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Case Reports & Case Series

Primary atypical teratoid/rhabdoid tumor of spinal canal in a child: Case report and the literature review

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ABSTRACT

Atypical teratoid/rhabdoid tumor (AT/RT) is a rare embryonal tumor of the central nervous system (CNS), which accounts for approximately 1–2% of all CNS neoplasms. Spinal canal is extremely rare. To our knowledge, there are only 16 cases of spinal AT/RT in children reported in English literature so far. Here we first presented a rare primary spinal AT/RT in a child with multiple vertebral subarachnoid metastasis. She was accepted surgical treatment and the diagnosis was established pathologically. She died 3 months later with no further therapy. We discuss the clinical presentation, imaging, pathology, treatment and prognosis of spinal AT/RT in children with the literature review.

1. Introduction

Atypical teratoid/rhabdoid tumor (AT/RT) is a rare embryonal tumor of the central nervous system (CNS), which accounts for approximately 1–2% of all CNS neoplasms [1]. It is most commonly occurring under 3 year old but rare in adult. Even with multimodal therapy recently, the prognosis is still dismal with a median survival time less than 9 months. AT/RT has been reported in all CNS locations, mainly supratentorial and infratentorial, spinal canal is extremely rare. To our knowledge, there are only 16 cases of spinal AT/RT in children reported in English literature so far. Here we first present a primary spinal AT/RT in a child with multiple vertebral subarachnoid metastasis.

2. Case report

2.1. History and examination

A 3-year-old girl, presented with back pain and weakness of lower limbs for 1 week. Her prenatal period, birth and development were normal. She was diagnosed with a hydrocephalus 2 months before, a ventricular peritoneal shunt was carried out and the hydrocephalus was relieved. On this admission, physical examination revealed the decreased motor and sensation of lower limbs, disturbance of the urinary and defecation function was observed at the same time.

2.2. Neuroimaging

The magnetic resonance imaging (MRI) scan of the whole spinal canal revealed multiple round-shaped lesions located intradural subarachnoid space of the spinal canal, while the scan of the brain was only postoperative change of hydrocephalus, no tumor was found. Intraspinal lesions exhibited both hypointensity or isointensity on T_2WI and T_1WI, and heterogeneous enhancement after injected gadolinium (Fig. 1A–C). The diagnosis of a malignant tumor with multiple vertebral subarachnoid metastasis and the spinal cord compression were established.

2.3. Operation

Then the patient underwent a laminectomy via posterior median approach for partial tumor resection and spinal canal decompression. The boundary between the tumor and the spinal cord was not clear in the operation, so the subtotal resection was achieved. The nerve root pain was relieved and the motor and sensation of lower limbs were improved.

2.4. Pathological findings

The postoperative histopathological examination showed the diversities of the tumor cells, including the present of the characteristic rhabdoid cells, primitive neuroectodermal, epithelial and mesenchymal

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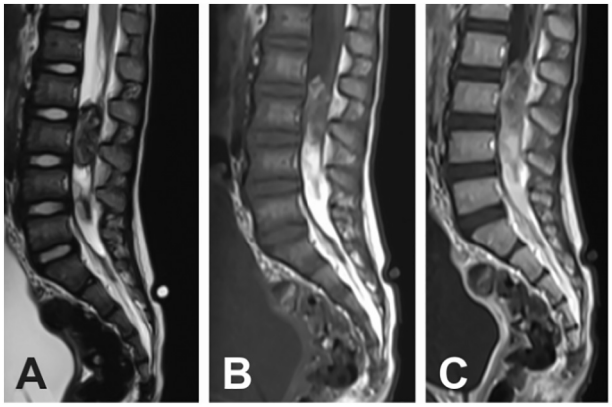

Fig. 1. MRI scanning suggested the multiple lesions in subdural arachnoid space of lumbar spinal canal with (A) low signal in T_2-WI, (B) mixed signal in T_1-WI, and (C) heterogeneous enhancement.

components. Immunohistochemical examination revealed the positive expression of epithelial membrane antigen (EMA), vimentin, glial fibrillary acidic protein (GFAP), neurofilament (NF), and cytokeratin (CK), and some cases were positive for synaptophysin (Syn), smooth muscle actin (SMA), a general germ cell antibody, and desmin. The expression of INI1/SMARCB1 was negative (Fig. 2A–D). The diagnosis of AT/RT was demonstrated pathologically. The patient gave up the further therapy and died 3 months later.

3. Discussion

AT/RT, first described in 1987, as one of the most malignant neoplasms of CNS, is predominantly in infants [2]. It is rare in elder children and adult. It can occur in any site in CNS, most commonly supratentorial, fourth ventricle, cerebellopontine angle [3], originated from cranial nerve also be reported accidentally [4]. However, primary spinal AT/RT in children is extremely rare. We have finished the standard research on PubMed/Medline using terms “Atypical Teratoid/rhabdoid Tumor, Spinal Canal, Child”. To our best knowledge, only 17 cases (include our present case) have been reported in English literature [5–20]. The clinical data of these 17 cases is concluded in Table 1, as the report is mainly single case the accurate prevalence is not clear. There are 11 females and 6 males, the average age is 31.59 months (range: 2 to 108 m). The locations are mainly cervical and thoracic level (82.35%), We believe that It is the first report of our case which demonstrated multiple vertebral subarachnoid metastasis in children.

