Radiology Case Reports

Volume 9, Issue 1, 2014

Hemorrhagic chondrosarcoma in a patient with Ollier disease: Case report and literature review

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We present a rare case of skull-base hemorrhagic chondrosarcoma in a patient with Ollier disease. Chondrosarcomas complicated by intracranial hemorrhage are very uncommon, with few reported cases in the literature. To our knowledge, this is the first such reported case.

Case report

The patient, a 19-year-old female previously diagnosed with Ollier disease, initially presented with a two- to three-month history of migraine-like headaches that were associated with nausea and vomiting. On admission, she had a decreased level of consciousness. Physical exam at the time revealed sixth- and seventh-nerve palsies and sensorineural hearing loss on the left side. An initial noncontrast computed tomography (CT) scan of the head showed an expansile mass lesion involving the left central skull base and petrous apex, with intracranial extension and curvilinear areas of intracranial hemorrhage. Severe mass effect was noted in the posterior fossa, with obscuration of the fourth ventricle and developing hydrocephalus (Fig. 1). Contrast-enhanced CT performed for stereotactic surgical guidance demonstrated mild contrast enhancement of the lesion (Fig. 2). The patient underwent a left retromastoid craniectomy for tumor debulking and hematoma evacuation with ventriculostomy catheter placement. She tolerated the procedure well, with no immediate complications. Pathology later confirmed the presence of a low-grade chondrosarcoma, World Health Organization (WHO) Grade 1 with

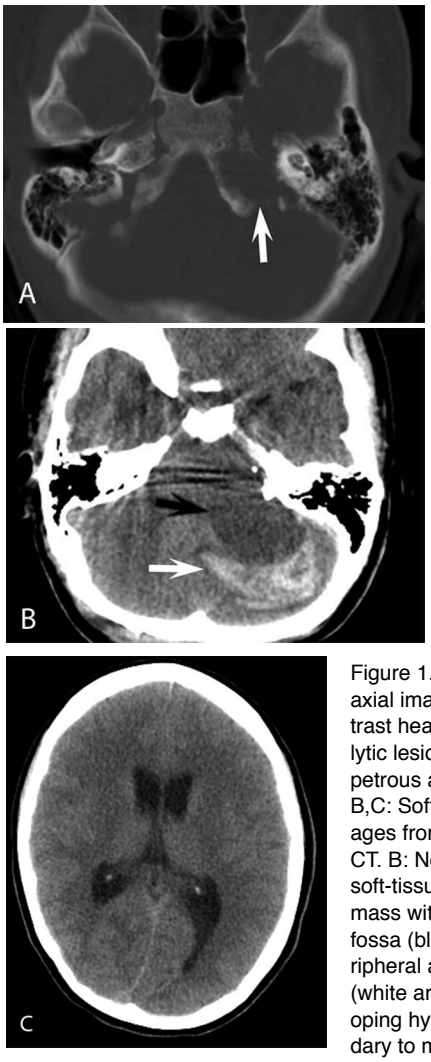

Figure 1. A. Bone-algorithm axial image from a noncontrast head CT demonstrates a lytic lesion within the left petrous apex (white arrow). B,C: Soft-algorithm axial images from a noncontrast head CT. B: Note low-attenuation, soft-tissue component of the mass within the left posterior fossa (black arrow), with peripheral areas of hemorrhage (white arrow). C: Note developing hydrocephalus secondary to mass effect on the fourth ventricle.

Citation: Hughes MA, Delfyett W, Gardner P, Arvanitis L, Rath T. Hemorrhagic chondrosarcoma in a patient with Ollier disease: Case report and literature review. *Radiology Case Reports*. (Online) 2014;1;889.

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Competing Interests: The authors have declared that no competing interests exist.

DOI: 10.2484/rcr.v9i1.889

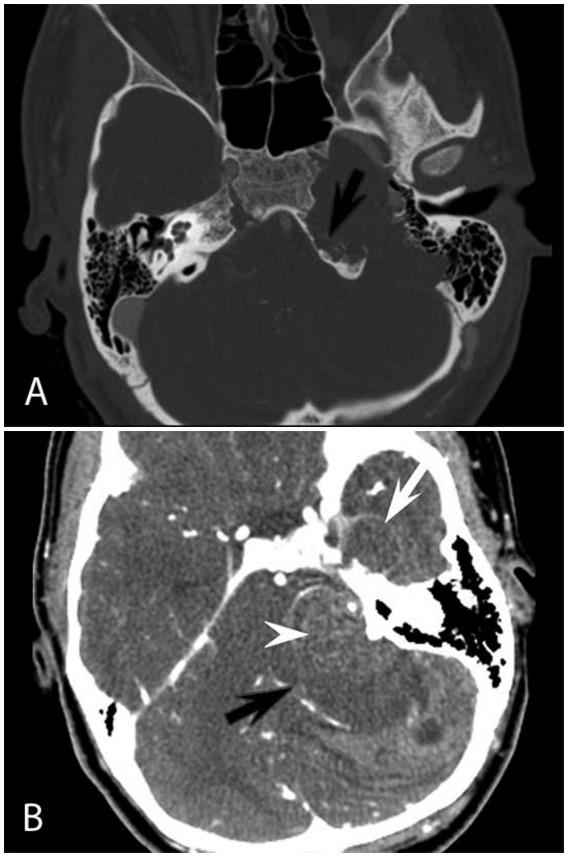

Figure 2. A. Bone-algorithm, axial CT image demonstrates the typical chondroid calcifications associated with chondrosarcomas (black arrow). B. Soft-tissue-algorithm, axial CT image post contrast demonstrates mild heterogeneous enhancement (white arrowhead) of the mass, with notable extension into both the middle cranial fossa (white arrow) and posterior fossa (black arrow).

extensive hemorrhage (Fig. 3). Subsequently, she underwent a transmastoid craniotomy and expanded endonasal approach anterior skull-base surgery for further tumor resection.

