Case Report

Congenital Intracranial Teratoma of The Lateral Ventricle With A Diverse Histologic Feature: A Case Report

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Summary

Comprising a very small percentage of all teratomas and all intracranial tumors, intracranial teratomas are extremely rare; however, they are the most common tumors among fetal intracranial tumors. The outcome of these fast-growing aggressive tumors has been reported to be generally poor and the long-term survival rates are seen only in a small number of patients. We present 5-year and 10-month-old girl who was prenatally diagnosed with a congenital intracranial teratoma arising from the left lateral ventricle and treated successfully by several surgical removals and adjuvant chemotherapy during the first year of life. Final histopathologic examination showed a teratoma consisting of mature and immature tissue elements at the resected specimen. A rare histopathologic feature for intracranial teratomas “nephrogenic differentiation” was also noted. Herein, a prolonged clinical course was presented along with the radiologic features and the outcome was discussed by a review of relevant literature in a congenital intracranial teratoma case.

Key words: Congenital; intraventricular; teratoma; nephrogenic differentiation

Lateral Ventrikülün Nadir Bir Histolojik Görünümü Sahip İntrakraniyal Teratoması: Bir Olgu Sunumu

Özet

Tüm teratoma ve intrakraniyal tümörlerin küçük bir bölümünü oluşturan intrakraniyal teratomalar, oldukça nadir ancak fetal hayatta en sık rastlanılan intrakraniyal tümörlerdir. Hızlı büyüyen bu agresif tümörlerin prognozu genel olarak kötüdür ve uzun dönem yaşam oranları yalnızca sınırlı sayıda hasta izlenir. Biz, sol ventrikül kaynağılı "konjenital intrakraniyal teratoma" olarak prenatal dönemde tanı edilmiş, birden fazla cerrahi operasyon ve adjuvan kemoterapi ile yaşamının ilk yılında başarılı olarak tedavi edilmiş 5 yaş 10 aylık bir kız çocuğunu sunuyoruz. Son histopatolojik tanı, "matür ve immatür doku elemanlarından oluşan teratoma" olup, intrakraniyal teratomalar için nadir bir histopatolojik görünüm olan “nefrojenik diferansiyasyon” tespit edilmiştir. Bu olgu bildirisinde uzun klinik süreç; radyolojik görünüm özellikleri beraberinde sunulmuştur ve klinik sonuç ilgili literatürün gözden geçirilmesiyle birlikte tartışılmıştır.

Anahtar Kelimeler: Konjenital; intraventriküler; teratoma; nefrojenik diferansiyasyon
INTRODUCTION

Congenital intracranial teratomas are relatively rare tumors. They consist of only 2-4% of all teratomas; however, they compose one third to one half of all congenital central nervous system (CNS) tumors and are referred to as the most common intracranial tumors in fetuses and neonates. Although teratomas may occur in several different locations, intracranial teratomas involve the midline brain structures preferentially. Prenatal ultrasonography (US) can easily detect these tumors. Magnetic Resonance Imaging (MRI) is the modality of choice in defining of tumor location and extension both in fetuses and newborns. It is well known that normal brain structures can be rapidly replaced by these fast-growing tumors. Furthermore, they may extend into extracranial structures. The prognosis of intracranial teratomas is generally poor due to a rapid growing mass, even though they have been diagnosed prenatally. The success of surgery is limited. Due to the potential complications of surgical procedure, a successful outcome can only be achieved in the limited number of cases depending on size and extension of tumor at the time of surgery. In this report, a case of a congenital intraventricular teratoma is presented along with lengthy follow-up results.

