CASE REPORT

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Atypical Teratoid Rhabdoid Tumors in Adult Patient with Multiple Lesions

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Primary atypical teratoid/rhabdoid tumor(AT/RT) of the central nervous system is a recently described, highly malignant neoplasm that usually occur in the posterior fossa of children. Although AT/RT usually occurs in young children, AT/RT is being recognized in adults with increasing frequency. The authors report 49-year-old man with multiple AT/RT lesions (right lateral ventricle, right temporal lobe and right cerebellum). Histopathologic findings showed typical rhabdoid cells with eccentric nuclei and prominent nucleoli. Eventhough the tumor was removed, a patient was dead in one month after surgery due to recurrence and rapid regrowth of the tumor.

KEY WORDS : AT/RT (Atypical teratoid rhabdoid tumor) · Adult.

Introduction

Atypical teratoid/rhabdoid tumor(AT/RT) of central nervous system is rare malignancy of infancy with high aggressiveness[7,9]. It has often been radiologically confused with a primary neuroectodermal tumor(PNET)/medulloblastoma (MB), but has a much worse prognosis, and is most common in the posterior fossa in children less than 2years[1,4,11]. Histopathologically, it is defined by the presence of rhabdoid cells and populations of primitive neuroectodermal, epithelial and mesenchymal cells[10].

Present knowledge is based on a small number of cases. Most treatments consisted of surgery, radiotherapy and chemotherapy[3,12]. Despite combined therapeutic efforts, the tumor has an extremely unfavorable prognosis due to local relapse and subarachnoid dissemination. It is almost always fatal within 1year of diagnosis. The authors report an AT/RT with multiple lesions in adult.

Case Report

A 49-year-old man presented with headache and vomiting for a week. Physical examination showed weakness on the left extremities (Grade IV). Brain magnetic resonance images(MRI) showed multiple mass lesions on the right lateral ventricle, right temporal and occipital lobes, and right cerebellum with inhomogenous enhancement. The lesion consisted of a solid and a cystic component. The cystic component appears to be slightly hypointense on the T1-weighted image, and hyperintense on the T2-weighted image. The solid component and cyst wall showed contrast enhancement with gadolinium both on axial and coronal images (Fig. 1). Subtotal tumor removal was performed via a temporal transcortical approach. After corticectomy at the middle and inferior temporal gyrus, yellowish, grayish, and dark purple colored tumor mass could be exposed (Fig. 2). It was measured by 1×2cm in the temporal base. The tumor was friable and showed well-developed vascularity with central necrosis and irregular margin. The tumor was extended from the temporal lobe and the temporal horn of the lateral ventricle to the trigone of the lateral ventricle posteriorly. Histopathology showed a typical rhabdoid cells with eccentric nuclei and prominent nucleoli (Fig. 3). In immunohistochemical findings, glial fibrillary acidic protein(GFAP), cytokeratin(CK), vimentin, epithelial membrane antigen(EMA) were expressed (Fig. 4). But, neurofilament protein(NFP) and smooth muscle actin(SMA) were negative in this case. Ten days after operation, his mental status was rapidly aggravated due to acute hydrocephalus.

