

Fetal arachnoid cyst: report of two cases

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ABSTRACT

Two new cases of antenatal diagnosis of arachnoid cyst are described. Diagnosis was made at 25 and 32 weeks' gestational age. In the first case, a slight increase in the diameter of the anechoic mass was observed in the second trimester. In the second case, ventricular dilatation was present prenatally. Appropriate surgical therapy was initiated at 2 months in the first case and at 8 days in the second and the neurological development of the two children is considered as normal at 18 months of age. Major features of *in utero* diagnosis, treatment and follow-up are discussed.

INTRODUCTION

Arachnoid cysts represent 1% of all intracranial masses in children¹. Arachnoid cysts are either congenital, resulting from abnormal leptomenigeal formation, or acquired, following *in utero* hemorrhage, infection or trauma. Diagnosis is usually made postnatally during childhood. However, with the development of ultrasound, recognition of arachnoid cysts is now possible prenatally. Only five cases of *in utero* detection of arachnoid cyst have been reported in the literature^{2–5}. However, in these cases, little is known about the circumstances of detection and follow-up of the children. We add two more cases of supratentorial arachnoid cyst depicted prenatally. Major features of *in utero* diagnosis, treatment and follow-up will be discussed.

CASE REPORTS

Case 1

A 20-year-old woman was referred to our University Hospital at 25 weeks' gestational age because of the presence of a cerebral anechoic mass at the second routine scan. On admission, the pregnancy was considered as normal. We performed a prenatal ultrasound examination using a 3.75 MHz transducer. The biparietal diameter, the abdominal diameter and the femoral length

were 6.2 cm, 5.8 cm and 4.3 cm, respectively. Fetal cerebral examination revealed the presence of an anechoic mass (Figure 1), measuring 3.0 × 2.2 cm, located supratentorially within the right parieto-occipital lobe. The anechoic mass had no connection with the lateral ventricle and did not induce a mass effect or ventricular dilatation. The rest of the fetal anatomy appeared to be normal. The amount of amniotic fluid was normal. A further prenatal ultrasound examination at 28 weeks'

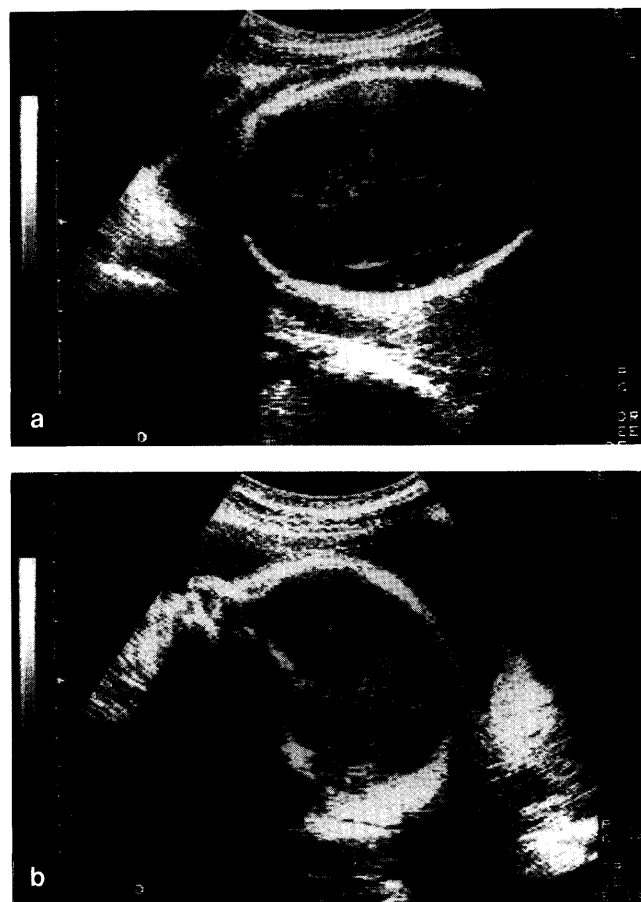


Figure 1 Case 1: prenatal ultrasound at 25 weeks' gestational age. (a) Axial view; (b) sagittal view. Note the cystic anechoic mass located in the right parietal lobe

