

Fetal tumors of the choroid plexus: is differential diagnosis between papilloma and carcinoma possible?

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KEYWORDS: choroid plexus carcinoma; choroid plexus papilloma; intracranial tumor; prenatal diagnosis

ABSTRACT

Fetal choroid plexus tumors are uncommon. The prognosis is widely variable and depends on the histological findings: papilloma or carcinoma. We report a case of prenatal diagnosis of choroid plexus mass detected by ultrasound at 33 weeks of gestation. Prenatal (T1, T2, T2 and diffusion weighted sequences) magnetic resonance imaging (MRI) was used to rule out a hematoma. Follow-up examination by ultrasound and MRI revealed a significant increase in the volume of the mass, suggesting a diagnosis of malignant tumor. A healthy neonate was delivered by Cesarean section at 38 weeks of gestation. Full surgical excision of the tumor was performed at 20 days after delivery and histological analysis revealed a papilloma. Copyright © 2011 ISUOG. Published by John Wiley & Sons, Ltd.*

CASE REPORT

We report the case of a 25-year-old primigravida with no history of familial or personal cerebral disease who had a normal pregnancy until the third trimester. At 33 weeks of gestation, the patient was admitted to our unit after abdominal trauma. Detailed ultrasound examination revealed an echogenic mass inside the fetal right cerebral ventricle, in the choroid plexus, measuring $29 \times 20 \times 19$ mm (Figure 1). Doppler examination showed peripheral vascularization. The velocity in the fetal middle cerebral artery was normal (53 cm/s). The right and left ventricular atria measured 11 mm and 6 mm, respectively. This abnormality was isolated.

Magnetic resonance imaging (MRI) sequences without gadolinium injection were performed and confirmed the presence of a solid mass in the choroid plexus, which was slightly heterogeneous, appearing isointense on T1-weighted sequences and isohyperintense on T2-weighted sequences. T2*-weighted images ruled out the presence of hemosiderin deposits, excluding the diagnosis of a hematoma. The observation of spots that were slightly hyperintense on T1-weighted imaging and slightly hypointense on T2-weighted imaging suggested the presence of small perilesional calcium deposits. On diffusion-weighted sequences, hypersignal appeared without diffusion restriction. A hyperintense signal was seen on T2-weighted sequences corresponding to peritumoral edema (Figure 2). These findings led to the diagnosis of choroid plexus tumor.

Two weeks later, examination by ultrasound and MRI revealed significant growth of the tumor, which now measured $36 \times 27 \times 24$ mm. A deviation of midline structures related to a mass effect had become obvious (Figures 1 and 2), as had the presence of major intracranial edema on T2-weighted and diffusion-weighted sequences and of intracranial hypertension shown by the complete absence of pericranial spaces.

Considering the growth potential of the tumor, the patient was informed of the high risk of a malignant tumor with its associated poor outcome. The patient did not consider termination of pregnancy (TOP) as an option. Labor was induced at 38 weeks of gestation, after checking for cephalic mensuration. A Cesarean section was performed because of fetal heart abnormalities. A healthy male neonate weighing 3310 g, with Apgar scores

