Case Report

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Cytologic diagnosis of atypical teratoid rhabdoid tumor based on touch imprint study: Report of a case with review of literature

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Abstract:

Atypical teratoid rhabdoid tumor (ATRT) is a rare malignant tumor with gloom destiny. Our case was a 4-year-old boy with a temporal lobe tumor that was then became evident of ATRT with recurrent happening. In a retrospective review of all cytologic slides, we found unique rhabdoid cells that are morphologically evident cells for ATRT in both times. Unfortunately, the cells were overlooked at the first time. We conclude if the pathologist is experienced to see rhabdoid cells noticing these cells is highly helpful for diagnosis ATRT, especially in frozen sectioning.

Keywords:

Atypical teratoid rhabdoid tumor, cytology, diagnosis

Introduction

Atypical teratoid rhabdoid tumor (ATRT) is a rare malignant tumor of the central nervous system with embryonal origin. Although it typically presents in infants and young children mostly <2 years old, it is reported in older ages even in adults. The most common location is in the cerebellum and cerebellopontine angles, in 50% of cases. Supratentorial location occurs in about 40% of patients.[1]

The fate of patients with ATRT is typically gloom, and the median time of survival has been reported about few months after the initial diagnosis.[2] Radiologic studies are less helpful due to nonspecificity of findings.[3] The definite diagnosis is now made classically on the H and E slides examination, but cytologic studies are newly considered promising procedures for the diagnosis.[4]

The histologic picture consists of rhabdoid and nonrhabdoid large cells and small primitive neuroepithelial cells. Rhabdoid cells, a hallmark of ATRT, are characterized by the distinct cytoplasmic border, hyaline eosinophilic cytoplasm, vesicular, eccentric nuclei with obvious nucleoli with or without fibrillary globoid inclusions.[5] These features can be seen in cytologic and imprint study that can be very helpful in intraoperative diagnosis. Even though the presence of these cells can be overlooked due to the reasons such as first - staining artifact due to proteinaceous background in the lesion that seems over eosinophilic and makes cell borders partially incomprehensible and second - screening error due to clustering or diffusely spreading of rhabdoid that leads to missing during the examination.[4]

The differential diagnoses include medulloblastoma/primitive neuroectodermal tumor (PNET), high-grade glial tumors, and also choroid plexus carcinomas in the aspect of histology, location, and age of the patients.[5]

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These rhabdoid cells are very helpful and pretty unique in diagnosis, especially in the cytologic evaluation of the tumor because primitive cells and epithelial differentiation of them are seen in other tumors. In the treatment of this tumor, surgery, radiation, and chemotherapy may all play a role,[6] but the response to treatments is not usually good.

We report a case of ATRT occurring in the temporal lobe of a 4-year-old boy with recurrence and evaluation of its cytologic studies.

Case Report

A 4-year-old boy, known case of previously diagnosed anaplastic oligodendroglioma of temporal lobe in 11 months ago, came to our Neurosurgery Department with convulsion. The radiologic study showed a mass at the same location of former one in temporal lobe [Figure 1]. The tumor was excised with an intraoperative consultation that got the diagnosis of the high-grade glial tumor. Then, the patient was referred to the Oncology Department for further treatment. The H and E study and afterward immunohistochemical (IHC) examination led us to the diagnosis of ATRT. Histomorphologic sections with H and E staining [Figures 2 and 3] showed a highly malignant tumor consists of cells distributed in a patternless fashion and some areas of necrosis. Some malignant cells had scant cytoplasm, hyperchromatic nuclei with high nucleus-to-cytoplasm ratio, and the others had large polyhedral characteristics. There also some cells with eccentric nuclei (rhabdoid cells) with plentiful eosinophilic cytoplasm, and large nuclei encompassing conspicuous nucleoli. There was a very high mitotic rate, some were in abnormal fashion. For confirmation, IHC study was performed that showed positive reaction for vimentin [Figure 4], epithelial membrane antigen, and cytokeratin [Figure 5]. Stains for glial fibrillary acidic protein and CD34 were negative. Ki-67 was positive in more than 60% of tumor cells.

The total feature of histologic and immunohistochemical was strongly in favor of ATRT diagnosis (World Health Organization Classification, Grade IV). The patient follow-up after 8 months of period revealed his death. We reviewed former set of slides of H and E, frozen sectioning, and touch imprints. Subsequently, we found enough number of typical rhabdoid cells in both samples [Figures 6 and 7]. These cells were overlooked in both times because of the inexperience of the pathologist as a matter of facing to ATRT in cytologic preparation. Retrospectively, it was evident in both cytologic preparations during the intraoperative consultation, and it could be diagnosed at both times by the help of cytologic touch and crush imprints in intraoperative consultation.