CASE PRESENTATION

A complex intracranial mass was detected in a female fetus on the second trimester ultrasonography (Fig. 1A). The presence of the complex mass was also confirmed on the fetal MRI (Fig. 1B). The pregnancy history was uneventful. The fetus was born via vaginal delivery with an Apgar score of 9/9/9 at 38 weeks of gestation determined by the last menstrual period. After the birth, neonatal brain MRI was conducted and it once again revealed a large, expansile, lobulated mass measuring 7 X 5 X 6-cm that originates from the left lateral ventricle extending into the periventricular white matter and the left thalamus. Mixed signal intensities with speckled signal void areas of calcifications within the mass were shown on both T1 and T2-weighted images (Fig.1C) and a heterogeneous enhancement was noted on post-contrast T1-weighted images. Diffusion Weighted Imaging displayed diffusion restriction. Axial computerized tomography (CT) images showed focal fat densities and calcifications within the mass (Fig. 1D). On the fifth day of life, surgery was performed for removal of the mass. Final histopathologic diagnosis was stated as "teratoma with mature and immature elements". Two more tumor resections were performed due to residual tumor in one and seven months after the first surgery, respectively. Residue tumor was revealed as an enhancing mass on both CT and MR images (Fig. 2). Along with mature and immature components within the tumor, kidney tissue was noted to be a finding rarely reported in teratomas, (Fig. 3A, Fig. 3B). The fast-growing residual mass caused obstructive hydrocephalus and a ventricular shunt catheter was placed at the time of second surgery. Adjuvant chemotherapy was given to lower the risk of recurrence. After the third surgery, no residual tumor was shown on MR images. Follow-up MRI showed postoperative signal changes in the surgical bed with mild ventricular dilatation. She developed remote symptomatic refractory left hemispheric seizures 18 months after the third surgical operation and she was loaded on the antiepileptic treatment. The last MRI was conducted at the age of 5 years and 10 months (70 months) and revealed mild to moderate enlargement of sulci consistent with surgical tissue loss and atrophy on the left hemisphere (Fig. 4). She attends school with specialized classes, making progress with. She has right-sided weakness due to the left hemispheric tissue loss. Growth percentiles based on "CDC 2-20 Years Data Chart" (Developed by the National Center for
Health Statistics in collaboration with the National Center for Chronic Disease Prevention and Health Promotion (and the bone age were found to be appropriate with her chronological age.

Fig.1A: The second trimester ultrasound image shows large, heterogeneous echogenic mass predominantly in the left lateral ventricle (arrows). Fig.1B: A large, heterogeneous intraventricular mass (arrows) with hydrocephalus shown on Fetal MRI (axial T2-weighted image). Fig.1C: Coronal T2-Weighted MR image shows multicystic areas and mixed signal intensities in complex, heterogenous mass centered predominantly in the left lateral ventricle. The cystic components reveal extension to the left cerebral hemisphere (arrows). Markedly low signal intensities in some of cystic structures are suspicious for blood products. Fig.1D: Preoperative non-contrast head CT image shows fat and calcifications (arrows) in the mass.

Fig.2: Postoperative T1-Weighted post-gadolinium image shows hydrocephalus and heterogeneously enhanced residue mass (arrows) measured 3 cm in diameter.
DISCUSSION

The term “congenital tumor” refers to tumors presenting within fetal life and the first 2 months of life[1]. Congenital teratomas are thought to have developed from the “misplaced” embryonic germ cells embedded in the head, mediastinum, or sacrococcygeal region during formation of the primitive streak in the 3rd week of life[13,16]. All teratomas show an incidence of 1:4000 live births, whereas congenital intracranial teratomas consist of 0.5–1.9% of all pediatric tumors[15]. Despite this relatively rare incidence, teratomas are the most common congenital intracranial tumor in fetus and newborn[3]. Congenital intracranial teratomas commonly originate from the midline structures of supratentorial region and the majority occurs in suprasellar region. Contrary to the congenital counterpart, intracranial teratomas frequently arise from infratentorial structures[13,16]. The pure intraventricular location is quite a rare entity and very few cases reported in the literature[4,6].
Teratomas are germ cell tumors characterized by excessive growth of multiple tissues that are normally not found at the site of involvement. These tissue components are often derived from two or all of three germinal cell layers; ectoderm, mesoderm and endoderm. According to "WHO Classification of CNS Tumors", all teratomas are histologically classified as "mature", "immature" or "teratoma with malignant component". The histologic features of congenital teratomas show some differences compared to those seen in adults. Congenital teratomas commonly compose of immature elements. Those elements do not reflect poor prognosis contrary to teratomas occurred later in life. In fetal teratomas, ectodermal and mesodermal tissues are predominantly found. Endodermal components that mostly consist of respiratory epithelium and gastrointestinal tissues are the least common\(^{(13)}\). Nephrogenic differentiation formed by renal glomeruli has rarely been reported in the literature. Jain et al. presented the nephrogenic differentiation in two spinal teratoma cases\(^{(9)}\). To our best knowledge, it has not been reported previously for intracranial teratomas.

