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Intracranial Mesenchymal Chondrosarcoma

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A case of intracranial mesenchymal chondrosarcoma (IMC) was treated surgically at our institution. CT images of IMC have not been previously published; and a review of the literature found only one reported case with CT correlation [1]. The oldest patient with a primary IMC previously reported was 51 years old [2]. We present a case occurring in a 61-year-old woman and describe the CT and angiographic findings.

Case Report

A 61-year-old woman presented with a 3-month history of nausea, difficulty with gait, morning headaches in the frontal region, and subjectively decreasing upper extremity strength bilaterally. Neurologic examination revealed a trace of left upper extremity weakness and no evidence of papilledema. A conventional radionuclide bone scan demonstrated increased tracer uptake in the right parietal region. Skull radiographs revealed dense flocculent calcification measuring 3 × 4 cm just off the midline and posterior to the coronal suture on the right. A CT scan of the head showed heavy calcification in the right frontal area with some mass effect. The calcification was not contiguous with the calvaria (Fig. 1A). Another CT scan, taken after administration of contrast material, revealed a large, inhomogeneously enhancing mass surrounding the calcification with some edema posteriorly (Fig. 1B).

A right internal carotid arteriogram showed no neovascularity associated with the calcification (Figs. 2A and 2B). There was no feeding of the tumor from the right external carotid vessels. Dural calcification extending into the interhemispheric fissure was well demonstrated on the anteroposterior view.

At surgery, a large, calcified mass attached to the right parietal convexity dura and falx was removed in part. Radiation therapy was planned and the patient is doing well postoperatively.

Discussion

Mesenchymal chondrosarcoma is a rare, malignant tumor first described by Lichtenstein and Bernstein in 1959 [3]. Initially thought to be a neoplasm of bone, approximately one-half of cases are extraosseous in origin [4]. The meninges are the most frequently reported site of extraskeletal origin [5]. Since Dahlin and Henderson's report [6] of an IMC in 1964, 16 additional cases including our own have been documented in the literature [1, 2, 4–12].

Primary cartilaginous tumors account for approximately 16% of all intracranial neoplasms [13]. Pathologically, these include benign chondromas, chondrosarcomas, and mesenchymal chondrosarcomas. Mesenchymal chondrosarcoma is easily distinguished from classical chondrosarcoma histologically as the former consists of a more primitive background of fusiform cells alternating with well-differentiated islands of cartilage [9, 14]. As might be expected, IMC has a much worse prognosis than classical chondrosarcoma, with a marked tendency for local recurrence and distant metastases [8]. Classical chondrosarcoma tends to occur at the skull base, have a peak manifestation in the fourth and fifth decades, and be avascular at angiography [14–17]. IMC tends to occur in the frontoparietal region (12/17 cases), have a peak manifestation in the third decade (age range, 7–61 years), and be highly vascular at angiography (approximately one-half of the cases) [1, 18].

In our case, oligodendroglioma was considered the most likely diagnosis on the basis of the CT and angiographic findings. Meningioma was thought unlikely in the absence of external carotid arterial supply, hyperostosis, or homogeneous contrast enhancement on CT.

The only previously described CT scan of IMC demonstrated a large, lobulated enhancing mass in the frontoparietal convexities bilaterally [1]. Calcification was not mentioned. Angiographically, the tumor mimicked an arteriovenous malformation. IMC should be considered in the differential diagnosis of a calcified, enhancing mass in the frontoparietal region. This is important to the referring neurosurgeon. Owing to the tumor's highly aggressive nature, local postoperative radiation as well as pre- and postoperative chemotherapy have been recommended, particularly in cases of subtotal resection [8, 18].

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REFERENCES


Fig. 1.—A and B, CT scans of brain without and with contrast show flocculent calcification with contrast enhancement in right frontal vertex with associated edema and bowing of a thickened falx cerebri.

Fig. 2.—A, and B, Right internal carotid arteriogram. Anteroposterior and lateral views show no neovascularity in region of dense calcification corresponding to the lesion seen on CT. Note dural calcification extending into interhemispheric fissure and involving the falx.