Case 4: Malignant mesenchymal neoplasm (NOS) with rhabdoid features
(presented by F. Giangaspero)

The neoplasms is a solid-appearing moderately cellular spindle cell neoplasms with abundant intercellular collagen deposition. The tumor is moderately vascular, but without the typical “staghorn vessels”. A number of the blood vessels are hyalinized. In some field the cells have “rhabdoid” features with abundant eosinophilic cytoplasms and eccentrically displaced nuclei. Only occasional mitoses are seen. By immunohistochemistry, the tumor cells show strong membrane pattern immunoreactivity for CD99. The INI-1 (BAF47) stain shows diffuse nuclear immunoreactivity consistent with retained expression. Stains for smooth muscle actin, CD31, and CD34 highlight the rich vascular network, although the cells are negative. The tumor is also negative for BCL-2, synaptophysin, GFAP, and epithelial membrane antigen (EMA). The Ki67 labelling index is low (3%). Ultrastructural analysis performed from paraffin material did not show any evidence of meningothelial differentiation.

Differential diagnosis:

Solitary fibrous tumor
Based on pure morphology this diagnosis was favored. In support of this interpretation, the CD 99 showed diffuse embramnous immunoreactivity. However against this diagnostic possibility was the lack of either CD34 or BCL-2 immunoreactivity.

Hemangiopericytoma
The non-specific immunohistochemical pattern would fit with the diagnostic consideration of hemangiopericytoma WHO grade II, however the typical vascular pattern was not present.

Meningothelial (rhabdoid) meningioma
No immunohistochemical (EMA, S100 negativity) or ultrastructural support for meningothelial differentiation.

Rhabdoid tumor
The retained nuclear expression of INI-1 (BAF47) protein excludes the possibility that this lesion represents a rhabdoid tumor.

Discussion

Meningial tumors are considerably less common in infants and children than in adults. Only 2.5% of all primary pediatric central nervous system (CNS) tumors are meningeal in origin. Although some of these tumors have identical features to their adult counterparts, many possess unique properties in children. Non-meningothelial mesenchymal tumors arising as dural-based masses constitute a heterogeneous group that includes a number of entities better known in sites outside the CNS. Sarcomas represent the most common category of primary non-meningothelial neoplasms arising from or involving the dura in childhood. Hemangiopericytoma is the most frequent of the primary dural sarcomas in children and adults alike. Other dural-based spindle cell sarcomas in children include fibrosarcoma, extraskeletal myxoid chondrosarcoma, malignant fibrous histiocytoma, and undifferentiated sarcomas lacking specific histologic and immunophenotypic features. Some are associated with a history of prior radiation therapy, but most are sporadically occurring. Leiomyosarcomas have been described in immunosuppressed children, particularly in the setting of AIDS and are an apparent consequence of Epstein-Barr virus (EBV) infection. The other category of sarcomas with a predilection for children are those with an undifferentiated “small blue cell” or round cell appearance such as mesenchymal chondrosarcoma and Ewing sarcoma-peripheral primitive neuroectodermal tumor (EWS-pPNET).

The case here presented lacked any specific histological, immunophenotypical and ultrastructural features that could fulfill the diagnostic criteria for one of the formerly mentioned sarcomas. A descriptive diagnosis of low grade mesenchymal neoplasms (NOS) with rhabdoid features has been proposed.
The child died of the disease because the neoplasm did not respond to chemotherapy confirming the biological aggressiveness of the tumor. No similar cases have been reported in the literature so far. This case enlarges the spectrum of non-meningothelial dura based mesenchymal tumors that can occur in young children.

References
