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AJNR Am J Neuroradiol 1988, 9 (6) 1189-1193
http://www.ajnr.org/content/9/6/1189

Intracranial Dural Chondrosarcoma

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Three cases of intracranial dural chondrosarcoma are reported. The radiologic appearance of this slow-growing, extraaxial malignant tumor is different from the more familiar skull-base chondrosarcoma and may mimic an atypical meningioma. Dural chondrosarcoma tends to be less calcified or even to lack matrix calcification; it is associated with bone erosion but not with bone destruction or hyperostosis; and it usually appears avascular at arteriography.

Most cranial chondrosarcomas arise from the base of the skull, particularly in the parasellar region. There are only a few reported cases originating from the dura, and their radiologic appearance has not been described in detail. We report three cases of intracranial dural chondrosarcoma with radiologic description and differential diagnosis.

Case Reports

Case 1

A 32-year-old man presented with a diffuse headache of 3 months duration. He had a psychiatric history of progressive withdrawal and an attitude of indifference for several years. The neurologic examination was unremarkable except for bilateral papilledema. A skull series was unremarkable. A postcontrast CT scan revealed a large hypodense bifrontal mass attached to the falx with peripheral rim enhancement but no parenchymal edema (Fig. 1A). A few minute calcifications were noted within the tumor. The presumptive diagnosis was malignant glioma and an open biopsy was performed. The tissue was diagnostic of well-differentiated (grade-1) chondrosarcoma (Fig. 1B). After referral to our institution, a pre- and postcontrast CT scan confirmed the tumor to be largely cystic or necrotic, with the peripheral solid portion being hyperdense and enhancing (not shown). MR imaging (Figs. 1C and 1D) showed the tumor periphery to be hyperintense on both T1- and T2-weighted images. The central portion was hypointense on T1-weighted images and hyperintense on T2-weighted images. Cerebral arteriography revealed the tumor to be avascular and extraaxial. Total gross resection of a large gray-white, cystic, multilobulated mass originating from the falx was performed. The tumor was entirely subdural, without infiltration into the adjacent compressed frontal lobes. The histologic features of the neoplasm were again diagnostic of grade-1 chondrosarcoma. Dramatic improvement of mental status was noted in the immediate postoperative period.

Case 2

A 57-year-old woman presented with a history of tinnitus in the right ear of 2–3 years duration. Physical examination was unremarkable except for a right retrocochlear hearing loss. Skull radiographs were normal. Contrast CT showed a large dumbbell-shaped solidly enhancing extraaxial mass in the medial right middle cranial fossa extending over the petrous apex and into the posterior fossa (Figs. 2A and 2B). Minimal erosion of adjacent sphenoid bone was present, and a few flecks of calcification were evident within the tumor. MR revealed

Received January 27, 1988; accepted after revision April 21, 1988.

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AJNR **9:1189–1193, November/December 1988**
0195–6108/88/0906–1189