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Accepted: 15 December 2010

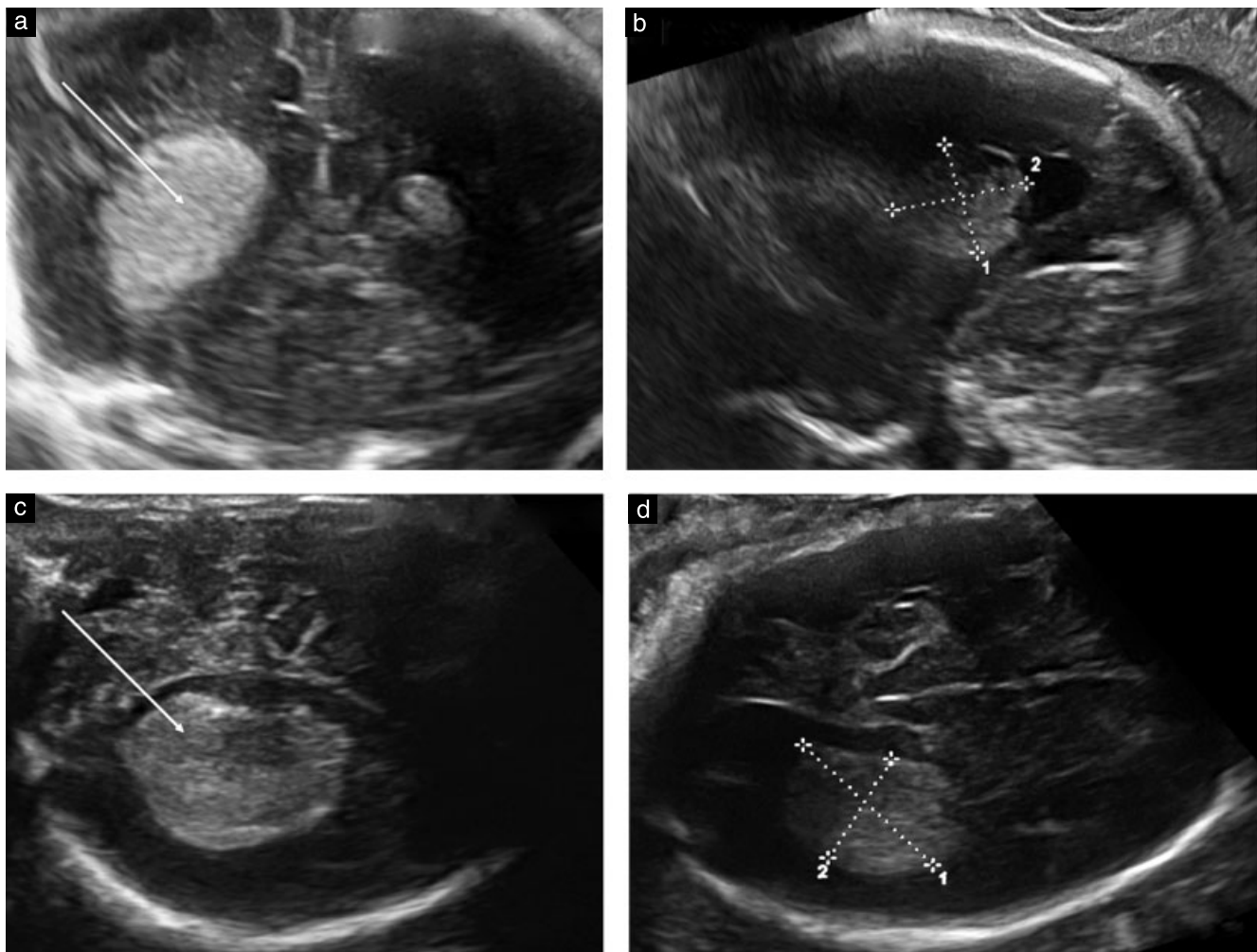


Figure 1 Prenatal ultrasound images. (a) Coronal view at 33 weeks of gestation showing an echogenic mass in the right choroid plexus (arrow). (b) Parasagittal view at 33 weeks of gestation; the tumor measures 29×20 mm. (c) Posterior transverse view at 35 weeks of gestation; the mass appears echogenic and slightly heterogeneous (arrow). (d) Transverse view at 35 weeks of gestation showing that the tumor had increased in size to 36×26 mm with deviation of the midline structures.

of 10 at both 1 and 5 min and cord blood pH of 7.24, was delivered. Neonatal head circumference was 34 cm. Postnatal ultrasound and MRI confirmed massive edema and a further significant increase in tumor volume. After gadolinium injection, marked contrast enhancement of the tumor was observed (Figure 3). Surgery was performed at 20 days of age. A wide frontotemporoparietal opening in the skull and a parietal corticotomy allowed access to the choroid plexus. After ligation of the pedicle, complete tumoral excision was achieved. During surgery, transfusion of more than double the infant's body mass of blood was necessary. The postoperative course was uneventful.

Neuropathological analysis revealed a papillary epithelioma. Immunohistochemistry was used to confirm the diagnosis with cytokeratin 18-positive marking. The cellular proliferation index assessed by MIB-1 immunostaining was approximately 20%. These findings confirmed the diagnosis of a benign tumor of the choroid plexus. At the last follow-up at the age of 1 year, the infant was healthy and MRI findings were normal.

DISCUSSION

Tumor of the choroid plexus is a rare type of intracranial tumor in children, but accounts for 42% of cerebral tumors in neonates¹. They occur in the lateral ventricle in most cases, but have also been described in the third or fourth ventricles². They can correspond to two different etiologies: papilloma, which is a benign tumor, and carcinoma of the choroid plexus. Total removal is possible for 96% of papillomas and for 61% of carcinomas. Surgical procedures are associated with significant operative morbidity and mortality due to uncontrolled bleeding. The prognosis is widely variable between the two forms, with a survival rate at 5 years of approximately 100% for papillomas and 40% for carcinomas³.