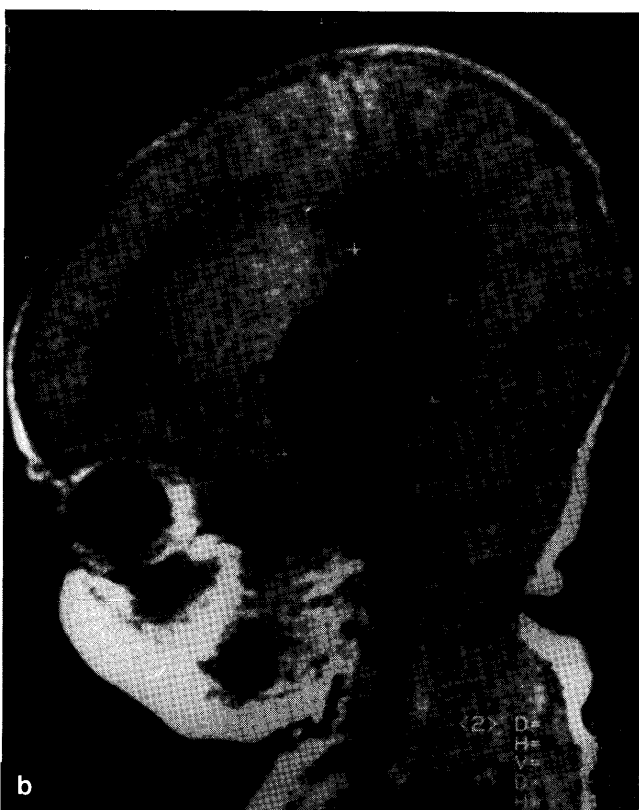


Figure 2 Case 1: postnatal magnetic resonance image 0.5 T GE 450/12 ms/90°. (a) Axial view; (b) sagittal view. Note the cystic mass (hyposignal) located in the parietal lobe and inducing a slight mass effect without evidence of hydrocephalus

gestation showed a slight increase in the diameter of the anechoic mass (4 cm), but without visible mass effect. Two other ultrasound examinations at 32 and 38 weeks showed no modification of the diameter. Based on these observations, the diagnosis of supratentorial arachnoid cyst was suggested. At 40 weeks of gestation, the mother gave birth vaginally to a boy weighing 3620 g. The head circumference was 36.5 cm and the biparietal diameter was 9.8 cm. The Apgar score was 10 at 5 min. No signs of hydrocephalus (bulging fontanelle, increasing head circumference) were noted at birth. Postnatal ultrasound examination and magnetic resonance imaging performed respectively at 1 day old and 5 days old showed the



Figure 3 Case 2: prenatal ultrasound: axial view at 33 weeks' gestational age. Note the anechoic cystic mass located in the mid-line with an important mass effect

presence of an arachnoid cyst located in the middle cranial fossa, right parieto-occipital lobe, inducing a slight mass effect with mid-line shift without evidence of hydrocephalus (Figure 2). The baby was discharged from the neonatal ward at 10 days old. At 2 months old, hydrocephalus developed (increase in head circumference, bulging fontanelle) requiring a cystoperitoneal shunt. At 18 months of age, this child is developing normally. The neuromotor examination was normal, as was the intelligence quotient, estimated by the Brunet-Lezine test.

Case 2

A 21-year-old woman was referred to our University Hospital at 32 weeks' gestation because of the presence of an intracranial anechoic mass at the second routine scan. On admission, the pregnancy was considered as normal and we performed a prenatal ultrasound examination. The biparietal diameter, the abdominal diameter and the femur length were 8.4 cm, 8.2 cm and 6.1 cm, respectively. Ultrasound examination revealed the presence of an anechoic mass of 6 cm diameter located supratentorially (Figure 3) in the mid-line, not communicating with the lateral ventricles. There was an important mass effect, leading to ventricular dilatation. The rest of the fetal anatomy was considered normal. The amount of amniotic fluid was normal. On the subsequent prenatal ultrasound examination at 37 weeks' gestation, no changes in the cerebral anechoic mass or fetal anatomy were noted. The biparietal diameter was within the normal range. Based on these observations, the diagnosis of suprasellar arachnoid cyst was suggested. A Cesarean section was performed at 40 weeks' gestation for contracted pelvis. The Apgar score was 10 at 5 min. The birth weight was 3080 g and the head circumference was 35.5 cm. No clinical signs of hydrocephalus were noted at birth. Postnatal ultrasound examination and magnetic resonance imaging performed at 1 day old and 5 days old, respectively confirmed the suprasellar arachnoid cyst. An important mass effect on the brainstem posterior fossa, optical chiasma and pituitary gland was noted in