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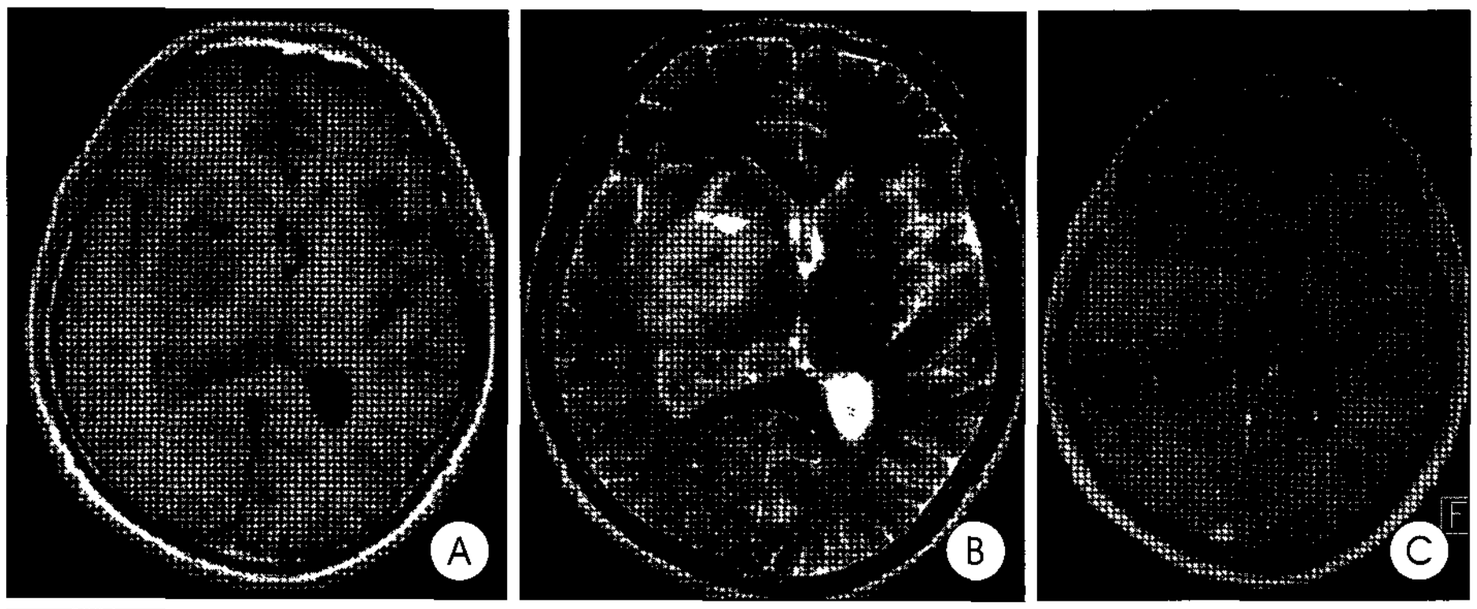

Fig. 1. The magnetic resonance(MR) image shows right temporal and lateral ventricular mass. The cystic component appears to be slightly hypointense on the T1-weighted axial image(A), and hyperintense on the T2-weighted axial image(B). The solid component and cyst wall show contrast enhancement after gadolinium administration on axial(C), coronal(D) and sagittal(E) images. Also, the mass is located in the cerebellum(D).

Fig. 2. *Intraoperative photography showing yellowish tumor mass.*

After extraventricular drainage was done, his mental status was improved. Several days later, however, his mental status was deteriorated day by day and follow-up brain computed tomography(CT) showed rapid regrowth of the tumor. He was dead in one month after surgery.

Discussion

AT/RT is a tumor of infancy and childhood with a median age at diagnosis of 16.5 months[11]. In 2000, Rorke and Biegel[9] reported that 98% of published cases had occurred in infants or children and 94% were 5years old or younger. Fifty-two percent occurred in the posterior fossa, 39% in the cerebral hemispheres or suprasellar region, 5% in the pineal region, 2% in the spinal cord and 2% were multifocal.

The radiologic features of AT/RT are nonspecific, showing multiple prominent cystic/necrotic areas associated with an inhomogeneous contrast-enhanced solid component[1]. It shows increased density on nonenhanced CT and heterogeneous contrast enhancement. Cystic change and intratumoral hemorrhage are common. On MRI, it has decreased signal intensity on T1-weighted images, iso- or decreased density on T2-weighted images (due to hypercellularity), and heterogeneous enhancement[13]. In our case, the tumor showed multiple mass lesions with inhomogenous enhancement. The lesion consisted of a solid and a cystic component. The cystic component appears to be slightly hypointense on the T1-weighted image, and hyperintense on the T2-weighted image. The solid component and cyst wall showed contrast enhancement after gadolinium administration. In multifocal AT/RTs, it is often confused with multiple brain metastasis, but, it can be distinguished from metastasis by typical histologic features.

