Midline Posterior Fossa Teratoma

—Case Report—

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Abstract

A 20-day-old female neonate presented with an immature teratoma in the midline posterior fossa. The tumor was totally removed but the patient died of pneumonia. Teratoma is a rare tumor, but very difficult to treat as the patients tend to be young, and the outcome is very poor.

Key words: posterior fossa, immature teratoma, hydrocephalus

Introduction

Intracranial teratoma has an estimated incidence of 0.3% to 0.6% of all intracranial neoplasms.²⁾ Teratomas are true congenital tumors, and are composed of tissue derived from all three germ cell layers.³⁾ Teratomas commonly present during infancy and childhood, and account for 2% of intracranial tumors in patients under the age of 15 years.⁷⁾ Teratoma in the posterior fossa is extremely rare. We treated a 20-day-old female with a midline posterior fossa immature teratoma.

Case Report

A 20-day-old female neonate presented with a 10-day history of rapid head enlargement, vomiting, refusal to feed, and irritable behavior. The neonate was delivered by cesarean section at full term, because of her large head. On examination she was irritable, emaciated, and had a low body weight. She had a bulging anterior fontanelle and visibly dilated scalp veins. The head circumference was 46 cm with prominent occiput. Computed tomography of the brain showed a large midline posterior fossa tumor (Fig. 1). The fourth ventricle was not separate from the tumor. The tumor was isodense and enhanced homogeneously after contrast medium administration. Supratentorial hydrocephalus was present with periventricular lucencies.

The tumor was excised totally through a midline

Fig. 1 Axial computed tomography scan with contrast medium showing a large midline posterior fossa teratoma with dilated lateral and third ventricles.

suboccipital craniectomy. The tumor occupied the fourth ventricle. The tumor was solid with varying consistency and was moderately vascular. The brain stem was not invaded by the tumor. The surgery was uneventful. The patient developed pneumonia and died on the 7th postoperative day.

Histological examination of the tumor showed the features of immature teratoma. The tumor was

Fig. 2 Photomicrograph of the tumor specimen showing cuboidal epithelial lining and cartilaginous tissue. HE stain, original magnification $\times 160$.

Fig. 3 Photomicrograph of the tumor specimen showing ependymal lining, papillary projections, and neuroglial tissue. HE stain, original magnification $\times 64$.

composed of highly cellular immature tissue with components from the three germ cell layers. The cells were rounded with scanty cytoplasm and hyperchromatic nuclei, and resembled primitive neuroectodermal cells. There were islands of cartilage, glandular epithelium, and tubular structures resembling neuroepithelial cells. There were no areas of hemorrhage or necrosis (Figs. 2–4). Autopsy showed ventriculomegaly. There was no evidence of posterior fossa tumor.

Discussion

Intracranial teratomas commonly originate near the midline, which is a location with great potential for misplacement of embryonal tissues.⁶⁾ Intracranial teratomas are common in the suprasellar and pineal region,¹⁾ and also occur in the cerebellopontine angle and the fourth ventricle.¹¹⁾ Malignant teratomas characteristically demonstrate elevated levels of alpha-fetoprotein and beta-human chorionic gonadotrophin in the cerebrospinal fluid.⁸⁾ Distant metastasis is common with malignant immature teratomas. The lung is the commonest site of metastasis from primary malignant intracranial teratomas and metastasis through the cerebrospinal fluid pathway has also been recorded.¹³⁾

Teratomas can be classified histologically into mature and immature types. Mature teratomas are benign in nature and contain well-differentiated tissues. Immature teratomas, as in our case, are composed of immature and undifferentiated fetal tissues, and usually undergo malignant transformation so are occasionally referred to as malignant immature teratoma.³⁾ Twenty cases of midline posterior

Fig. 4 Photomicrograph of the tumor specimen showing tubular structures lined by immature neural tissue. HE stain, original magnification $\times 160$.

fossa teratomas have been reported, 13 cases were benign mature teratomas and six cases were immature malignant teratomas,^{5,14)} with one case not known.⁵⁾

Intracranial teratomas have three presentations in neonates.^{3,6,7)} The first type is teratoma in patients who are either still born or die immediately after birth. These tumors are quite large and the brain is completely replaced by the teratomatous growth. The second type, as in our case, is teratoma in patients who are born with an enlarged head, who survive the delivery and the tumors are relatively small at birth. The third type is teratoma in children who are normal at birth, but experience rapid head enlargement during the early weeks of life. These

patients harbor the smallest tumors.

Only one of 73 cases of intracranial teratomas occurring in the neonatal period was infratentorial.¹⁶⁾ The first teratoma located in the midline posterior fossa was reported in 1912.⁹⁾ Since then, 20 cases of midline posterior fossa teratomas have occurred, with a predominance of females, compared to the male predominance found in teratomas of the pineal and suprasellar regions.⁵⁾ Our case, a 20-day-old neonate, is the youngest patient so far to have such a lesion.¹⁴⁾

Teratomas are difficult to treat because of the usually large size at presentation and young age of the patient. Only five of 13 patients with mature teratoma were alive during follow up.⁵⁾ Radical excision of the tumor, although difficult, is the ideal treatment of intracranial teratoma.⁴⁾ Adjuvant therapy using radiotherapy and chemotherapy is recommended in immature intracranial teratomas.^{4,15)} However, the use of radiotherapy treatment is restricted in young patients. Irrespective of the treatment, the prognosis of the immature teratomas is poor.¹⁰⁾ All seven patients with infratentorial immature teratomas including our case died after radical surgical excision of the tumor.⁵⁾ A one-month-old female neonate died on the first postoperative day because of hypotension.¹²⁾ A two and half-year-old female died 3 months later due to tumor recurrence.¹¹⁾

Immature teratomas in the posterior fossa occur at a very young age. The clinical outcome of these patients is poor, because of the age of presentation, malignant nature of the tumor, and limited role of adjuvant therapy.

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