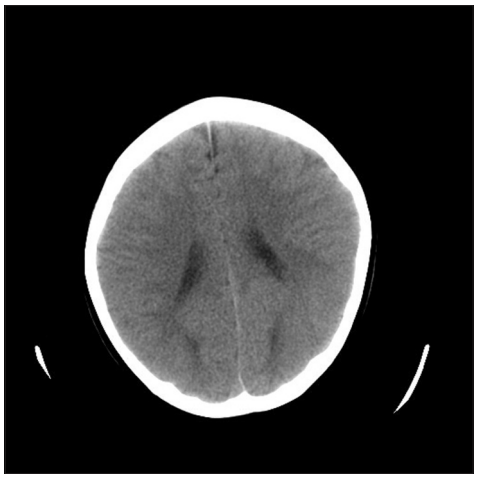

Figure 1: Radiologic study showed a mass at the same location of former one in temporal lobe

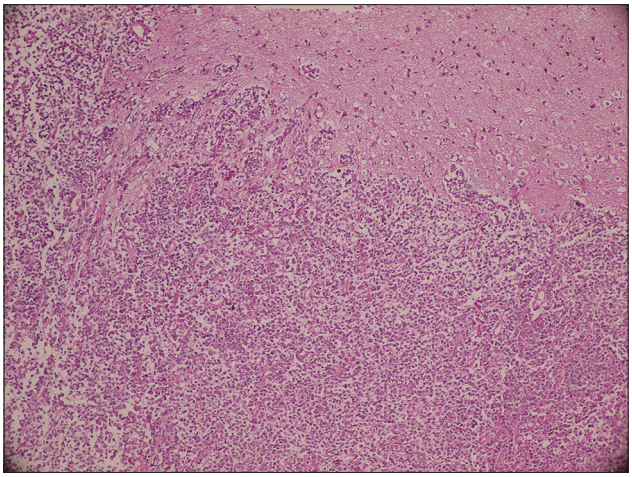

Figure 2: Tumor tissue with normal brain tissue (right upper part). Cellularity is high. Patternless distribution of malignant cells is obvious. (H & E stain, ×100)

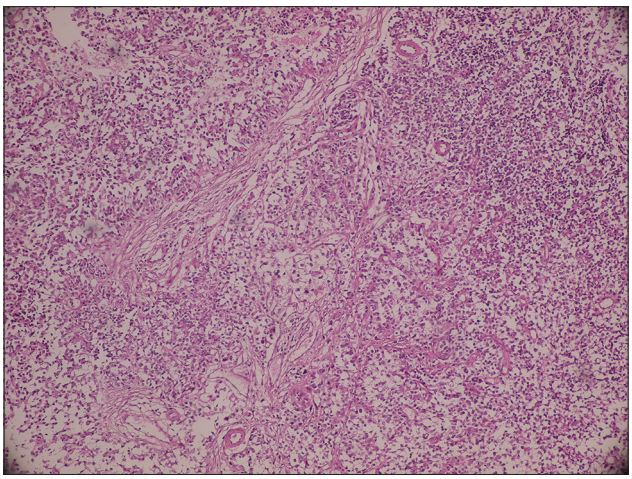

Figure 3: Histomorphologic view of tumor part shows rhabdoid cells beside spotty necrosis. (H & E stain, ×100)

At the first presentation of tumor, the patient brought with a complaint of convulsion with no other mentionable

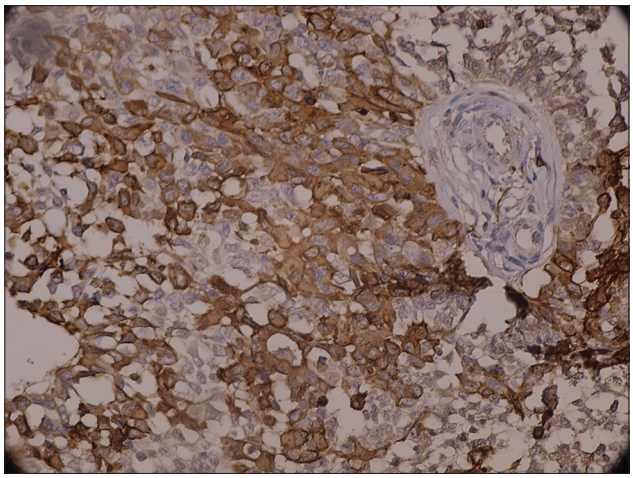

Figure 4: Immunohistochemistry staining for vimentin marker of tumor cells shows positive reaction (H & E stain, ×400)

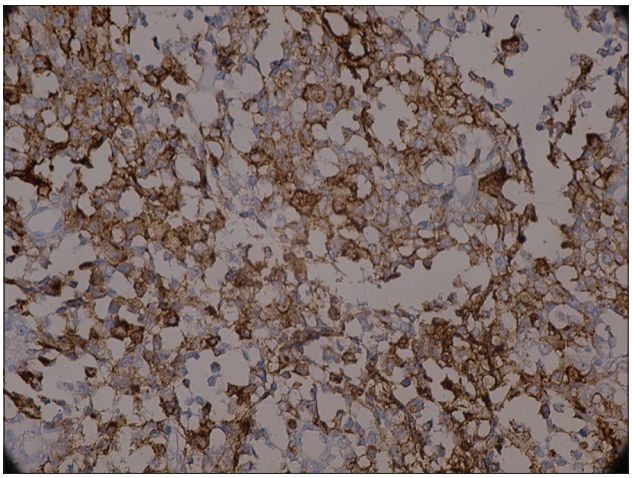

Figure 5: Immunohistochemistry staining for cytokeratin marker of tumor cells shows positive reaction (H & E stain, ×400)