Histologically, a tumor contain rhabdoid cells, usually with additional, variable components of primitive neuroectodermal, mesenchymal and epithelial cells[6]. The capacity for divergent differentiation and the presence of rhabdoid cells led Rorke et al. to coin "atypical teratoid/rhabdoid tumor" (AT/RT) as a descriptive diagnostic term[5]. The typical rhabdoid cell is medium-sized, round to oval, with an eccentric nucleus that commonly has a prominent nucleolus[8]. The mesenchymal component of AT/RT is characterized by cells with eosinophilic paranuclear inclusions of intermediate filaments. However, some tumors also contain spindle-shaped cells in fascicles or more loosely arranged. The neuroepithelial component is primitive, consisting of sheets of small poorly differentiated

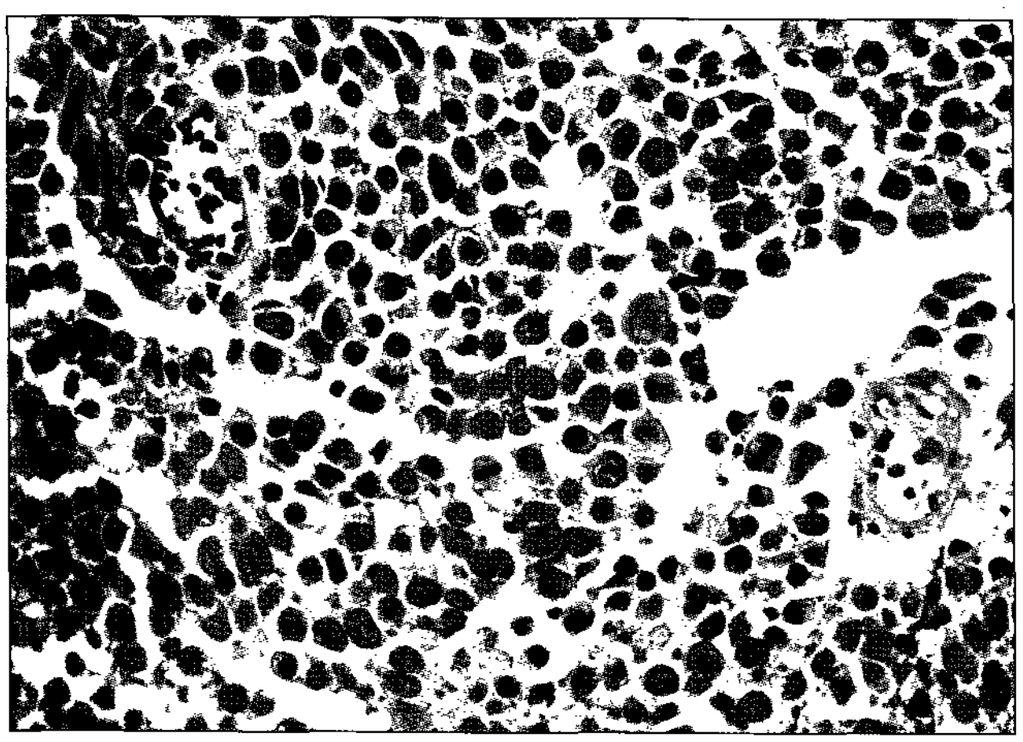

Fig. 3. Typical rhabdoid cells with eccentric nuclei and prominent nucleoli(H & E, ×200).

