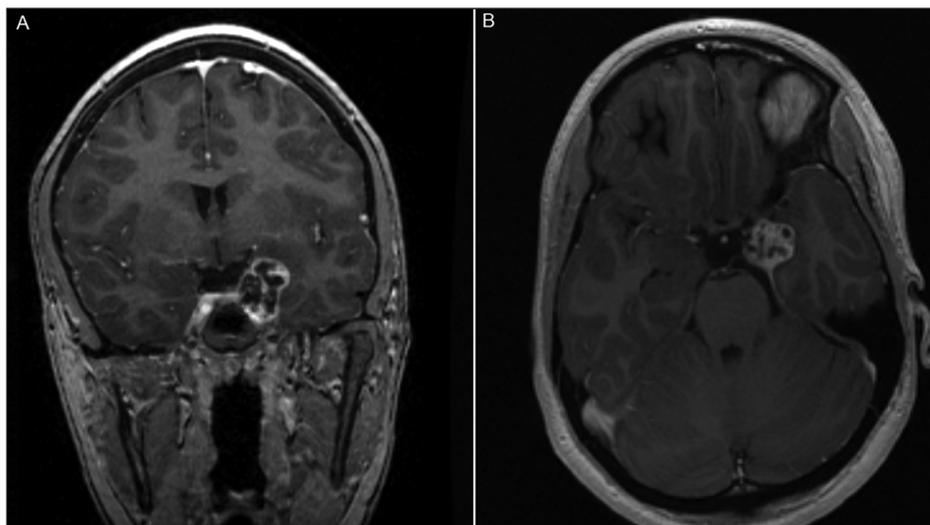


Figure 1: Coronal (A) and axial (B) enhanced T1-weighted magnetic resonance imaging scans showing lobulated mass (2.3 x 2.5 cm) within the left cavernous sinus with heterogenous enhancement.

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RECEIVED APRIL 16, 2013. FINAL REVISIONS SUBMITTED JULY 9, 2013.

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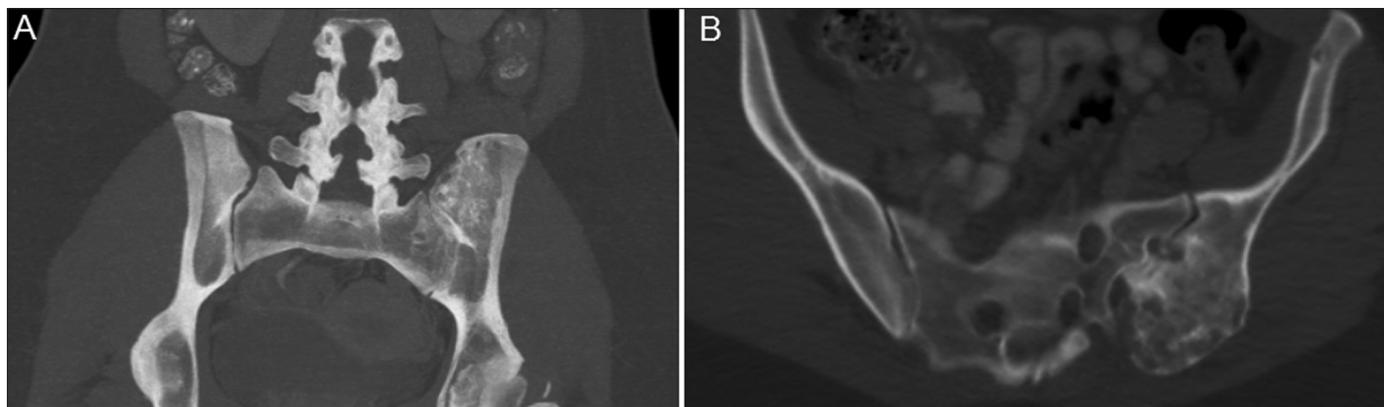


Figure 2: Coronal (A) and axial (B) plain computed tomography (CT) scans demonstrating classic features of enchondromatosis affecting bones of the pelvis.

of two and ten. While the pathophysiology of OD is not well understood, a mutation in isocitrate dehydrogenase 1 or 2 (IDH1/2) has been linked to OD⁴. Maffucci's syndrome also belongs to "enchondromatosis disorders", with the presence of soft tissue hemangiomas being the differentiating factor. Both syndromes are associated with a higher risk for developing brain tumors and hematological malignancies³.

Neurological involvement has been reported in several cases of OD³. The most common findings are related to cranial nerve palsies resulting in symptoms such as diplopia, hoarseness, dysphagia, facial dysesthesia, and hearing loss. The skull base, particularly the anterior and middle cranial fossa, is the common intracranial location for OD because the bones in this region form through endochondral ossification⁵.

Benign enchondroma and malignant chondrosarcoma are the two main differentials in skull base involvement in OD. Radiologically, enchondromas appear as hypo/hyperdense, well-demarcated lesions, with slight contrast enhancement on CT. On MRI, enchondromas are hypointense on T1 images, mixed hypo/hyperintense signal on T2 images with marked heterogeneous and honeycomb-like gadolinium enhancement. Strong enhancement is highly suggestive of chondrosarcomas due to rich blood supply⁶. Other lesions such as chordoma (usually midline) and meningioma (homogeneous enhancement) should be in the radiological differential diagnoses. The histological features of conventional malignant chondrosarcoma (increased cellularity and mitotic figures) cannot be applied in OD due to the hypercellular nature of their lesions; therefore, diagnosis depends on the combination of radiographical, clinical, and histological features².

Based on the unclear etiology of OD and the multiplicity of lesions, the management of this disease is challenging. The goal of skull base surgery in OD is safe gross total resection, which is often difficult given the delicate neurovascular anatomy of this region. Adjuvant radiotherapy is usually indicated in cases of chondrosarcomas because of their high recurrence rate³.

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