PRIMARY CNS EWING SARCOMA: A REPORT OF 2 CASES
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Introduction: Pediatric small round blue cell tumors of the central nervous system (CNS) include several families of malignant tumors. Primary Ewing’s sarcoma/peripheral primitive neuroectodermal tumor (ES/pPNET) of the CNS is a rare entity and we report two cases here.

Case 1: The patient is a 35 month-old girl who presented with partial seizure involving left facial twitching and lip smacking. MRI showed a large frontal, cortically-based, well circumscribed, largely solid, enhancing mass that abutted the meninges and caused moderate peritumoral edema. Grossly, the tumor was largely solid but contained a small centrally located cystic space. Histologically, it was featured by sheets of small blue cell tumor in a background of collagenous fibers suggestive of a sarcoma.

Case 2: The patient is a 28 month-old boy who presented with left side weakness, unsteady gait, and frequent falls. MRI showed a largely cystic frontotemporal mass touching the meninges and contained a small enhancing solid nodule. The tumor caused midline shift and mild peritumoral edema. Histologically, it was composed of solid sheets of small blue cells with occasional perivascular arrangement but no intervening collagen fibers. The histological features suggested a primitive neuroectodermal tumor (PNET) or ependymoma.

Special studies: For both cases, immunohistochemistry for CD99 showed strong membranous staining. Immunohistochemistry on Case 2 was suggestive of a central PNET. Both tumors were positive on PAS stain but negative on PAS with diastase. EWSR1 translocations were demonstrated by FISH.

Discussion: ES/pPNET tumors are rare primary tumors in the CNS and it is important to recognize them for appropriate treatment. Radiographically, they are typically circumscribed tumors, sometimes connected to the meninges. Histologically, the differential diagnosis includes primary and metastatic sarcoma, central PNET and medulloblastoma, ependymoma, metastatic neuroblastoma, and, less likely, atypical teratoid/rhabdoid tumor and hematopoietic tumors. The second case that we are reporting here is particularly challenging as it possesses some histologic and immunohistochemical features of central PNET and ependymoma. A high index of suspicion is mandatory for diagnosis. Immunohistochemistry for CD99 in combination of PAS stain with and without diastase provide an efficient and inexpensive mean to avoid misdiagnosis and FISH is a valuable adjunct for diagnosis.