Tumors of the choroid plexus have been described previously in the fetus^{4–7}. In two cases of choroid plexus papilloma reported at 21 weeks of pregnancy, the diagnosis was suspected because of ventriculomegaly. Both pregnancies were terminated and pathological examination showed bilateral papillomas of the choroid

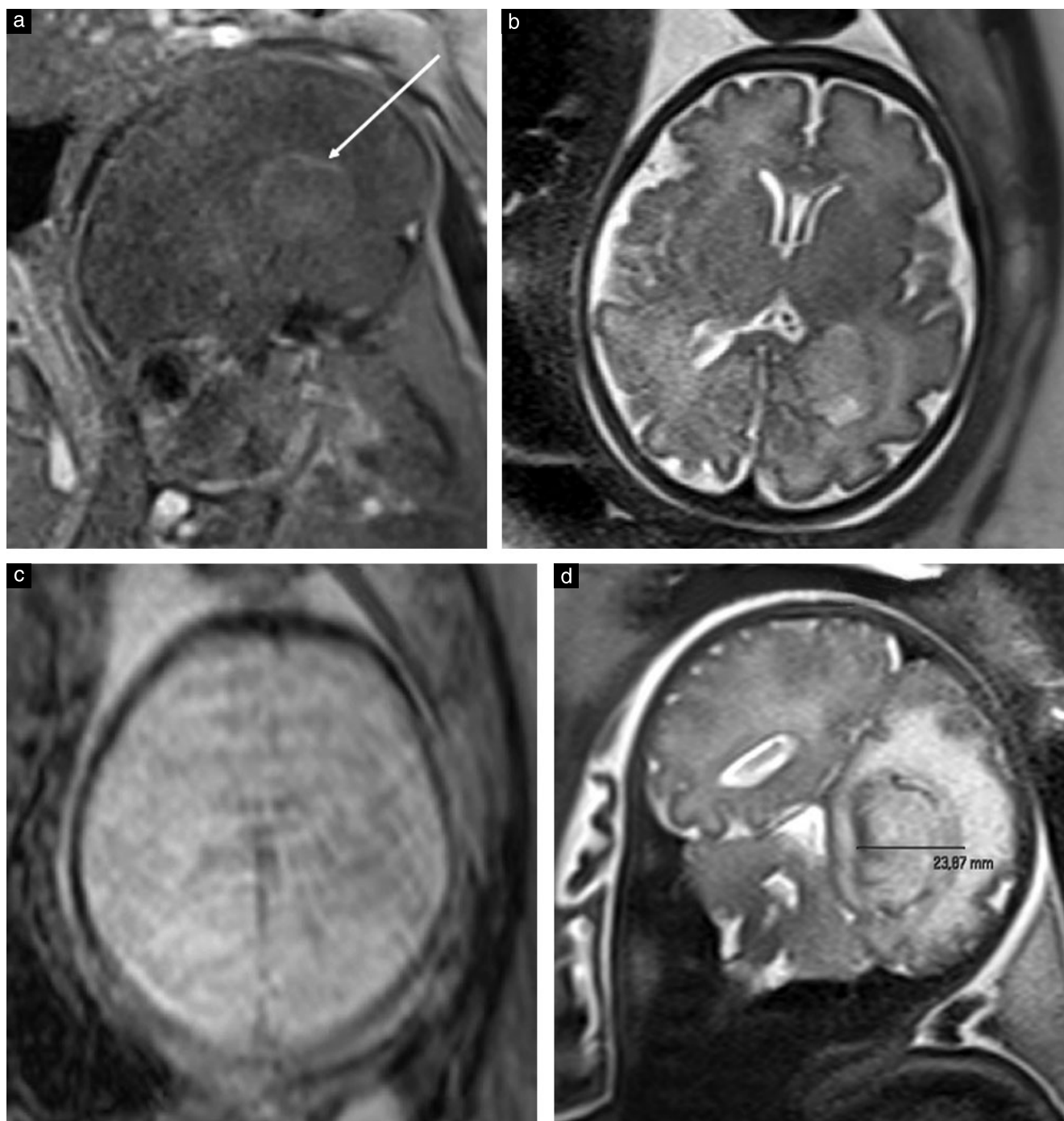


Figure 2 Prenatal magnetic resonance images. (a) Parasagittal T1-weighted image at 33 weeks of gestation showing an isointense mass in the right choroid plexus (arrow). (b) Transverse T2-weighted image at 33 weeks of gestation, the mass appears with hyperintense signal. (c) Absence of signal of hemosiderin on T2*-weighted imaging at 33 weeks of gestation ruling out diagnosis of hematoma. (d) Coronal T2-weighted image at 35 weeks of gestation showing increased tumor size and peritumoral edema.

plexus⁴. Antenatal investigations in these cases were limited, possibly because of legal restrictions regarding the gestational age at TOP.

