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Atypical Teratoid/Rhabdoid Tumor Mimicking Tuberculous Meningitis

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ABSTRACT

Atypical teratoid/rhabdoid tumor of the central nervous system is a highly malignant neoplasm in infants and young children. We report a 6 year-old girl with atypical teratoid/rhabdoid tumor. Based on cerebrospinal fluid examination MRI scan and family history of tuberculosis; we diagnosed tuberculous meningitis. There was inadequate response to the antituberculosis therapy; so we performed stereotactic brain biopsy. Pathologic result revealed high grade atypical teratoid/rhabdoid tumor.

Key words: *Atypical teratoid/rhabdoid tumor, Tuberculous meningitis.*

INTRODUCTION

Primary atypical teratoid/rhabdoid tumors (AT/RT) are extremely rare malignant intracranial neoplasms. To date, approximately 200 cases of atypical teratoid/rhabdoid tumor (AT/RT) of the central

nervous system have been described in the literature(1). It is a highly aggressive neoplasm, often has central nervous system dissemination, does not respond to therapy and typically is fatal within 1 year(2).

CASE REPORT

A 6-year-old girl was admitted to our clinic with confusion, headache, vomiting, aphasia, and right hemiparesis for last two months. She had family history of tuberculosis. On admission, the patient was lethargic. She was not alert and oriented. On clinical examination right hemiparesis was detected. Cerebrospinal fluid (CSF) examination revealed 350 lymphocytes/mm³, protein 40.8 mg/dL and glucose 36 mg/dL. At the same time blood glucose was 136 mg/dL. MRI brain showed marked leptomeningeal involvement and basal meningitis. The initial working diagnoses included tuberculous meningitis and malignant infiltration. Spinal MRI was normal. Cerebrospinal fluid was negative for viral serology and PCR for *Mycobacterium tuberculosis*. No malignant cell were demonstrated in cerebrospinal fluid. Chest radiography did not reveal any abnormality. The tuberculin test was negative. Antitubercular and antiedema treatment was initiated. Post-treatment course showed an improvement in neurological status, but two weeks later the child developed ptosis and loss of vision. Cranial CT scan showed meningeal enhancement in the extracerebral sub-arachnoid spaces. There was minimal dilatation of third and lateral ventricles, and homogeneous contrast enhancement on both temporal lobes, right insular cortex, ambient cistern, and left sigmoid sinus. These findings were concluded as suggestive of tuberculous meningitis. Lumbar puncture revealed 120 lymphocytes/mm³ with protein 129 mg/dL and sugar 19 mg/dL.

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Antitubercular treatment was continued. After the fourth week, enhanced cranial contrast CT scan showed a 2 cm diameter mass of left temporal region. The lesion mimicked a tuberculoma. The child also developed fever and seizure by this time. The cranial MRI revealed meningeal hyperdensity and triventricular hydro-cephalus and also multiple tuberculoma lesions in both temporal lobes. A ventriculo-peritoneal shunt was performed. CT scan revealed that the solid enhancing mass has become bigger than before. The neuroradiologist concluded this mass as a fungus ball, abscess, or tumor. Stereotactic brain biopsy was performed. Two tissue specimens with surface covered leptomeninges were taken from both cerebellar hemispheres. Microscopically, malignant tumor cells were seen in the biopsy specimens. Histologically, the tumor specimens had many rhabdoid cells with prominent nuclei and eosinophilic cytoplasm and polygonal or oval cells with round nuclei, large cytoplasm and dispersed chromatin. It showed necrosis. Malignant tumor cells have attached to the meninges. The morphologic features of the tumor resembled teratoid/rhabdoid tumor. The tumor cells were immunoreactive for epithelial membrane antigen and pankreatin. Synaptophysin, glial fibrillary acidic protein and neuron specific enolase immunoreactivity was absent. Pathologic result was high grade atypical teratoid/rhabdoid tumor. The family refused chemotherapy and radiotherapy; the child died 3 months after admission.

DISCUSSION

We had mistakenly diagnosed this child as tuberculous meningitis, and also antituberculous treatment was initiated. The child did not respond clinically or radiologically; rather showed increasing size of the space occupying lesions, which were thought of as tuberculomas(3). Finally, brain biopsy clinched the diagnosis of rhabdoid tumor.

The initial diagnosis of atypical teratoid/rhabdoid tumor is made with a radiographic study (MRI or CT). Yet, there are no specific imaging features of AT/RT and neuroradiologists rarely mention atypical teratoid/rhabdoid tumor in their differential diagnosis. Heterogeneous masses with

calcifications, eccentric cysts and off midline location in the posterior fossa in children younger than 2 years should alert the radiologist to the possibility of atypical teratoid/rhabdoid tumor in the differential diagnosis of the mass(4). Leptomeningeal dissemination is common in patient with AT/RT and indicates poor prognosis(5). Cytological examination of the cerebrospinal fluid is important as 1/3 of these patients will have intracranial dissemination with involvement of the CSF. Lu, *et al.*(6) reported a 2-year-old girl with AT/RT. It is important to keep in mind the possibility of brain tumor in non-resolving tuberculomas.

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