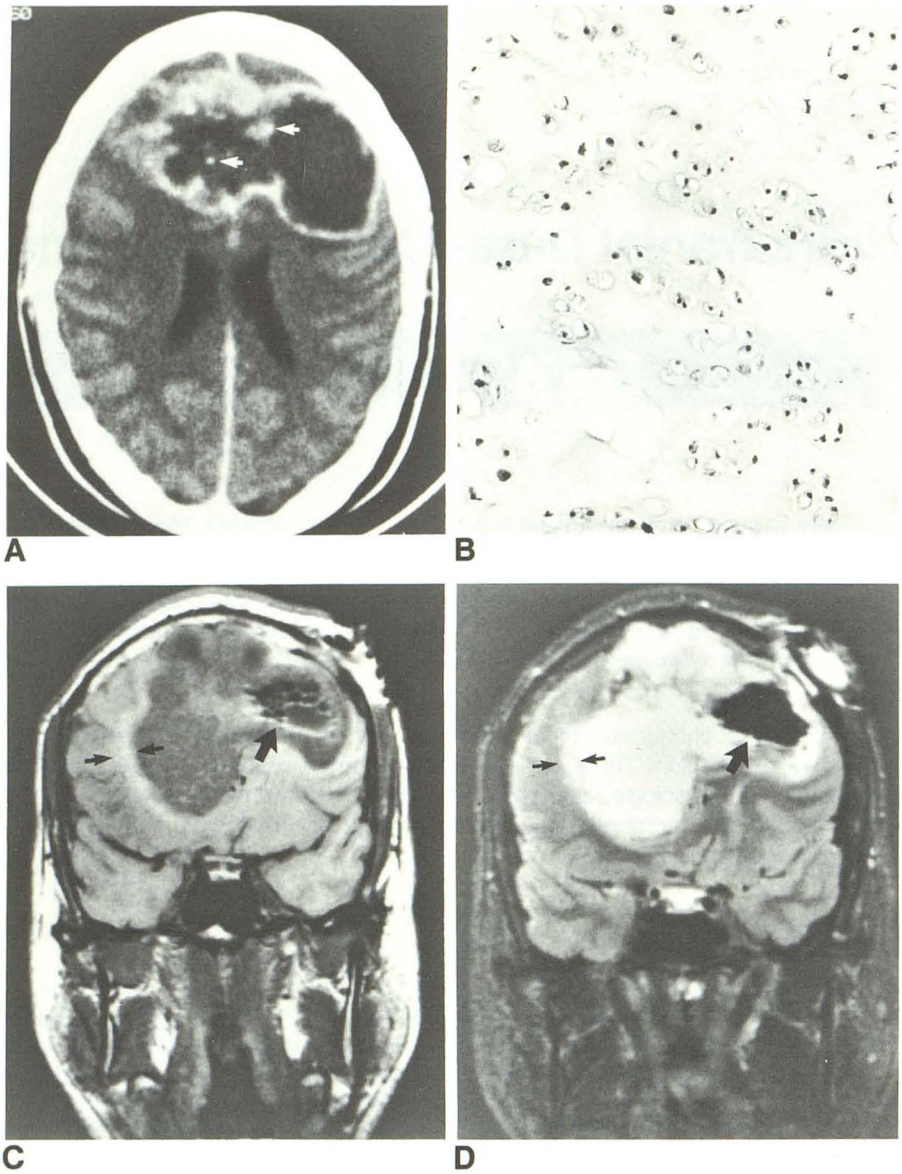

Fig. 1.—Case 1.

***A*, Contrast CT. Minute calcifications (*arrows*) within a large, cystic, rim-enhancing bifrontal mass with no appreciable parenchymal edema. Origin of tumor appears to have been in the anterior falx.**

***B*, Grade-I chondrosarcoma. There is minimal hypercellularity, some loss of lobular architecture, and some increase in nuclear/cytoplasmic size and detail. Occasional binuclear cells are present. (H and E, ×140)**

***C* and *D*, T1-weighted (800/25) (*C*) and T2-weighted (2000/80) (*D*) coronal MR images. Tumor rim (*small arrows*) appears hyperintense in both T1- and T2-weighted images. There is an intratumoral air collection after biopsy and partial drainage (*large arrows*).**

the tumor to be hypointense on T1-weighted images and hyperintense on the proton-density-weighted series (Figs. 2C and 2D). Arteriography was not performed. The working diagnosis was probable trigeminal neuroma. A transtemporal surgical approach was carried out, and a gray-tan subdural tumor was partially resected. The histologic diagnosis was chondrosarcoma, grade 2.

Case 3

A 41-year-old woman with Ollier disease gave a history of nonspecific headache and anosmia of several months duration, followed by diplopia for 3 weeks. She also reported the removal of a right fifth metatarsal cyst, which was diagnosed as enchondroma. Physical examination revealed a right sixth cranial nerve paralysis. The skull radioagraphs were unremarkable. CT showed a large, hypodense but solidly enhancing mass in the right middle cranial fossa, with minimal erosion of the lateral wall of the sphenoid sinus and cranial floor (Figs. 3A–3C). Cerebral arteriography showed the tumor to be avascular and extraaxial (Fig. 3D). Tentorial meningioma was the working diagnosis, although the external carotid vessels were normal. At surgery the tumor was found to be well encapsulated and to arise from the dura of the middle cranial fossa. The histologic features of the tumor were diagnostic of grade-2 chondrosarcoma. Further radiologic workup after craniotomy revealed two small enchondromas of the right scapula and a chondrosarcoma of the right ilium.

