Calcified Middle Cranial Fossa Mass

A 21-year-old male presented for evaluation of transient loss of consciousness and was found to have a hyperdense mass in the left middle fossa. He underwent craniotomy for tumor resection. Intra- and extradural invasion was noted. Gross total resection was achieved. Pathology demonstrated a densely cellular neoplasm with predominately spindle cell morphology in a collagen-containing stroma, areas of vascular proliferation, focal mineralization, and regions of cartilage formation. High mitotic index and regions of necrosis were seen. Based on the final diagnosis of osteosarcoma, the patient was referred for chemotherapy and radiation. Intracranial osteosarcoma is a nonmeningiomaticous mesenchymal tumor. Most osteosarcomas are meningeval-based and supratentorial. Presentation depends on tumor location and may include focal neurologic deficits, cranial neuropathy, seizures, or symptoms of increased intracranial pressure. Given the relative rarity of intracranial osteosarcoma, there are no established guidelines for treatment, and therapy is guided by experience with systemic osteosarcoma. Gross total resection is recommended whenever feasible. Both chemotherapy and radiation therapy are used as adjuvant therapy. Regardless of treatment, osteosarcoma remains a highly aggressive malignancy with a poor prognosis. Morbidity and mortality may be the result of local recurrence or development of pulmonary or skeletal metastasis.
Well-defined regions of cartilage formation were noted (Fig. 5). High mitotic index and regions of necrosis were seen. Immunohistochemistry for S100, epithelial membrane antigen (EMA), glial fibrillary acidic protein (GFAP), and synaptophysin was negative. Based on the final diagnosis of osteosarcoma, the patient was referred for chemotherapy and radiation.

Intracranial osteosarcoma belongs to the class of nonmeningiomatous mesenchymal tumors, which encompass benign lesions, such as lipoma, and more malignant lesions, such as hemangiopericytoma, osteosarcoma, and chondrosarcoma. These tumors are relatively rare, with sarcomas comprising between 0.2 and 2.5% of intracranial tumors. In contrast to long bone osteosarcoma, intracranial osteosarcoma is more common in adults. There is no clear gender predilection. Most osteosarcomas are meningeal-based and supratentorial. Presentation depends on tumor location and may include focal neurologic deficits, cranial neuropathy, seizures, or symptoms of increased intracranial pressure.

Radiographic evaluation of osteosarcoma typically demonstrates intralesional calcification. Osteolysis is common. The tumors are typically isointense on T1-weighted MRI and hyperintense on T2-weighted imaging. MRI may demonstrate areas of hemorrhage or necrosis. Although osteosarcoma may appear well-circumscribed on imaging, there is typically some degree of parenchymal infiltration.
Differential diagnosis includes hemangiopericytoma, mesenchymal chondrosarcoma, synovial sarcoma, anaplastic meningioma, gliosarcoma, melanoma, and metastatic spindle cell carcinoma. Immunostaining is helpful to eliminate meningioma on the basis of EMA negativity, while GFAP negativity rules out astrocytic tumors. Evidence of direct osteoid formation by the tumor cells is key to the proper diagnosis.5

Given the relative rarity of intracranial osteosarcoma, there are no established guidelines for treatment, and therapy is guided by experience with systemic osteosarcoma. Gross total resection is recommended whenever feasible. Both chemotherapy and radiation therapy are used as adjuvant therapy.4,5 Regardless of treatment, osteosarcoma remains a highly aggressive malignancy with a poor prognosis. Five-year survival is between 25 and 37%. Median survival of 12 to 18 months has been reported.1,5 Morbidity and mortality may be the result of local recurrence or development of pulmonary or skeletal metastasis.

References