Antenatal choroid plexus tumors are described most often during the third trimester, presenting as a large homogeneous mass. They can also be revealed by the presence of hydrocephalus or ventriculomegaly because of overproduction of cerebrospinal fluid or obstruction of the foramen of Monro by the mass. Intraplexic

hematoma is a differential diagnosis but may also be a complication of plexus tumors. Barber *et al.* reported findings of a papilloma located in the posterior horn that appeared upon ultrasound examination as a hyperechoic mass corresponding to intracranial hemorrhage, with an irregular surface and perilesional vascularization⁵. In one case, the tumor was considered not accessible for surgery, and autopsy after postnatal death revealed a choroid plexus papilloma with intraventricular hemorrhage⁶.

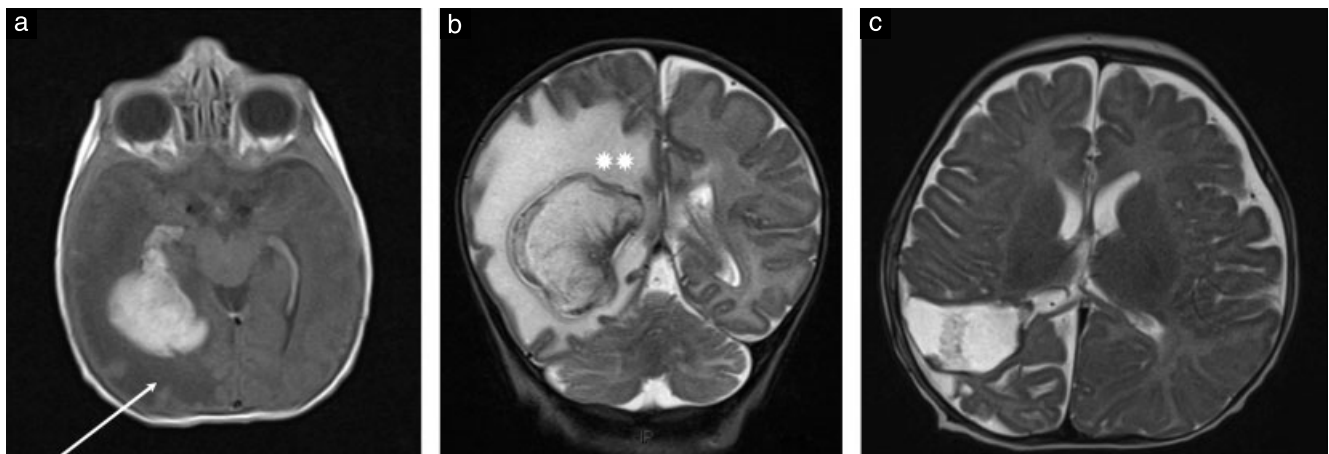


Figure 3 Postnatal magnetic resonance images. (a) Transverse T1-weighted image with gadolinium showing strong contrast enhancement of the tumor (arrow). (b) Coronal T2-weighted image showing peritumoral edema as hyperintense signal (**). (c) Transverse T2-weighted image after surgery showing persistence of porencephalic cavity.

Fetal MRI has been described previously in only one case⁵. MRI appears to be useful to rule out a hematoma and to determine the precise location of the tumor and the presence of peritumoral edema.

We can hypothesize that neurosurgical experience with neonatal brain tumors could be transposed to antenatal cases to help determine the prognosis. According to published series, almost half of these tumors are carcinomas, but the incidence could be different for prenatal cases. In a recent series of 38 cases of plexus tumors operated on at a mean age of 22 months, histology revealed 25 papillomas and 13 carcinomas, and in a more recent study of seven neonates, all cases were papillomas, with one anaplastic papilloma (unpubl. observ.). It appears that during the neonatal or antenatal period, papilloma is more frequent than carcinoma. Radiological findings show no difference between papilloma and carcinoma. Hydrocephalus is observed in both papilloma and carcinoma but rapid growth is usual in papilloma, particularly in neonatal cases.

The prognosis for cases of intraventricular tumor remains difficult to assess based on antenatal imaging findings even with high resolution and specific MRI sequences. The volume of the mass and the findings on color Doppler imaging do not allow discrimination between benign and malignant tumors. Considering that

papilloma and carcinoma are both possible etiologies, and since histology cannot be assessed prenatally, prudent counseling is particularly important in such cases.

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