Discussion

Chondrosarcoma, a malignant but slowly growing cartilaginous tumor, constitutes approximately 11% of malignant primary bone tumors and rarely occurs intracranially [1]. Most of the previously reported cranial chondrosarcomas were found at the base of the skull and presumably arose from cartilaginous synchondroses [2–5]. There are only a few reported cases originating above the skull base and all of these have been associated with the falx except for one, which was in the vermis of the cerebellum [2, 6–12]. It is not

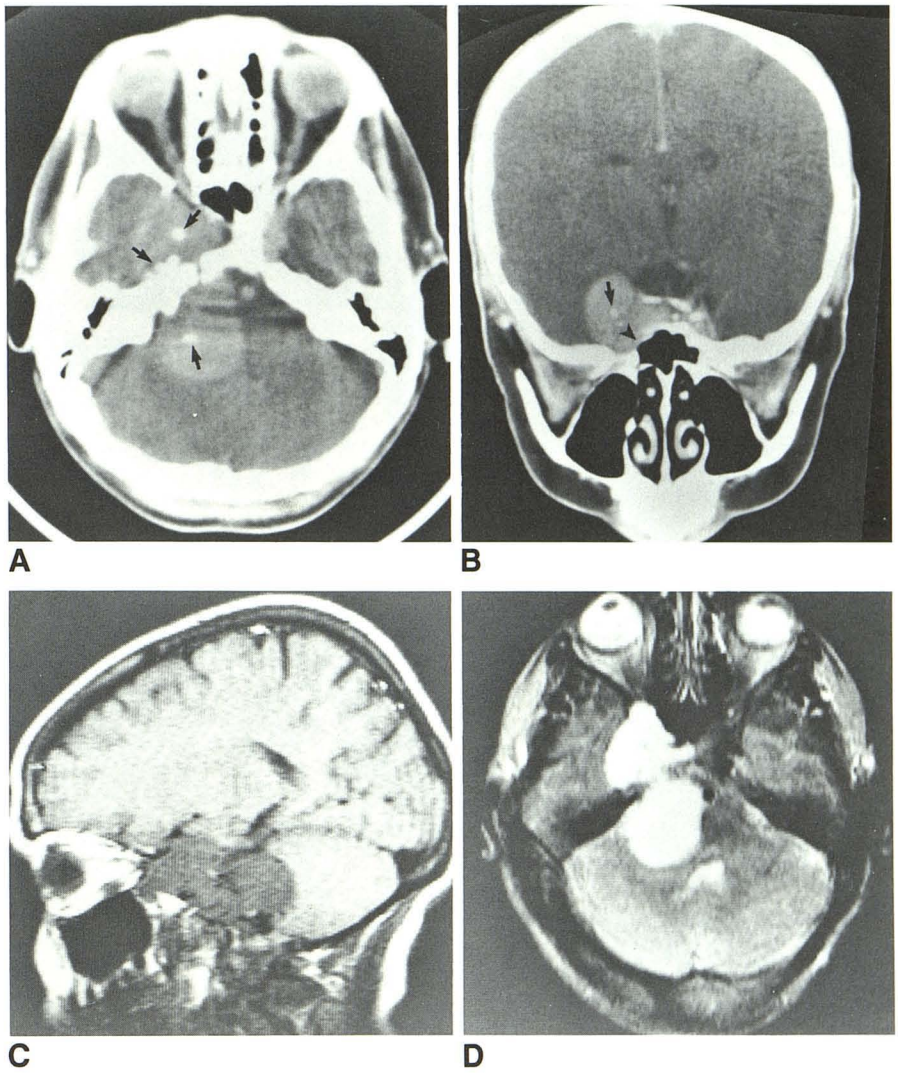

Fig. 2.—Case 2.