Discussion

Chondrosarcomas of the skull base are typically centered at the petro-clival synchondrosis. CT findings include expansile lytic lesions that may demonstrate the characteristic chondroid ring and arclike calcifications. Variable enhancement is noted by CT. Typical magnetic resonance (MR) findings include high T2 signal, intermediate T1 signal, and variable enhancement (1). Associated hemorrhage, as noted in this case, is a very uncommon finding, with four reported cases in the literature (2-5).

Harada et al reported a case of a 23-year-male with a clival chondrosarcoma and intratumoral and subarachnoid hemorrhage; the patient was successfully treated with a combination of surgical resection and gamma-knife radiosurgery (2). A petrous-bone chondrosarcoma with cerebellar invasion and associated hemorrhage in a 52-year-old female was described by Ohshige et al (3). The patient underwent multiple surgical resections and experienced at least three episodes of tumor bleeding, ultimately dying from complications of this tumor (3). Gallia et al reported a case of a paracavernous chondrosarcoma with intratumoral hemorrhage that extended into the adjacent temporal lobe and into the subarachnoid space; this patient was successfully treated with a combination of surgical resection and proton-beam therapy (4). Fukuchi et al reported a case

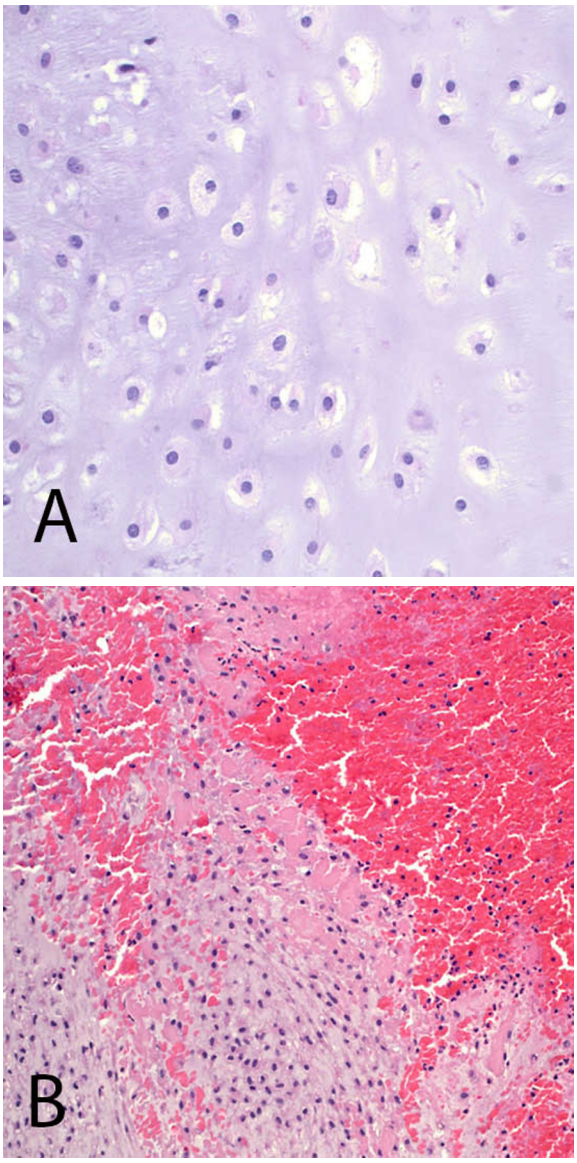

Figure 3. A. H&E section shows a conventional low-grade chondrosarcoma, WHO grade 1, with mildly atypical chondrocytes of various sizes and shapes in a prominent hyaline cartilage matrix (x400). B. In addition, there are extensive areas of acute and organizing hemorrhage with degenerating tumor cells (H&E, x200).

of a skull-base chondrosarcoma involving the petrous apex, clivus, and parasellar region in a 75-year-old female. Spontaneous intratumoral and peritumoral hemorrhage occurred, resulting in brainstem compression, respiratory arrest, and ultimately the patient's death (5). So, while skull-base chondrosarcomas are typically WHO grade 1 tumors and associated hemorrhage is exceedingly rare, when hemorrhage does occur it can be catastrophic, as two of the four previously reported cases reflect.

Enchondromas are typical benign intraosseous cartilaginous tumors. Ollier disease is characterized by multiple enchondromas, with an incidence of approximately 1 in 100,000 people (6). Malignant degeneration of enchondromas into chondrosarcomas is a known complication (7). The risk of malignant degeneration of an enchondroma into chondrosarcoma is increased in the setting of Ollier disease (8). Skull-base chondrosarcoma associated with hemorrhage is a very rare entity, with only four previously reported cases in the literature (2-5). To our knowledge, this is the first reported case of a hemorrhagic chondrosarcoma in a patient with Ollier disease.

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