Prenatal ultrasonography and fetal MRI can easily recognize intracranial teratomas with their complex heterogeneous appearance. The prenatal diagnosis can be usually made as early as the 27\(^{th}\) weeks of gestation. Those that are detected before 24 weeks of gestation constitute 10 % of all cases\(^{(14)}\). MRI is the best modality in defining of the tumor location and extension both in fetuses and newborns. The characteristic MR feature of an intracranial teratoma is commonly a large, heterogeneous mass with cystic components\(^{(1)}\). Various elements such as fat, calcification, cystic components, soft tissue structures may lead to heterogeneous signal characteristics on T1- and T2-weighted images without obvious differences between mature and immature teratomas. Contrast-enhanced studies reveals heterogeneous enhancement of the mass. Similar imaging characteristics are shown on CT, as well. CT has an advantage in detecting of small calcifications compared to MRI.

These tumors have a tendency to present with larger tumor size and the massive tumor may invade extracranial structures. Obstructive hydrocephalus and macrocephaly are frequently seen caused by the massive tumor and a cesarean section for delivery may be required due to enlarged fetal head. The majority of these cases have resulted in stillbirth or perinatal death. Cardiovascular or respiratory failure may occur as a result of compression or replacement of brainstem or other brain tissues and these cases mostly conclude in stillbirth or death just following the birth. Location and size of the tumor seem to be more important than the malign potential in foreseeing the outcome. The larger size and propensity to recurrence poorly affect the prognosis. Depending on the tumor size and extension, their pseudo-encapsulated distinct margins may allow to complete resection and the outcome can be satisfactory in the limited number of patients. The complete resection with chemotherapy may delay the regrowth\(^{(8)}\). In the present case, it might be thought that relatively far location from brainstem structures could be a good prognostic factor allowing to the near to complete surgical resection and following the third resection, adjuvant chemotherapy might have helped to prevention from recurrence. Another important point to mention is that vaginal delivery did not lead to serious complication in this case.

According to some authors, radiation therapy is not recommended due to the potential risk of severe damage in developing CNS and does not offer a consistent advantage for unresectable tumors\(^{(3,17,18)}\). In a series of intracranial germ cell tumors, the 10-year survival rate of mature teratomas, which have undergone total or subtotal resection, was 92.6%. In the same series, the overall 10-
The year survival rate of malignant teratomas was 70.7% and the authors indicate that extensive removal is an effective way when surgery is followed by postoperative radiation therapy and chemotherapy\(^{(12)}\).

The dissemination potential of immature teratomas should always be considered and included in follow-up planning\(^{(5,12,14)}\). \(\ddot{I}\)şik et al. reported that the prognosis of congenital intracranial teratomas was poor and death usually occurs shortly after birth; however, there were also some rare reports such as prolonged survival up to 3.5 years after tumor resection\(^{(8)}\). Compared to the reported cases in literature, our patient's age is older. Shih-shan et al. reviewed one hundred twenty-three papers for their study of "surgical treatment of brain tumors in infants less than 6 months of age". They have concluded that teratoma is the most common tumor among tumors seen in the first 6 months of life and approximately 83.7% of all congenital intracranial teratoma patients died. Although, imaging studies may help to identify and distinguish potentially curable lesions such as choroid plexus papillomas, Shih-shan et al. stated that predicting of the tumor type is a challenge based on the preoperative imaging and the prognosis of many infantile tumors remains poor.

Surgery provides a way to establish a certain diagnosis and a complete treatment for some intracranial teratoma cases. The outcome frequently depends on the extent of resection, and/or response to the adjuvant therapies. Nevertheless, the prognosis is overall poor contrary to the longest survival seen in choroid plexus papillomas and astrocytomas\(^{(2,10)}\).

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