Tumor and Tumorlike Masses in Pediatric Patients that Involve Multiple Spaces

Thangamadhan Bosemani, MD*, Andrea Poretti, MD

KEYWORDS

• Tumor • Tumorlike • Multiple spaces • Intracranial • Children • MR imaging

KEY POINTS

- The pediatric central nervous system is affected by tumors and tumorlike masses that involve multiple spaces.
- Neuroimaging plays an important role in distinguishing tumors from tumorlike masses.
- MR imaging may represent the best predictor of neurodevelopmental abnormalities, seizures, and requirement of neurosurgery in neurocutaneous melanosis.
- Histiocytic disorders, infectious or inflammatory processes, and neurocutaneous syndromes should be considered potential tumor mimics.

INTRODUCTION

Brain tumors are the most common solid tumor of childhood, the second most common malignancy overall after leukemia and the leading cause of mortality among all childhood cancers.[1] Primary brain tumors have been categorized by the World Health Organization classification to include tumors of the neuroepithelium, cranial nerves, meninges, and sella and those of hematopoietic and germ cell origin.[2] Brain tumors typically form masses, which infiltrate or compress brain parenchyma. The tentorium cerebelli divides the brain parenchyma into supratentorial and infratentorial compartments. The meningeal layers covering the brain create potential anatomic spaces such as epidural, subdural, and subarachnoid. Masses may be confined to a single compartment/space or may extend across multiple compartments/spaces. Not every brain mass, however, represents a tumor or neoplasm. Tumorlike masses of the pediatric brain include histiocytic disorders, inflammatory lesions, vasculitis, ischemia, hemorrhage, vascular malformations, and malformations of brain development.[3]

The recognition of a brain mass on imaging is the first step in the workup. This recognition is followed by verifying its location, identifying it as a tumor and distinguishing it from tumorlike mimics, characterizing the tumor grade and type, and describing its characteristics (including vascularity, vascular permeability, and metabolic spectrum). Hence, imaging plays a vital role in establishing the correct diagnosis, establishing a therapeutic strategy, and influencing prognosis.

Disclosure: The authors report no conflicts of interest.
Section of Pediatric Neuroradiology, Division of Pediatric Radiology, The Russell H. Morgan Department of Radiology and Radiological Science, Charlotte R. Bloomberg Children's Center, The Johns Hopkins School of Medicine, Sheikh Zayed Tower, Room 4174, 1800 Orleans Street, Baltimore, MD 21287-0842, USA
* Corresponding author.
E-mail address: tbosema1@jhmi.edu

Neuroimag Clin N Am 27 (2017) 135–153
http://dx.doi.org/10.1016/j.nic.2016.08.006

MR imaging, including conventional (anatomic) and advanced (functional) sequences, is the study of choice.

This article reviews pediatric brain tumors that involve multiple spaces, including lymphoproliferative disorders, melanocytic lesions, metastasis, tumors of meningothelial cells, and tumors in children with neurofibromatosis type 1 (NF-1), neurofibromatosis type 2 (NF-2), and von Hippel-Lindau (VHL) disease. In addition, tumorlike masses, which include histiocytic disorders, inflammatory processes, infectious etiologies, and neurocutaneous syndromes, are reviewed. For each entity, epidemiology, pathophysiology, and neuroimaging findings are discussed. For tumorlike masses, features and findings that allow the correct differentiation from neoplastic masses are discussed.

TUMORS ACROSS MULTIPLE SPACES

Lymphoproliferative Disorders

Lymphoproliferative disorders in childhood include central nervous system (CNS) involvement in childhood leukemia and non-Hodgkin lymphoma (NHL), primary central nervous system lymphoma (PCNSL), and posttransplant lymphoproliferative disorder with CNS involvement (CNS PTLD). Lymphoproliferative disorders may extend across multiple spaces, such as the brain parenchyma, meninges, bone marrow, or calvarium.

Central nervous system involvement in childhood leukemia

Acute lymphoblastic leukemia (ALL) is a common hematologic malignancy of the bone marrow in which precursor lymphoblasts, blocked at an early stage of differentiation, proliferate rapidly and replace normal hematopoietic cells of the bone marrow.[4] ALL is the most common childhood malignancy, accounting for approximately 25% of cancers and 80% of all leukemias in children.[4]

CNS involvement is typically clinically occult and discovered at the time of lumbar puncture. Children may present with cranial nerve deficits, seizure, altered mental status, headache, or other neurologic deficits.[5]

Primary neuroimaging manifestations of ALL include enlargement of the lateral ventricles (present before therapy, hence, representing hydrocephalus rather than atrophy), leukemic infiltration of CNS structures (meninges, parenchyma, bone marrow, orbit, and spine; **Fig. 1**A–D), and cerebrovascular complications (hemorrhage, cerebral infarction).[6]

Leukemic infiltration may involve the leptomeninges, subarachnoid space, or epidural space. Meningeal thickening and enhancement can be either focal or diffuse on postcontrast T1-weighted images. Leptomeningeal enhancement of the brain may result from CNS leukemia/relapse, infection, or, rarely, both. Parenchymal leukemic infiltration is very uncommon, it is often hyperdense on CT, it is contiguous with a meningeal surface, and it enhances after contrast material administration.[6] On MR imaging, parenchymal leukemic CNS infiltration tends to be slightly hypointense on T1-weighted images and isointense to mildly hyperintense on T2-weighted and fluid-attenuated inversion recovery (FLAIR) images compared with gray matter.

Chloroma, also known as *granulocytic sarcoma*, is a focal extramedullary mass of immature myeloid cells of granulocytic lineage that infiltrate bone and soft tissue, most frequently occurring in myelogenous leukemia.[7] Chloromas most commonly arise in the skull, orbits, and sinuses (**Fig. 2**).

Spinal involvement is frequent in leukemia and lymphoma, either as initial manifestation or in relapse, with bone marrow or meningeal infiltration (**Fig. 1**E, F). MR imaging of the bone marrow shows low-signal-intensity leukemic infiltrates on T1-weighted images.[6] Subarachnoid nodules, enhancement, and sugar coating of the nerve roots and cauda equina consistent with leptomeningeal seeding are seen on contrast-enhanced T1-weighted images.

Primary cerebrovascular complications are related to leukocytosis, thrombocytopenia, sepsis, or coagulopathy. Hemorrhage is the most common cerebrovascular complication. Hemorrhage can be identified on conventional MR imaging sequences such as T1 and T2 and on advanced sequences such as susceptibility-weighted imaging. Hemorrhage may show mass effect, midline shift, and perifocal edema.

More common than primary leukemic infiltration and primary cerebrovascular complications are treatment-related changes of the CNS including infection, acute neurotoxicity, demyelination, and hemorrhage.[6]

Central nervous system involvement in childhood non-Hodgkin lymphoma

CNS involvement in NHL is identified by either malignant cells in the cerebrospinal fluid or by cranial nerve palsy on physical examination, which guides further management.[8] About 6% of childhood/adolescent NHL patients are CNS positive during the course of the disease.[9] On MR imaging, leptomeningeal disease with meningeal thickening and enhancement may be seen. MR imaging is highly sensitive for the presence