SHORT COMMUNICATION

GLIOEPENDYMAL AND ARACHNOID CYSTS: UNUSUAL CAUSES OF EARLY VENTRICULOMEGALY IN UTERO

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SUMMARY

In this report we describe two cases of fetal midline intracranial cyst presenting with ventriculomegaly at routine detailed second-trimester scan. In the first case, additional findings included a banana-shaped hypoplastic cerebellum and macrocrania; autopsy after termination of the pregnancy revealed a glioependymal cyst. In the second case, subsequent follow-up examination revealed a progressive increase in cyst size and worsening of ventriculomegaly; termination of pregnancy was performed at 24 weeks and autopsy confirmed an arachnoid cyst. These cases document interhemispheric cyst as a cause for early ventriculomegaly in utero.

KEY WORDS: arachnoid cyst; glioependymal cyst; intracranial cyst; prenatal diagnosis; ultrasonography; ventriculomegaly

INTRODUCTION

The detection of an anechoic structure in the fetal brain, other than choroid plexus cyst, is an uncommon finding at prenatal ultrasonography; differential diagnosis includes arachnoid cyst, porencephalic cyst, aneurysm of the vein of Galen, cystic neoplasm, and Dandy-Walker cyst (Sauerbrei and Cooperberg, 1983; McGahan et al., 1988; Romero et al., 1988; Nyberg and Pretorius, 1990; Langer et al., 1994; Sepulveda et al., 1995). In this report we describe two unusual cases of fetal intracranial cyst, one derived from the arachnoid and the other from glioependymal tissue, both presenting as a midline intracranial cystic mass in association with ventriculomegaly at routine second-trimester anomaly scan.

CASE REPORTS

Case 1

A 26-year-old woman, gravida 2, para 1, was referred for ultrasonographic evaluation at 18 weeks’ gestation because of the detection of fetal ventriculomegaly at routine detailed scan. Ultrasonographic examination at referral showed a singleton fetus in cephalic presentation, a high anterior placenta, and normal amniotic fluid volume. Detailed examination revealed several abnormal findings including a biparietal diameter of 49 mm, a head circumference of 176 mm, a cerebral ventricle to hemisphere ratio of 0·57, and a cerebral ventricular atrium of 12 mm; all these measurements were above the 95th centile for the gestational age (Filly et al., 1991; Snijders and Nicolaides, 1994). An avascular cystic structure measuring 14 mm × 9 mm was clearly identified in the midline of the fetal brain (Fig. 1A). In addition, the cerebellum was banana-shaped with
Fig. 1—(A) Case 1. Ultrasonographic view of the fetal head at 19 weeks' pregnancy shows a gliopendymal cyst (arrow) and ventriculomegaly (calipers). (B) Ultrasonographic view of the posterior fossa shows a banana-shaped hypoplastic cerebellum (between arrows).

Fig. 2—Photomicrograph of the cyst wall shows a ciliated ependymal layer (e) with an underlying glial layer (g) and loose connective tissue.

A transverse cerebellar distance of 13 mm, well below the fifth centile for the gestational age (Snijders and Nicolaides, 1994) (Fig. 1B). However, there was no evidence of spina bifida or other abnormalities. In view of the location of the cyst and associated ventriculomegaly, the diagnosis of an arachnoid cyst was suspected. The parents were counselled of the poor prognosis in view of the associated brain abnormalities and they opted for termination of the pregnancy, which was carried out with vaginal prostaglandin pessaries. Amniocentesis performed before termination revealed a normal 46,XX fetal karyotype. Autopsy revealed a female fetus weighing 420 g, bilateral cerebral ventricular dilatation, a midline cyst lying between the parietal and occipital lobes, and a hypoplastic cerebellum. Histology of the brain revealed that the cyst wall was lined by glial and ependymal tissue (Fig. 2). The neuronal migration in the cerebellum appeared generally abnormal with patchy irregularity in the cortex. The aqueduct of Sylvius was completely absent in one section of the midbrain and represented by a few ependymal rosettes in the other. In the cerebellum the dentate and midline nuclei appeared dysplastic. There were no other abnormalities.
A 30-year-old woman, gravida 2, para 1, was referred at 17 weeks' gestation because of bilateral ventriculomegaly and the suspicion of an enlarged cavum septum pellucidum at routine detailed scan. Ultrasonographic examination at referral demonstrated a singleton fetus with biometry consistent with dates, a high posterior placenta, and normal amniotic fluid volume. Detailed examination of the fetal anatomy demonstrated bilateral ventriculomegaly with atrium measurements of 13 mm but also a midline cystic structure in the anterior half of the brain measuring 8 mm × 8 mm (Fig. 3). The diagnosis of ventriculomegaly secondary to an arachnoid cyst was made. The parents were counselled as to the difficulties in prognosticating the neurodevelopmental outcome of arachnoid cyst in the presence of ventriculomegaly, but a close follow-up to assess the cyst size and progression of ventriculomegaly was felt to be critical to establish the final prognosis. The parents opted for karyotyping by amniocentesis and expectant management with regular ultrasonographic follow-up. Amniocyte culture confirmed a normal 46,XY fetal karyotype. Repeat scans demonstrated progressive increase of the cyst size and worsening ventriculomegaly, which progressed to a severe degree at 24 weeks. At this stage, several management options were discussed with the parents, including ventriculocentesis, serial cyst aspiration, cyst wall resection, preterm delivery and postnatal shunting, and termination of the pregnancy (Stein, 1981; Locatelli et al., 1987; Raffel and McComb, 1988; Jones et al., 1989). They decided on termination of the pregnancy, which was effected by serial administration of prostaglandin pessaries following intracardiac potassium chloride injection to achieve fetal asystole. A male stillborn infant weighing 886 g was delivered vaginally. Autopsy confirmed the presence of a midline cyst with ventriculomegaly and no associated malformations. Histologically, the cyst contained arachnoid tissue.