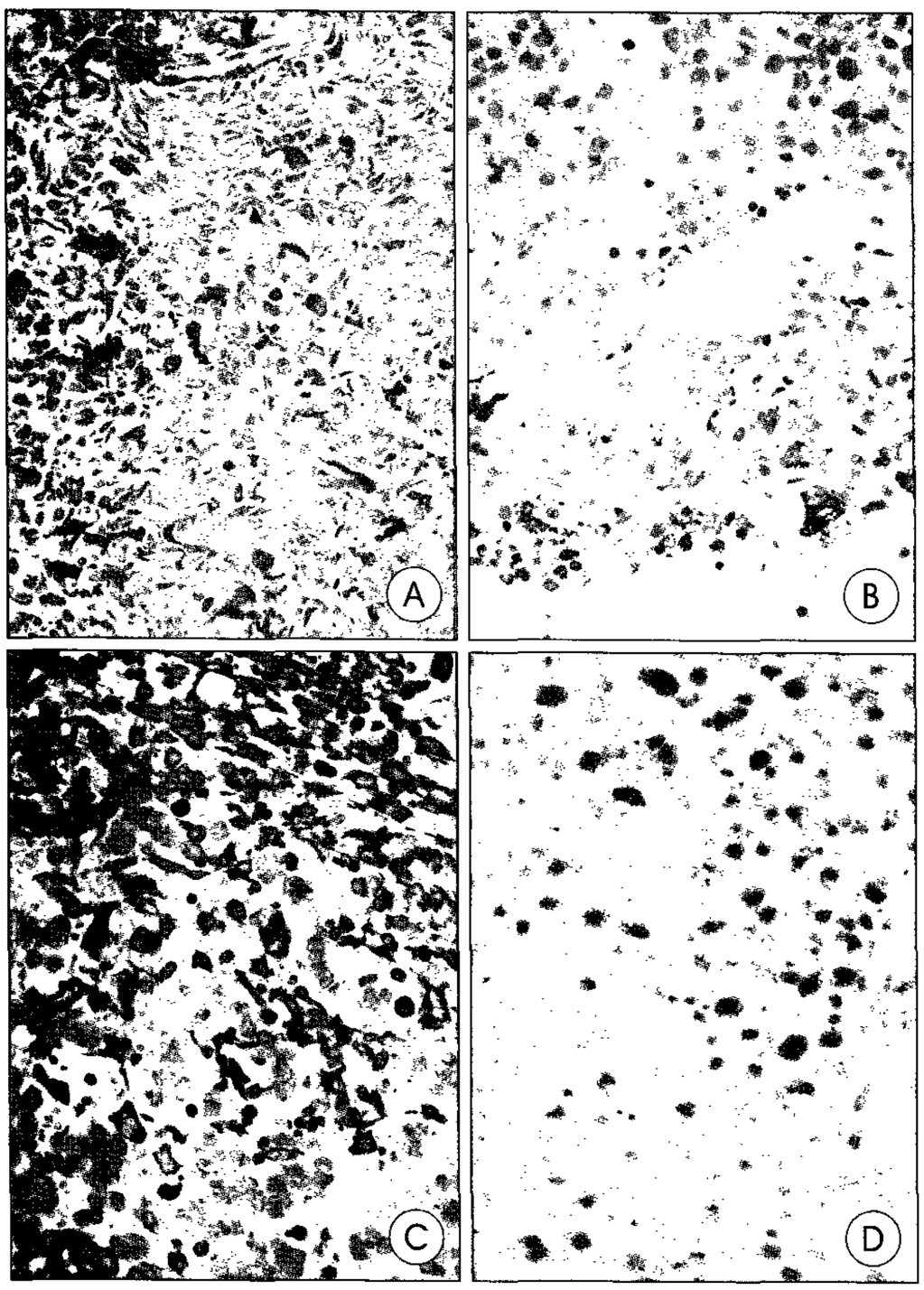

Fig. 4. Immunohiotochemical stains in tumor cell. A : glial fibrillary acidic protein (GFAP, ×200) : (+), B : cytokeratin (CK, ×200) : (+), C : vimentin (×200) : (+), D : epithelial membraqne antigen (EMA, ×200) : (+).

cells. Epithelial cells are poorly differentiated, squamous or adenomatous. There were many typical rhabdoid cells with pinkish cytoplasm and eccentric nuclei and prominent nucleoli in our case. Immunohistochemical investigation is most helpful in diagnosis. The three antibodies of greatest value are vimentin, epithelial membrane antigen(EMA) and smooth muscle actin(SMA). The rhabdoid cells almost always express EMA, vimentin, and SMA in various degrees[2]. In author's case, the tumor expressed glial fibrillary acidic protein(GFAP), cytokeratin(CK), vimentin, epithelial membrane antigen(EMA), so we can confirm AT/RT.

According to a review of reported patients in the literature by Elizabeth Weiss et al, mean survival for all AT/RT patients was 11months[12]. Patients with subarachnoid tumor spread and a primary infratentorial tumor location survived only 2.5months on average. Because of a high risk of local relapse and subarachnoid dissemination, AT/RT of the brain has an unfavorable prognosis. In our case, the patient was dead 1month after tumor resection due to local relapse.

Conclusion

This report provides additional evidence documenting the existence of AT/RT in adults with multiple lesions. Multifocal AT/RTs are rare and often confused with multiple brain metastasis, but, it can be distinguished from metastasis by typical histologic findings. To confirm AT/RT, immunohistochemical investigation is most helpful.

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