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Case Report / Olgu Sunumu

Immature teratoma of the posterior fossa in an infant: case report

Bir bebekte posterior fossanın immatür teratomu: olgu sunumu

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Abstract

Teratoma is a rare tumor of the central nervous system that belongs to intracranial germ cell tumors. We report a 2-month-old male child with an immature teratoma of the posterior fossa. Physical and laboratory examinations were normal. Though a radiologic examination was characteristic for this neoplasm, it was insufficient to make a definite diagnosis. Combining the radiologic findings with a histopathologic examination contributed to diagnosing immature teratoma and differentiating it from other subtypes of intracranial germ cell tumors. Our aim was to provide a greater understanding of immature teratoma by reporting this case.

Keywords: Immature teratoma, intracranial germ cell tumor, posterior fossa

Öz

Teratom, merkezi sinir sisteminin nadir görülen bir tümörü olup intrakranial germ hücreli tümörlerden birisidir. Bu makalede, posterior fossanın immatür teratomu bulunan 2 aylık bir erkek çocuğu sunmaktayız. Fizik bakı ve laboratuvar bulguları normal saptandı. Radyolojik inceleme bu neoplazma açısından belirgin bulunmasına rağmen, kesin tanı koymak için yetersizdi. Radyolojik bulguların histopatolojik inceleme ile birleştirilmesi, immatür teratomun tanısına ve immatür teratomun intrakraniyal germ hücreli tümörlerinin diğer alt türlerinden ayırt edilmesine katkıda bulunmuştur. Bu olguyu sunarak, immatür teratomun daha iyi anlaşılmasını sağlamayı amaçladık.

Anahtar sözcükler: İmmatür teratom, intrakraniyal germ hücreli tümör, posterior fossa

Introduction

Teratoma is rare in central nervous system (CNS) tumors, there are different occurrence rates in different nations (1). They mainly occur in young people and the pediatric population (2). Immature teratoma is a subtype of CNS germ cell tumor (3), and tends to be malignant. Immature teratoma is composed of naive mesenchyme, epithelial dysplasia of embryo, and immature primitive nerve tissue. The prognosis of this tumor is not good compared with mature teratoma (4, 5). Thus, a correct and timely diagnosis can contribute to facilitating healthy recovery of patients. Herein, we report a 2-month-old boy with immature teratoma, his clinical symptoms, and physical, radiologic, and histopathologic examinations.

Case

A 2-month-old boy who had been born following a full-term pregnancy and normal delivery and no history of asphyxia after birth, represented with both eyes strabismus to the left for 5 days, with intermittent vomiting. A physical examination was normal. Laboratory examinations were normal.

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Magnetic Resonance Imaging (MRI): Non-enhancement MRI (Figure 1a, b) showed a heterogeneous signal intensity mass in the right side of the posterior fossa. The mass had a well-defined margin, the right cerebellar hemisphere, vermis and brachium pontis were involved. The mass mainly appeared slightly hypo-intense on T1-weighted images and slightly hyper-intense on T2-weighted images with multiple round-like signal intensity within the mass. The fourth ventricle, and aqueduct of the mid-brain and brain stem were clearly compressed and deformed. The supratentorial ventricles were dilated with hydrocephalus. There were patchy slight hypo-in-