DISCUSSION

This report describes two cases of midline intracranial cyst causing early ventriculomegaly in utero, one of these being the first description of the prenatal ultrasonographic appearance of cerebral glioependymal cyst. Arachnoid cysts are fluid-filled cavities lined by the arachnoid membrane which can be associated with ventriculomegaly but no other cerebral anomalies (Romero et al., 1988; Nyberg and Pretorius, 1990). On the other hand, glioependymal cysts are derived from displaced neuro-ectodermal tissue and they are usually associated with other brain abnormalities such as agenesis of the corpus callosum, brain heterotopias, and other cerebral dysplasias (Friede and Yasargil, 1977). The gross features of
glioependymal cyst may be indistinguishable from those of arachnoid cyst, although it is possible to differentiate them with light microscopy. Histologically, the wall of the cyst comprises glioependymal tissue in a glioependymal cyst, whereas it contains arachnoid connective tissue in an arachnoid cyst. In doubtful cases, however, electron microscopy is needed for confirmatory diagnosis (Friede, 1985).

Although the frequency of arachnoid cysts and glioependymal cysts differs along the neural axis, glioependymal cysts are far less common than arachnoid cysts. Arachnoid cysts are commonly found in the convexity of the cerebral hemispheres or in the spinal cord, whereas both types of cyst can be found in the supracollicular or retrocerebellar space (Friede, 1985). When the cyst is located in the intracerebellar region, it is almost always glioependymal in origin. Isolated reports have documented ependymal cyst in the spinal canal (Robertson et al., 1991) and Sylvian fissure (Patrick, 1971) but glioependymal cysts in the meninges and cerebral hemisphere are rare.

To our knowledge, only four cases of interhemispheric glioependymal cyst have been reported so far (Barth et al., 1984), none of them prenatally detected by ultrasonography. In our case, the presenting features were a midline anechoic lesion in association with macrocephaly and ventriculomegaly. Although the mechanism of ventriculomegaly is probably quite similar to arachnoid cyst, i.e., compression of the aqueduct of Sylvius by an expanding space-occupying mass, autopsy revealed partial agenesis of the aqueduct of Sylvius, a feature that has not been previously reported in association with glioependymal cyst. Furthermore, it is noteworthy that a banana-shaped cerebellum (Nicolaides et al., 1986) without any spinal abnormality was also noted prenatally; a dysplastic, hypoplastic cerebellum was confirmed at postmortem. To the best of our knowledge, this is the first case documenting a banana-shaped cerebellum in association with an abnormality of the central nervous system other than spina bifida.

Langer et al. (1994) recently reviewed the literature on prenatal diagnosis of arachnoid cysts. All seven cases reported were detected between 22 and 36 weeks' gestation, located in the supratentorial area, and three were associated with ventriculomegaly. Two infants were lost to follow-up, one died at 2 months, two were developing normally at 18 months after surgery, and no information is available from the report on the remaining two. In contrast, our two cases of intracranial cysts were detected before 20 weeks' gestation, at which time the management options include termination of the pregnancy.

To date, there is no available information regarding the prognostic difference between arachnoid cyst and glioependymal cyst, but the association with progressive ventriculomegaly in utero at an early gestational age suggests a poor neurodevelopmental prognosis. Close examination of the interhemispheric area for cystic lesions should be routinely undertaken whenever early ventriculomegaly is detected on prenatal ultrasonography.

REFERENCES