Table 1 Arachnoid cysts diagnosed prenatally

Report	Location	Age at diagnosis (weeks)	Associated prenatal signs	Outcome
Diakoumakis <i>et al.</i> ²	suprasellar	32	hydrocephalus; polyhydramnios at 35 weeks	surgery; lost to follow-up
Meizner <i>et al.</i> ⁴	parieto-occipital	22	isolated	no surgery; lost to follow-up
McGahan <i>et al.</i> ³	parieto-occipital	32, 36	—	—
Raman <i>et al.</i> ⁵	posterior fossa	24	hydrocephalus	surgery; died at 2.5 months
Langer <i>et al.</i> (case 1)	parieto-occipital	25	isolated	surgery at 2 months; development normal at 18 months
Langer <i>et al.</i> (case 2)	suprasellar	32	hydrocephalus	surgery at 8 days; development normal at 18 months

Table 2 Number and location of arachnoid cysts

Report	Middle cerebral fossa	Posterior fossa	Convexity	Supra- and infratentorial
Diebler and Dulac ⁶	30	19	8	11
Pascual-Castroviejo <i>et al.</i> ⁷	26	26	5	5
Menezes <i>et al.</i> ⁸	9	13	—	—
Raffel and McComb ⁹	19	7	5	—
Locatelli <i>et al.</i> ¹⁰	13	10	6	—
Total (n)	97	75	24	16
(%)	46	35	11	8

Table 3 Differential diagnosis of supratentorial arachnoid cyst

	Porencephalic cyst	Supratentorial arachnoid cyst	Aneurysm of Galen vein	Cystic neoplasm
Ultrasound appearance and Doppler	anechoic fluid-filled space	anechoic fluid-filled cystic mass	anechoic space venous and/or arterial Doppler signals	heterogeneous mass polyhydramnios
Ventricular communication	+	—	—	±
Subarachnoid space communication	+	+	—	—
Mass effect (mid-line shift)	±	±	±	±
Hydrocephalus	±	±	±	±
Location	brain parenchyma	middle cranial fossa convexity suprasellar	mid-line Galen vein quadrigeminal plate cistern superior and posterior to the thalamus	brain parenchyma

association with a mild ventricular dilatation (Figure 4). The cyst diameter was 6 cm. Because of the importance of the mass effect, cystoperitoneal and ventriculoperitoneal shunts were performed at 8 days old. Following shunt placement, the cyst diameter decreased to 4 cm 8 days later. The infant was discharged from the neonatal ward at 20 days old. She is developing normally at 18 months old. The neuromotor examination was normal as well the intelligence quotient, estimated by the Brunet-Lezine test.

DISCUSSION

To date, relatively few cases of arachnoid cyst have been diagnosed on prenatal sonography²⁻⁵. In the cases reported in the literature (Table 1), prenatal diagnosis was made between 22 and 36 weeks. Three cases were extensively described. In two cases, the arachnoid cyst

was associated with antenatal hydrocephalus and required surgery during the neonatal period. In our two cases, diagnosis was made at 25 and 32 weeks of gestation. In only one case was hydrocephalus present prenatally and associated with mass effect. In five important series reporting location of an arachnoid cyst in children, supratentorial and middle cranial fossa were the most common sites (Table 2). When the arachnoid cyst was detected prenatally, the location was not particularly different (Table 1).

The arachnoid cyst is seen as a thin-walled cystic mass, exhibiting an anechoic signal, located supratentorially, not communicating with lateral ventricles and showing sometimes a mass effect as in case 2. When fetal supratentorial arachnoid cysts are suggested on ultrasound examination, the differential diagnosis includes porencephalic cyst, cystic neoplasms or arteriovenous malformations of the Galen vein (Table 3). However,