***A* and *B*, Axial (*A*) and coronal (*B*) contrast CT scans. A dumbbell-shaped, solid, enhancing, extraaxial mass occupies both right middle and posterior cranial fossae with intratumoral calcifications (*arrows*) and erosion of right sphenoid wall (*arrowhead*).**

***C* and *D*, Sagittal (300/32) (*C*) and axial (3500/32) (*D*) MR images. Tumor is hypointense on T1-weighted image and hyperintense on proton-density-weighted image. Tumor calcification and bone erosion are not identified.**

possible to identify the exact number of cases of chondrosarcoma of dural origin that have arisen along the base of the skull, because they have been grouped together in the literature with those arising from the cranium.

The histogenesis of intracranial chondrosarcoma remains obscure and largely a matter of conjecture. The dura itself does not normally contain cartilage. However, the meninges contain primitive multipotential mesenchymal cells that might explain the development of dural chondrosarcoma [11, 13–14]. An alternative explanation for their origin is aberrant embryonal rests [8], supported by the discovery of an ectopic nodule of cartilage in the falx of a kitten [15]. Chondrosarcomas have been classified histologically into three subtypes: classical, mesenchymal, and dedifferentiated. The classical chondrosarcomas can be further subdivided into grades I, II, and III, on the basis of their mitotic rate, cellularity, and nuclear size [16].

The radiographic appearance of cranial chondrosarcomas arising from the base of the skull has been described [2–4]. These tumors are densely calcified and originate from adjacent, destroyed bone. They generally demonstrate contrast enhancement on CT and are usually seen as avascular, extraaxial masses at arteriography. Recent advances in CT and MR have enabled the demonstration of the exact extent of tumor and the site of tumor origin. On the basis of our own experience and the few cases illustrated in the literature, it appears that the CT features of dural chondrosarcomas are more like those of meningioma than skull-base chondrosarcomas [2, 4, 10–12]. Dural chondrosarcomas tend to be less calcified or even to lack matrix calcification, in contrast to the skull-base lesions. The adjacent bone is usually eroded from the pressure effect of these slow-growing tumors, and the bone destruction common to skull-base chondrosarcomas is not seen. Case 2 shows CT and MR characteristics suggestive of meningioma [17, 18], although no precontrast CT or arteriography was performed. However, the absence of hyperostosis is against the diagnosis of meningioma. Case 3 could suggest a lipomatous meningioma, and case 1 a cystic