According to studies of Hirth at al [21], the prognosis of AT/RT from spinal canal is poorer than intracranial, the median survival time is only 6 months. As the prognosis is dismal, the early diagnosis is importance for the effective treatment. Kodama at al [9] found that the irregularly curved annular enhancement of the tumor edge suggested necrosis maybe the typical change of intracranial AT/RT. However, the imaging characteristic of spinal AT/RT has been described nonspecific.

Table 1
Published cases of spinal AT/RT in children.

References	Age (m)	Sex	Location	Treatment	Outcome (m)
Rosemberg et al.	24	F	C6-T1	CMT	DOD/2
Howlett et al.	9	M	T5-T10	Surg/CMT/RT	DOD/2
Tamiya et al.	7	F	T7	Surg	DOD/2
Cheng et al.	24	F	T12	NS	DOD/2
Tanizaki et al.	10	F	T10	Surg/CMT/RT	DOD/3
Bannykh et al.	48	M	T9-L1	NS	NED
Kodama et al.	9	M	C4-T6	Surg/CMT/RT	DOD/20
Moeller et al.	108	M	T11-L2	Surg/RT	NED
Yang et al.	84	M	Lumbar	Surg	NS
Se no et al.	5	F	C4-T3	Surg/CMT	DOD/2
Yano et al.	21	F	Cervical	Surg/CMT/ABMT	DOD/4
Tinsa et al.	48	F	T1	NS	DOD/2
Niwa et al.	72	M	C3-C6	Surg	NS
Fridley et al.	13	F	Cervical	Surg/CMT	DOD/4
Stabouli et al.	2	M	C1-C5	CMT	DOD/6
Shiflett et al.	17	F	Cauda equina	None	DOD/1
This case	36	F	Spinal canal	Surg	DOD/3

ABMT, autologous bone marrow transplantation; CMT, chemotherapy; DOD, dead of disease; F, female; M, male; NED, no evidence of disease; NS, not specified; RT, radiation therapy; Surg, surgery.

The early diagnosis is difficult, the confirmed diagnosis is depending on pathology. According to the cases reported, the signal intensity characteristics of this tumor are usually heterogeneous on pre-contrast images as well as post-contrast images, probably because of necrosis, hemorrhage, cysts, and/or calcification, our case also has the same imaging change. So it is import to suspect the diagnosis of AT/RT in a spinal tumor, especially in infant patient.

The differential diagnosis of spinal AT/RT should include primitive neuroectodermal tumor (PNET), astrocytoma, ganglioglioma, especially PNET. As the imaging characteristic is similar, the confirmed diagnosis maybe the pathology. Kim et al. [22] reviewed 25 PNET and made the comparison with spinal AT/RT, maybe the spinal AT/RT has a younger age and a poorer prognosis. The other differential characteristic is that the leptomeningeal dissemination rate of spinal AT/RT is higher than PNET. As most report described, the rate of leptomeningeal dissemination of spinal AT/RT may achieve 40% [3].

There is no standard treatment for spinal AT/RT, the treatment experience is referred to intracranial AT/RT. The surgical resection is the main therapy, followed by chemotherapy and radiotherapy. However, there is no difference in prognosis between total resection or not, revealed by most studies [5]. Despite aggressive therapy, the prognosis of spinal AT/RT is poorer than intracranial as its malignance, with a median survival time less than 6 months [23]. The death reasons are recurrence and dissemination. The metastasis is usually observed before the clinical manifestation as our case presented. We hypothesis that the hydrocephalus was the result of the reabsorption disorder due to leptomeningeal metastasis. So, if multiple lesions are revealed

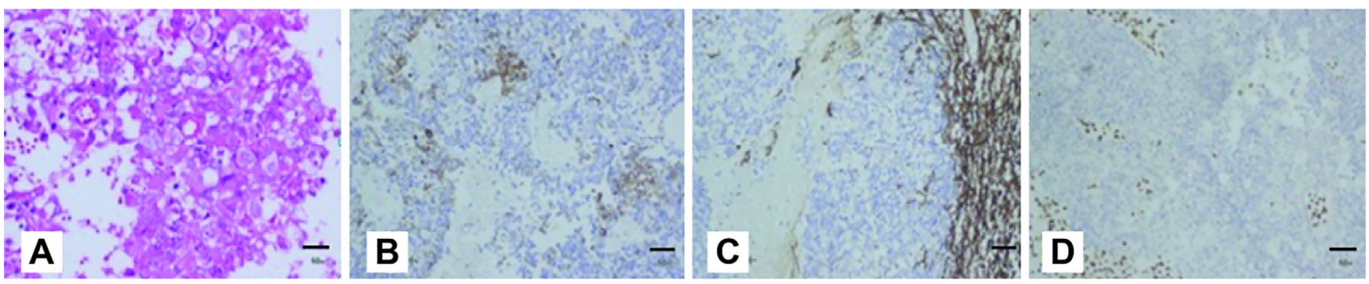

Fig. 2. Pathological examination suggested that (A) the characteristic tumor cells exhibited a rhabdoid morphology in H&E stain, and (B) CK (+), (C) GFAP (−), and (D) INI 1(−) in immunohistochemical examination.

intracranially or intraspinally by MRI in infant or young child, AT/RT should be considered. We also recommend that all patients with spinal AT/RT should have imaging of their brain once diagnosis has been established to exclude leptomeningeal metastasis.

4. Conclusion

AT/RT is a rare tumor of CNS, primary spinal AT/RT in children is extremely rare. The clinical manifestation and imaging characteristic are not special, the demonstrated diagnosis depends on pathology. The early diagnosis and multimodal therapy is important. However, the prognosis is still dismal because of recurrence and early dissemination.

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