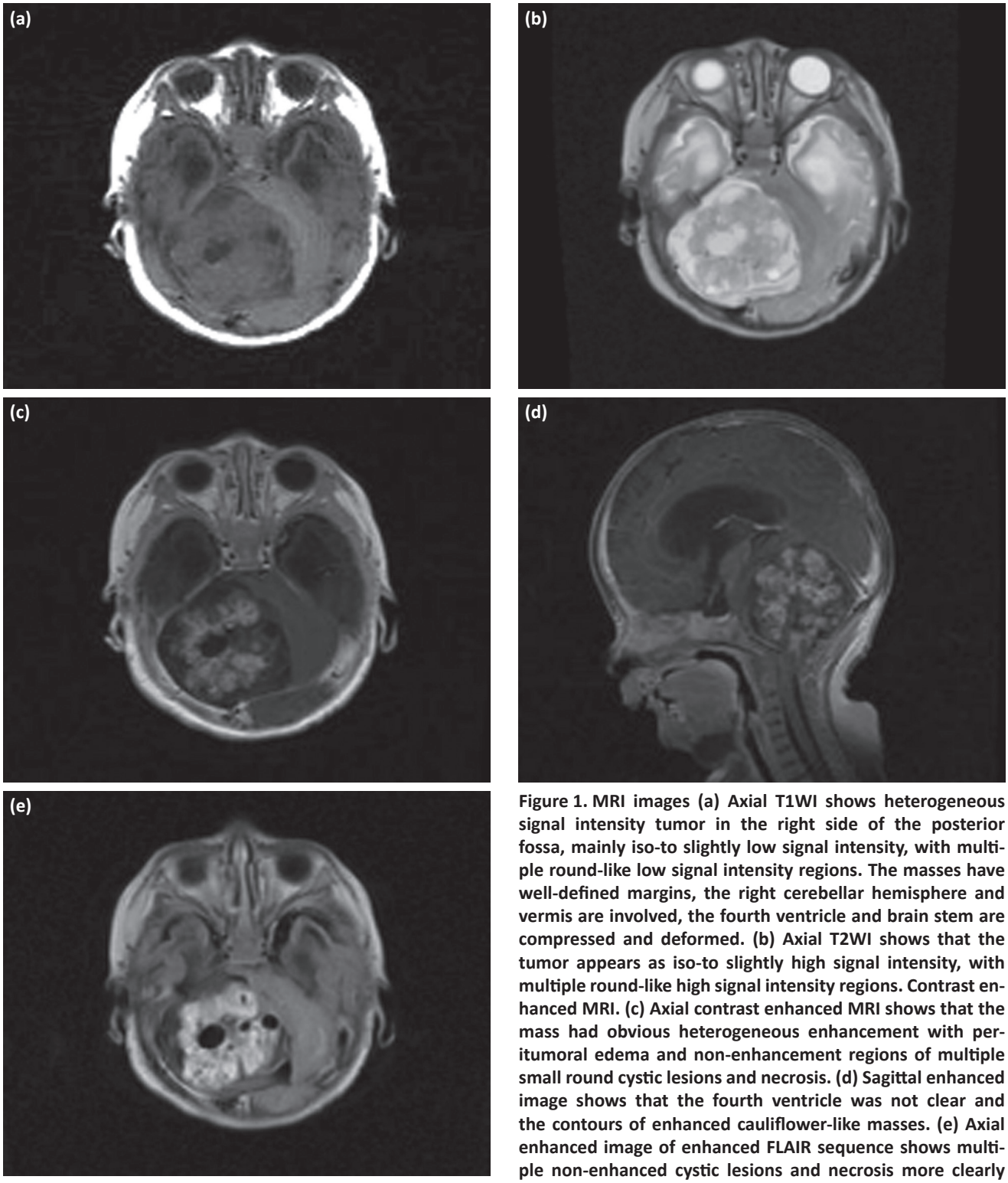

Figure 1. MRI images (a) Axial T1WI shows heterogeneous signal intensity tumor in the right side of the posterior fossa, mainly iso-to slightly low signal intensity, with multiple round-like low signal intensity regions. The masses have well-defined margins, the right cerebellar hemisphere and vermis are involved, the fourth ventricle and brain stem are compressed and deformed. (b) Axial T2WI shows that the tumor appears as iso-to slightly high signal intensity, with multiple round-like high signal intensity regions. Contrast enhanced MRI. (c) Axial contrast enhanced MRI shows that the mass had obvious heterogeneous enhancement with peritumoral edema and non-enhancement regions of multiple small round cystic lesions and necrosis. (d) Sagittal enhanced image shows that the fourth ventricle was not clear and the contours of enhanced cauliflower-like masses. (e) Axial enhanced image of enhanced FLAIR sequence shows multiple non-enhanced cystic lesions and necrosis more clearly

tensities on T1-weighted images and hyper-intensities on T2-weighted images in the white matter around bilateral ventricles with ill-defined margins. Contrast-enhanced MRI scan (Figure 1c–e): The mass showed obvious heterogeneous enhancement, with multiple cystic regions.

Surgical findings: After opening the dura in a "Y" shape, the tumor appeared greyish and looked like "cauliflower" tissues.

Histopathology investigations: Immature teratoma [World Health Organization (WHO) grade III, posterior fossa], contained multiple primitive neural tubes, choroidea, squamous epithelium, appendix organs of skin, cartilage, bone, columnar epithelium, and muscle tissues (Figure 2). Immunohistochemistry: CK (+), EMA (+), 10% Ki67 (+), Syn (+), NF (+), GFAP (+), sporadic LCA (+), some desmin (+).

Written informed consent was obtained from the parents of the patient.