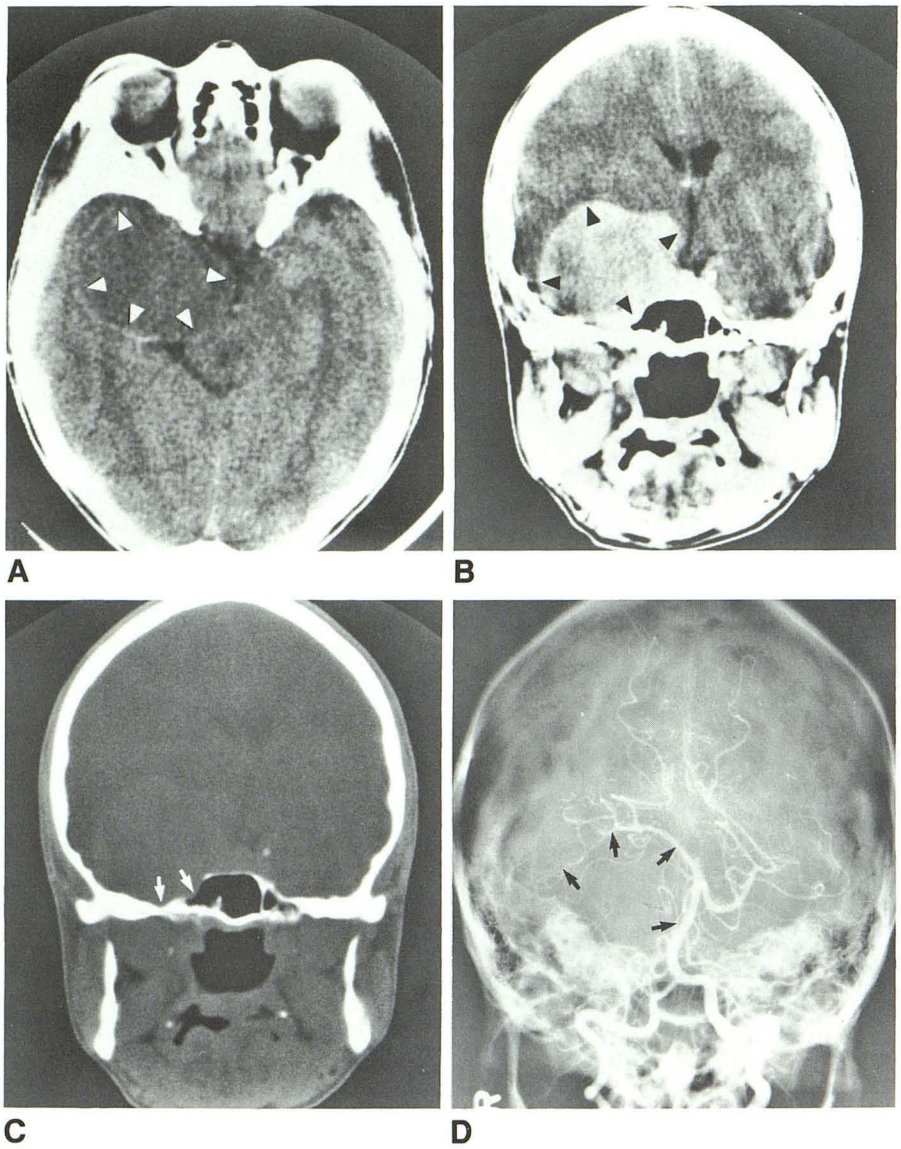

Fig. 3.—Case 3: Precontrast axial (*A*) and postcontrast coronal (*B and C*) CT scans. Tumor is hypodense and solidly enhanced (*arrowheads*), without edema. Minimal bone erosions (*arrows*) are demonstrated on the bone-setting images. *D*, Anteroposterior left vertebral arteriogram. Tumor appears extraaxial and avascular (*arrows*).

or necrotic meningioma [19]. However, there were no associated hyperostotic changes, and cerebral arteriography revealed that the tumor was avascular in both cases.

The CT and MR appearance of intracranial dural chondrosarcoma is not pathognomonic, but should suggest a relatively slow-growing extraaxial tumor. The lesions to be differentiated from dural chondrosarcoma vary depending on the location of tumor. The differential diagnosis for such lesions along the floor of the cranial fossae or in the parasellar region includes: chordoma, craniopharyngioma, neuroma, carotid aneurysm, and meningioma. Chordoma characteristically appears as a more centrally located mass arising from the clivus and destroying adjacent bone, with or without calcification [20]. Craniopharyngioma may occasionally present with a large, calcified parasellar extension, but usually the suprasellar and/or intrasellar components will serve to differentiate it from other lesions. A cranial nerve neuroma may be indistinguishable from a dural chondrosarcoma, but the absence of tumoral calcification and enlargement of the corresponding cranial foramina may lead to a correct diagnosis. A carotid artery aneurysm may present on CT as a calcified, enhancing lesion in the parasellar region and can be excluded by carotid arteriography or, in some cases, MR. The main entity in the differential diagnosis of falcial chondrosarcoma is the more frequently occurring meningioma, as is the case with such lesions along the floor of the cranium. Classically, meningioma presents on CT as a hyperdense, solidly enhancing extraaxial mass with adjacent reactive bone changes. Atypical CT features such as cyst formation, necrosis, or lipomatous infiltration may be confusing [19], but often the arteriogram will demonstrate classic tumor hypervascularity and enlarged feeding dural arteries characteristic of meningioma. The very rare falcial osteosarcoma can be confusing radiologically and pathologically [21].

In conclusion, intracranial dural chondrosarcoma appears as a slow-growing, extraaxial mass with bone erosion and

may mimic an atypical meningioma. However, the absence of hyperostosis and bone destruction as well as avascularity at angiography tend to lead the diagnosis away from meningioma. Dural chondrosarcoma is a possible entity in the differential diagnosis of a presumed meningioma, particularly when atypical features are present.

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