Case Presentation

A 6-month-old boy presented to the emergency department after falling from his bed onto a wood floor. The mother of the patient reported soft-tissue swelling over the posterior aspect of the head. Unenhanced computed tomography (CT) of the brain was performed, followed by contrast-enhanced MRI of the brain (Figure 1A-D).

**FIGURE 1.** Axial noncontrast CT image through the head (A) demonstrates a mixed attenuation mass with specks of calcification in the right peri-sylvian region. The peripheral component of the mass is hyperdense compared to brain parenchyma. There is mass effect on the underlying parenchyma with effacement of the right lateral ventricle and midline shift to the left. Coronal T2 MR image (B) better depicts central cystic appearing T2 hyperintense and peripheral solid T2 iso- to hyperintense components and confirms the extra-axial nature of the mass with displacement of underlying gray matter. Coronal (C) and axial (D) T1 postcontrast MR images reveal a broad dural base laterally (D) and avid enhancement of the peripheral solid components with extension into the right orbital apex and suprasellar region (D). There is scalloping of the right sphenoid wing (D), which was better demonstrated on initial CT imaging through the skull base (not shown).
Key Imaging Findings
- Dural-based mass in an infant

Differential Diagnosis
- Rhabdomyosarcoma
- Metastatic Neuroblastoma
- Leukemic masses
- Meningioma

Discussion
Brain tumors represent the second most common malignancy in childhood after leukemia, comprising 20%-25% of all neoplasms in children under 15 years of age.1 Supratentorial neoplasms are more prevalent in children under 3 years of age, while infratentorial tumors are more prevalent in children over 3 years of age. The patient age and tumor location help guide an appropriate list of differential considerations. In the setting of a dural-based tumor in an infant, the primary considerations include rhabdomyosarcoma, metastatic neuroblastoma, leukemic mass, and rarely meningioma.

Rhabdomyosarcoma
Rhabdomyosarcoma represents the most common soft-tissue tumor subtype in pediatrics, with lesions commonly originating in the head and neck. Almost 65% of cases are diagnosed in children under 16 years of age. Rhabdomyosarcomas of the head and neck occur most often in the parameningeal areas (ie, middle ear, infra temporal region, paranasal sinuses, nasopharynx, etc.), orbit, scalp, parotid gland, and thyroid gland, among other areas.2 Clinical symptoms are site-dependent with chronic otitis from temporal bone/middle ear involvement, sinusitis from paranasal sinus involvement, cranial nerve palsies that may result from multiple sites of involvement, and proptosis from orbital involvement.3

CT demonstrates a soft-tissue mass that is often ill-defined with bony destruction. MR imaging is preferred due to its superior soft-tissue evaluation, demonstrating a heterogeneously enhancing soft-tissue mass in a location typical of rhabdomyosarcoma (usually the head and neck soft tissues with or without intracranial extension). Perineural and meningeal spread is characterized by abnormal contrast enhancement on MR. Fat-suppressed T1-weighted imaging can be employed to evaluate for spread into the skull base; however, CT is most useful for osseous involvement. Treatment options include chemotherapy, radiation therapy, and surgery. Parameningeal locations have the worse prognosis, with a 5-year disease-free survival of 57%.3

Metastatic neuroblastoma
Neuroblastoma is a malignant tumor that originates from primitive neural crest cells.4 Most commonly, neuroblastoma arises from the adrenal glands or retroperitoneum but can occur anywhere along the sympathetic chain; < 5% of cases arise from the cervical sympathetic chain ganglia.5 The head and neck is the most common site of distant metastases, with metastases involving osseous, lymphatic, and dermal structures.6 The most common site of intracranial metastases is the dura; contiguous invasion into the brain parenchyma is rare since the dura acts as a barrier to spread.5 Symptoms of intracranial metastases are often attributable to lesion location with headache, nausea, and emesis being the most common symptoms.6

In the setting of osseous metastases, CT may demonstrate aggressive periosteal reaction (often termed “hair-on-end” appearance), lytic defects, and separation of sutures.5 Parenchymal metastases tend to be cystic with variable degrees of internal calcification and heterogeneous enhancement of solid components.6 It is theorized that the cystic components result from prior hemorrhage, while most lesions tend to have a variable degree of hemorrhage both on imaging and pathologic evaluation. MR imaging demonstrates dural-based masses with aggressive osseous destruction, often with internal calcification and hemorrhage.5 Increased T2 signal can be seen surrounding the mass from vasogenic edema.

Leukemic masses
Leukemic involvement of the head is variable but mainly involves the calvarium, orbital/periorbital region, and/or meninges.7 Brain parenchymal involvement is rare but is seen more often during relapse.8 Imaging findings of calvarial involvement consist of homogeneously enhancing mass(es) with expansion of the calvarium and adjacent meningeal enhancement. Orbital and periorbital involvement typically include enhancing soft-tissue masses mostly homogenous in appearance.

Meningioma
Meningeal tumors in children are rare, accounting for approximately 0.4-1% of intracranial tumors.9 There is a known predilection for meningioma formation, however, in patients with neurofibromatosis type 2 (NF-2), with a 19%-24% incidence.10 Meningiomas may also be seen with a higher frequency in children previously treated with whole-brain radiation for other central nervous system neoplasms. In contrast to adults, there is a male predominance with a male-to-female ratio of 2.4:1.11 Meningiomas in children tend to be more aggressive and behave differently than lesions seen in adulthood.9 Signs and symptoms at presentation vary according to location of tumor, with the most common symptom being headache. Infants may present with signs and symptoms of increased intracranial pressure, such as vomiting and/or increased head size; however, as brain tumors are relatively rare in infants, symptomatology can be frequently overlooked.

Pediatric meningiomas have a variety of tumor subtypes, as well as...
imaging characteristics. The various histologic subtypes of meningioma include meningothelial, fibroblastic, sclerosing, angioblastic, sarcomatous, transitional, and malignant. The malignant variants include papillary, anaplastic, and rhabdoid subtypes, with the papillary subtype classified as a WHO grade III malignant tumor that accounts for 1.0%-2.5% of all meningiomas in children.

On CT, meningiomas often present as enhancing partially solid and cystic extra-axial masses with variable degrees of internal calcification and surrounding edema. Dural-based lesions are not as common as in adults. MR imaging demonstrates iso- to hypointense extra-axial masses on T1 and T2 sequences with heterogeneous contrast enhancement. Prognosis of meningiomas in pediatric patients relates to degree of tumor resection, tumor location, and pathologic grade. If the diagnosis of meningioma is confirmed in a child without a prior history of radiation therapy, additional work-up for an underlying genetic disorder such as NF-2 is warranted. This patient had a positive work up for NF-2; a few small schwannomas were detected along the spinal nerve roots.

**Diagnosis**

Meningioma (papillary variant with rhabdoid features, WHO grade III)

**Summary**

The differential considerations for brain neoplasms in children depend on patient age and tumor location. When presented with an extra-axial dural-based mass in an infant, the primary considerations include rhabdomyosarcoma, metastatic neuroblastoma, leukemic infiltrates, and rarely meningioma. The imaging appearance of these entities have some overlap. However, in general, rhabdomyosarcomas originate from the head and neck soft tissues with or without intracranial extension; neuroblastomas have more osseous involvement and are likely to calcify; and leukemic infiltrates tend to be more homogeneous and less destructive in appearance. The diagnosis of meningioma in infants is rare in the absence of NF-2. When present, meningiomas in children are typically of higher grade with heterogeneous enhancement and cystic changes.

**References**