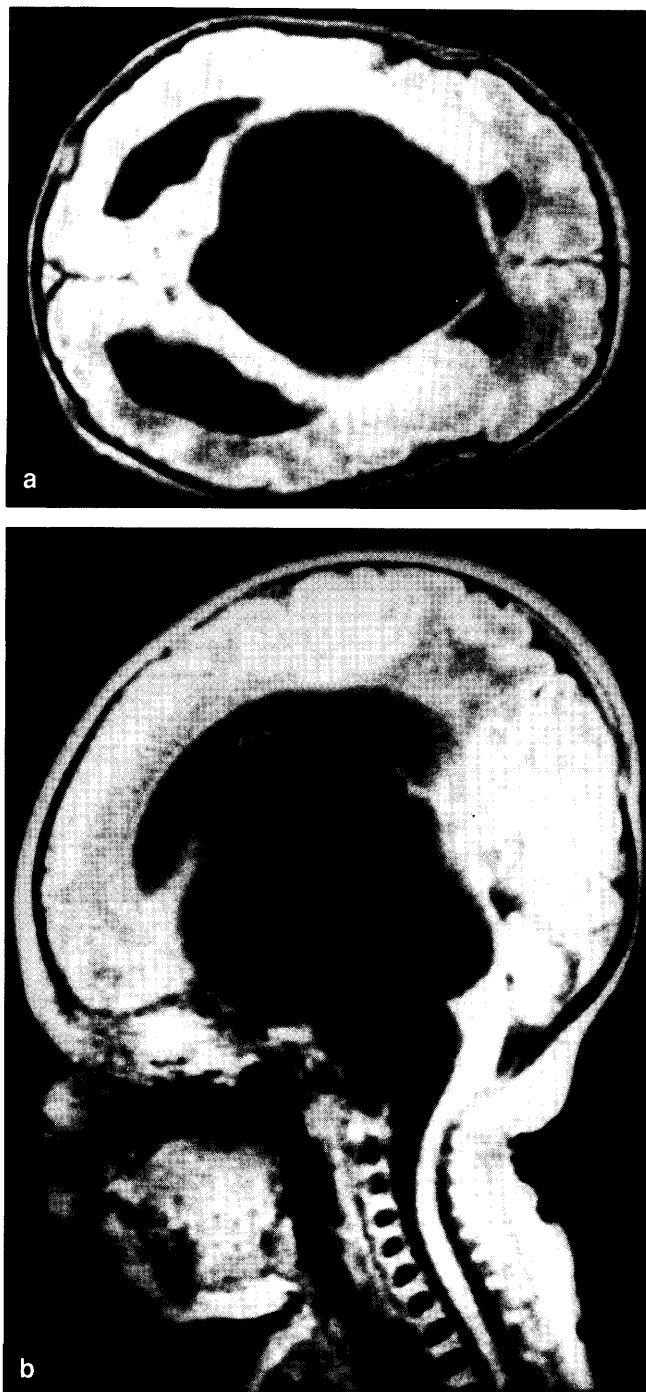


Figure 4 Case 2: postnatal magnetic resonance image 0.5 T GE 450/12 ms/90°. (a) Axial view; (b) sagittal view. Note the large cystic mass (hypointense) with the subsequent mass effect on pituitary gland, brainstem and posterior fossa and the dilated lateral ventricles

differential diagnosis between an arachnoid cyst, often associated with a good outcome, and loss of brain tissue, remains difficult with usually a severe prognosis.

In our cases, prenatal ultrasonography has been used clearly to demonstrate a supratentorial fetal arachnoid cyst. It is noteworthy that prenatal ultrasonography has delineated the cyst with equally good definition as that of postnatal neuroimaging (ultrasound or magnetic resonance imaging).

Furthermore, in our cases, the arachnoid cysts were isolated. No evidence of an abnormal karyotype was

Table 4 Malformations associated with arachnoid cyst

Report	Malformation	n
Menezes <i>et al.</i> ⁸	tetralogy of Fallot	1
Galassi <i>et al.</i> ¹¹	cervical syringomyelia	1
	absent septum pellucidum	2
Jones <i>et al.</i> ¹²	sacroccygeal tumor	1
Pascual-Castroviejo <i>et al.</i> ⁷	agenesis of corpus callosum	8
	deficient cerebellar lobulation	4
	Chiari type 1 malformation	2
	neurofibromatosis type 1	1
	arteriovenous malformation	1

noted in association with the arachnoid cysts. However, many other associated malformations have been reported (Table 4); some of them, especially a major complication like corpus callosum agenesis, should be detected by ultrasound prenatally.

Both children required surgery in the first 2 months of life. Our two patients were scanned at least once in the early period of the second trimester. Apart from the cysts ultrasound examination proved normal at this stage. We observed a slight increase in size of one of the arachnoid cysts during the last trimester. It can be suggested that the arachnoid cyst grows rapidly during the second trimester of pregnancy and increases in size only slightly during the last. The growth potential of a cyst differs from one fetus to another and, in some cases, hydrocephalus and mass effect may occur before birth. Thus, it seems important not only to detect this malformation during pregnancy, but also to follow its growth by repeating ultrasound examinations. This survey is essential to determine the mode of delivery and the early management of the infant. In fact, the prognosis of an isolated arachnoid cyst appears to be excellent, when appropriate surgical therapy is initiated early.

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