Discussion

Intracranial germ cell tumors (GCT) are a heterogeneous group of tumors that account for about 3–5% of all CNS tumors, two-thirds of which are germinomas (6). Teratoma is rare in the CNS, and they belong to GCTs (3). Teratomas originate from remaining embryonic tissues, which are aberrant because abnormal migrations of primitive germ cells between the third and fifth week of embriyonic development result in remaining germ cells in the midline position. They are pluripotent cells with multidirectional development potential and transform into teratoma in the effect of multiple factors. More than 50% of teratomas occur in the pineal region, 20% in the suprasellar region, and they can also be seen in the posterior fossa, middle fossa, and ventricles (2). They are more common in males than in females, usually in patients aged under 20 years, especially in children (2). The tumor is the most common GCT in infants aged under 1 year, and also the most common intracranial tumor of newborns, accounting for 50% of intracranial tumors in 2-month-old infants (7). Clinical symptoms are lack of specificity, mainly related to the site of the tumor.

Teratoma was classified among GCTs in the 2016 WHO classification of tumors of the CNS (3), belonging to borderline tumors. Teratomas contain tissue components derived from three germ layers (endoderm, mesoderm, and ectoderm), composed of bone, cartilage, hair, fat, lipid, epithelium, muscle, and neural tissues. They are sorted as benign and malignant tumors, depending on the biologic behavior, and no absolute boundary is defined between them. Benign teratomas may transform into

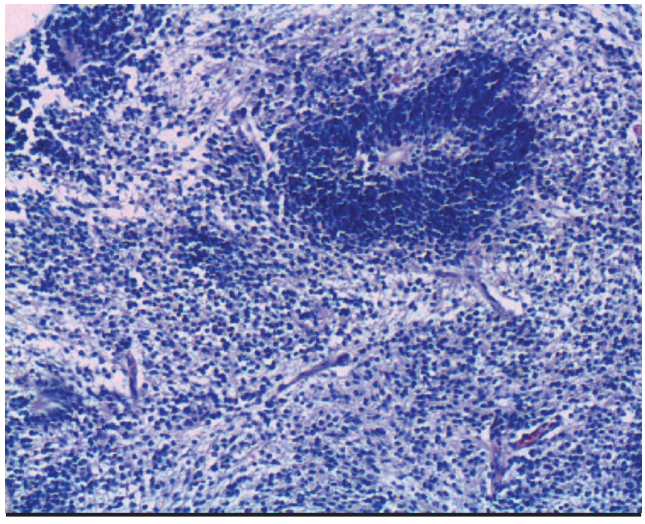

Figure 2. Microscopic examinations of the tumor show many primitive neural tube, choroidea, squamous epithelium, appendix organ of skin, cartilage, bone, columnar epithelium, and muscle tissues

malignant tumors. According to the macroscopic structure, it is classified into cystic and solid tumors, which are incompletely independent of each other. Generally speaking, tumor mainly comprising cystic contents are possible to be benign, whereas tumors that have mainly solid contents are possible to be malignant. In addition, they can be categorized as mature teratoma, immature teratoma, and teratoma with malignant transformation, according to the degree of histologic differentiation. Mature teratomas are benign, the latter two types are malignant. Mature teratomas contain well-differentiated components of ectoderm, mesoderm, and endoderm. Immature teratomas are poorly differentiated. Most teratomas have well-defined margins and integral membranes, and typically appear as round or multilobulated masses. However, malignant tumors have poorly-defined margins with peritumoral edema. Mature teratomas can be diagnosed easily because of their characteristic tissues, such as bone, cartilage, fat, soft tissue; calcification and fat are rare in immature teratomas. The main magnetic resonance imaging (MRI) feature is that the signal intensity of the mass is heterogeneous, associated with calcification, hemorrhage, cystic components, and fat within the mass. On contrast-enhanced MRI, the cystic contents of the tumor are not enhanced and the solid components may or may not be slightly enhanced. Obvious enhancement of a tumor indicates that it is highly malignant.

We reported a case of an immature teratoma in a 2-month-old male infant, the incidence rate of teratoma agreed with the report. The increased intracranial pressure caused by hydrocephalus was mainly due to the mass on the right side of posterior fossa. The compression of the

fourth ventricle, involvement of the right brachium pontis, and compression of the brain stem may have injured the oculomotor nerve.

The differential diagnosis should include other common tumors of the posterior fossa such as medulloblastoma, astrocytoma, and ependymona. The imaging of immature teratomas has no characteristics, thus pathology should be combined to make a final diagnosis. In addition, some authors (4, 5) found that the levels of alpha-fetoprotein (AFP) and human chorionic gonadotrophin (hCG) were increased in serum and/or cerebrospinal fluid of patients with each subtype of germ cell tumors (including immature teratoma), thus assays of AFP and hCG may contribute to the early diagnosis of this disease.

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