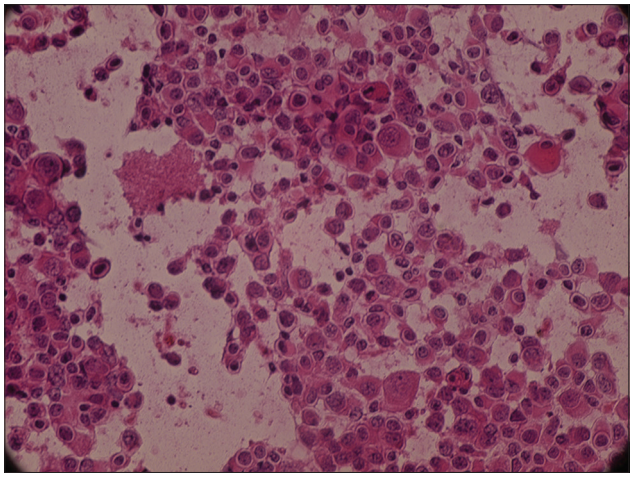

Figure 6: Touch preparation cytology of tumor shows noticeable rhabdoid cells (H & E stain, ×400)

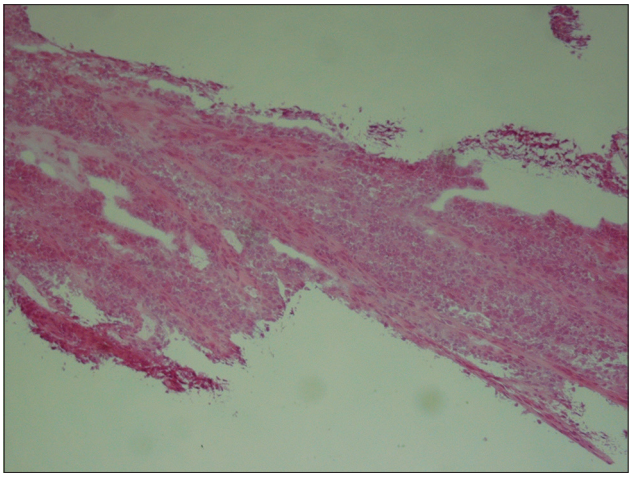

Figure 7: Frozen sectioning of the brain tumor (H & E stain, ×100)

neurologic signs or symptoms. He had stable vital signs without fever. He was mentally alert, complaisant, and had normal cortical functions. Her upper and lower limbs were normotonic.

Radiologic imaging of his brain had been performed and showed a large mass in temporal lobe with solid cystic appearance without significant hemorrhage and minimal peripheral edema and mass effect [Figure 1]. Radiologic differential diagnoses included dysembryoplastic neuroepithelial tumor, pilocytic xanthoastrocytoma, PNET, and ganglioglioma. After excision of tumor, histopathologic diagnosis of high-grade glial tumor most probably oligodendroglioma was wrongly made.

Discussion

ATRT is a high-grade tumor of the brain that has a relationship with other RTs of other sites such as lung and kidney, especially in children.[7] It has an aggressive behavior with high recurrence and mortality rate with median survival rate about 6 months spatially in children under 3 years of age. The most cases of ATRT are seen not only in children <2 years old and in cerebellar and posterior fossa location but also in suprasellar, pineal, and temporal lobe, and overall cerebral hemispheres have been detected.[8] There is also slightly male predominance. It has been mentioned in literature that the diagnosis of primary malignant RT of the brain can be made only histopathologically.[7] ATRT reveals different histomorphological appearances, such as epithelial, mesenchymal, PNET-like, and rhabdoid. Hence, diagnosis is difficult, especially in tiny samples and in intraoperative consultation, where IHC study, especially INI1, is not available. In some studies, it has been stated that imprint cytology expands intraoperative identification.[4,9,10]

In this case report, our patient was a 4-year-old boy and the tumor involved temporal lobe. He experienced

recurrence of the tumor after about 11 months that a confirmation for better survival of patients over 3 years of age.[2,7] Radiologic imaging of the tumor was not that beneficial to help diagnosis. It was not diagnosed in the first place even though unique rhabdoid cells were there. A former pathologic study led to misdiagnosis of ATRT with high-grade glial tumor (oligodendroglioma) that is also rare in these age patients. Then, after about 11 months, we received the specimen of recurrent tumor. After reviewing and reevaluation of the slides of frozen sections, especially touch imprint and then permanent sections, we found typical rhabdoid cells with high mitotic rate that helped us to diagnosis correctly.

Conclusion

A tumor with hypercellular smears while intraoperative consultation with noticeable rhabdoid cells and without other known tumoral pattern must bring us to the diagnosis of ATRT. At the end, we recommend the use of touch imprint cytology for the intraoperative report as an efficient tool for the diagnosis of ATRT.

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Conflicts of interest

There are no conflicts of interest.

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