AJNR

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AJNR Am J Neuroradiol 1995, 16 (8) 1727-1728
http://www.ajnr.org/content/16/8/1727

Cranial Rhabdoid Tumor with Marginal Tumor Cystic Component and Extraaxial Extension

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Summary: We report the radiologic findings in a case of primary central nervous system malignant rhabdoid tumor with a cystic marginal component in a 4-month-old infant. Marginal tumoral cysts are unusual in tumors other than esthesioneuroblastoma.

Index terms: Brain neoplasms, magnetic resonance; Children, neoplasms

Malignant rhabdoid tumor is a rare childhood neoplasm that typically arises within the kidneys and is characterized by an aggressive clinical course. Since its recognition in 1978, 14 cases involving the central nervous system have been reported (1–6). We have had the opportunity to image an infant with this tumor involving the central nervous system (CNS) diagnosed in early infancy. The tumor had marginal cyst formation and an extraaxial nasosinusal component, mimicking tumors originating in the olfactory tract.

Case Report

A 4-month-old boy was referred to the emergency pediatric ward because of cough and mild fever for 3 days. His mother's pregnancy was uneventful, and he was delivered at term without complications. The only remarkable abnormality was macrocephaly. A cranial ultrasound was ordered, and an intracranial mass was detected. The abdominal ultrasound was normal. The blood test revealed nonspecific leukocitosis. The patient was then referred to our institution. Plain and enhanced computed tomography was performed (not shown). The study demonstrated a huge bifrontal intracranial tumor with a tiny calcification, extensive surrounding edema, diffuse enhancement, and a cystic marginal component. The mass was attached to the base of the anterior cranial fossa with a small amount of tumor extending into the upper ethmoid sinus and nasal cavity. Magnetic resonance was performed with T1-, T2-, and proton density–weighted sequences and an enhanced T1-weighted sequence in several planes. The MR demonstrated a large mass with a nasoethmoidal component and involvement both frontal lobes. There was extensive vasogenic edema and invasion of the ventricular system. The tumor also showed a diffuse irregular enhancement and had a marginal superior cyst (Fig 1). Relaxations times of the tumor were nonspecific. At this time, enhanced subarachnoid nodules were detected, suggesting extensive leptomeningeal spread. Angiography was performed. The tumor was fed by many branches of the internal and external carotid arteries with an extensive tumoral capillary network. A frontal craniotomy was done, and a modest tumoral resection was achieved. The patient died 3 days later. Pathologic examination showed a moderately vascularized neoplasm. A high proliferation of undifferentiated polygonal cells with eccentric nuclei were discovered. These cells also showed a plasmacytoid epithelioid aspect with hyaloid inclusions. Immunohistochemical staining was positive for vimentin, cytokeratin, and epithelial membrane antigen. The final pathologic diagnosis was malignant rhabdoid tumor.

Discussion

Although its origin is uncertain, malignant rhabdoid tumor may arise from the neural crest. The mean age of patients in whom malignant rhabdoid tumor is detected is 2.7 years (2). The CNS location of this tumor is diverse and includes hemispheric supratentorial, infratentorial, and multifocal, with an aggressive extension including intraventricular. Postsurgical imaging of patients operated on showed not only local relapse but devastating leptomeningeal spread within few months (3, 4).

Imaging studies of malignant rhabdoid tumor showed large masses with multiple necrotic/cystic foci and diverse degrees of enhancement with associated parenchymal edema in both computed tomography and magnetic reso-

Received November 3, 1994; accepted after revision March 16, 1995.

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AJNR 16:1727–1728, Sep 1995 0195-6108/95/1608–1727

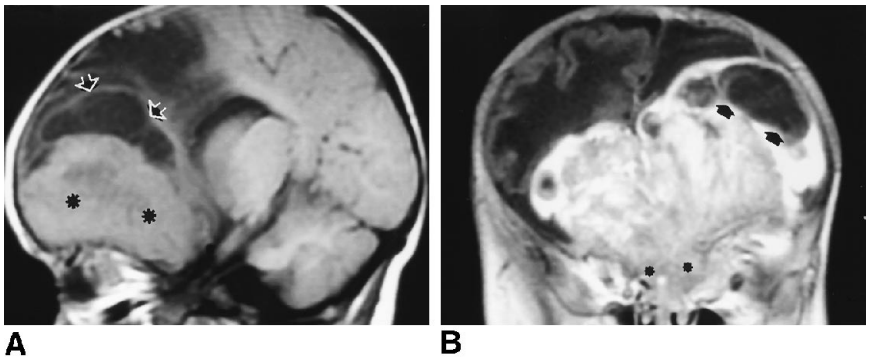

Fig 1. Sinonasal and subfrontal extension of a malignant rhabdoid tumor in a 4-month-old infant.

A, Parasagittal nonenhanced T1-weighted image. A tumor (*asterisks*) with nonspecific signal, marginal cystic component (*arrows*), and extensive surrounding edema is identified.

B, Coronal enhanced T1-weighted image at the level of the orbits shows a tumor that involves the central sinonasal cavity (*asterisks*) and extends through both frontal lobes. Notice left apical marginal cystic component (*arrows*).

nance. Calcification may be present. Subarachnoid dissemination at the time of diagnosis often is present (2, 3, 4).

Pathologically, malignant rhabdoid tumor is characterized by polygonal or round undifferentiated cells with eosinophilic cytoplasm, containing hyaloid inclusions, and roughly ovoid vesicular eccentric nuclei under light microscopic examination. By electronic microscopy, these are whorls of filaments in cytoplasm. Immunohistochemical staining indicates that these filaments are positive for cytokeratin and vimentin but negative for desmin, neurofilament, and myoglobin (4, 5).

We believe that this case fulfill the criteria for a malignant rhabdoid tumor involving the CNS on both biological and pathologic grounds. This case deserves interest in two aspects. First, the extensive extraaxial sinonasal extension mimicking sinonasal primary tumor with an intracranial extension is rare. Its primary sinonasal origin cannot be ruled out, although we found no previous reports of this origin. In fact, on the basis of magnetic resonance imaging, a esthesioneuroblastomalike tumor was proposed despite the rarity of esthesioneuroblastoma to appear at this age. The second aspect is the adjacent intracranial cyst component. According to Som et al (7), the association of marginal tumor cysts along the intracranial margin of sinunasal mass is very characteristic of esthesioneuroblastoma. Although this statement is correct by their extensive and expert experience, some exceptions that might "prove the rule" also may be possible. This case demonstrates that marginal cysts can occur with other histologies, although the phenomenon is certainly rare.

Finally, we point out the aggressivity of this tumor. In our patient, both the extraaxial and intraaxial tumor components were present. Moreover, intraventricular and subarachnoid extensions indicate extreme aggressivity. Although as many as 80% of previously reported CNS malignant rhabdoid tumors originate in the posterior fossa (6), our case may reflect the variety of potential CNS origins and extensions. The differential diagnosis of such aggressive tumors during infancy should include the primary neuroectodermal tumor, including medulloblastoma, and glioblastoma multiforme, as well as malignant teratoma. This differentiation on the basis of imaging is difficult, because all may share many radiologic findings. However, the presence of an extraaxial component as in our case, the age, and the presence of calcification will narrow the differential diagnosis.

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