Prenatal Diagnosis of a Suprasellar Arachnoid Cyst With 2- and 3-Dimensional Sonography and Fetal Magnetic Resonance Imaging

Difficulties in Management and Review of the Literature

Ali Gedikbasi, MD, Figen Palabiyik, MD, Aylin Oztarhan, MD, Gokhan Yildirim, MD, Ciler Eren, MD, Sezin Saygi Ozyurt, MD, Yavuz Ceylan, MD

Arachnoid cysts are intra-arachnoid sacs containing cerebrospinal fluid–like liquid and represent 1% of all intracranial masses in neonates.1 Although well reported, the precise etiology and pathogenesis of arachnoid cysts are uncertain. Being developmental in origin, arachnoid cysts can be detected on prenatal screening. Although sonography is used routinely for fetal assessment, it is difficult to account for some of the additional anomalies. We report a rare case of a suprasellar arachnoid cyst detected incidentally during a routine fetal sonographic examination at 24 weeks' gestation, with prenatal 2- and 3-dimensional sonography, fetal and neonatal magnetic resonance imaging (MRI), and postnatal follow-up.

Case Report

A 27-year-old gravida was scanned routinely in our fetal diagnosis and therapy unit at 24 weeks' gestation (Voluson 730 Expert; GE Healthcare, Zipf, Austria). The fetus had grown appropriately for gestational age and had normal findings other than the cranium. A round anechoic mass, approximately 25 mm in diameter and without Doppler flow, was detected in the midline suprasellar or third ventricle region of the fetal brain (Figure 1). The lateral ventricular size was slightly increased (11 mm), and no other abnormalities were seen. The family was informed about these findings and given a differential diagnosis that included arachnoid cysts and cystic neoplasms of the sellar or suprasellar region. Porencephalic cysts were excluded because they create a mass effect. In addition, intracranial hemorrhage was also excluded because of its form and anechoic appearance. Infectious marker screening results were negative.

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One week later, a second scan was performed to evaluate progressive findings. The cystic structure and lateral ventricle had enlarged to 28 × 25 mm in diameter and 13 mm in size, respectively. Sagittal views showed displacement of the brain stem posteriorly with obstruction of the ventricular circulation (Figure 2, A and B). The corpus callosum showed normal development with regular blood flow and without findings of increased intracranial pressure (Figure 2C). Three-dimensional sonography showed an isolated round cystic structure (Figure 3). Fetal MRI was performed and confirmed the suprasellar location with the prenatal sonographic findings (Figure 4). An arachnoid cyst was suspected, and a ball valve mechanism with a possible anatomic communication between the cyst and the subarachnoid space was thought to account for the cyst enlargement.

Figure 2. A, Sagittal plane of the fetus after 25 weeks' gestation. B, The cyst and lateral ventricle enlarged in 1 week to 28 × 25 and 13 mm, respectively. C, Sagittal color Doppler image showing appropriate flow of the anterior artery of the corpus callosum.

Pediatric neurologic and neurosurgical counseling were given, and our hospital’s Ethical Committee and Perinatal-Neonatal Council offered the family a discussion of the prenatal findings, neonatal prognosis, and pregnancy management.

Figure 1. A, Sagittal plane of the fetus after 24 weeks' gestation with a midline cyst measuring 25 mm in diameter. B, Transverse plane of the fetus with borderline ventriculomegaly and a midline cyst.
ment and options, including termination of pregnancy. The family decided to continue the pregnancy and accepted karyotyping, which yielded normal results. The latter part of the pregnancy was followed by a 2-week interval with continued growth of the cyst and ventricles and the development of hydrocephaly. The final dimensions of the cyst and lateral ventricles at 36 weeks' gestation were 42 × 37 and 22 mm, respectively.

A 3380-g male neonate with Apgar scores of 7, 9, and 10 at 1, 5, and 10 minutes, respectively, was born abdominally at 37 weeks 3 days because of a breech presentation and a biparietal diameter of 104 mm. The neonate was hospitalized for follow-up and an extended examination in our neonatal intensive care unit. The neonate had stable vital findings and was discharged from the neonatal intensive care unit 2 days later.

Magnetic resonance imaging performed in the first week of life showed the cyst to be displacing the brain stem posteriorly and superiorly and obstructing the third ventricle. Marked dilatation of the third and lateral ventricles was observed (Figure 5). The head circumference at this time was 41 cm (>95th percentile). The neonate's head circumference continued to grow; therefore, a ventriculocystostomy was attempted under general anesthesia without success. Dilatation of the cyst continued, and an infection in the ventriculostomy occurred. The infected drainage was removed. Two weeks later (after antibiotic therapy), a second cystoventriculostomy was attempted with partial success. The continued dilatation of the cyst and lateral ventricles stopped but without resolution (Figure 6). A slight decrease in the size of the third ventricle, arachnoid cyst, and both lateral ventricles was observed. Neurologic development was near normal at 6 months of age. The head circumference remained stable and did not continue to grow. Thyroid-stimulating hormone, growth hormone, luteinizing hormone, follicle-stimulating hormone, and prolactin values were in the normal range. The family was counseled about surgical attempts and medical follow-up; definitive surgery was thus postponed for a later date.

Figure 3. Three-dimensional images of the fetus from the surface into the skull at 25 weeks' gestation. A and B, Surface section and facial image of the fetus. C and D, Deeper 3-dimensional sections of the fetal brain. E and F, Three-dimensional sections showing the arachnoid cyst (arrow) in the fetal brain.
Arachnoid cysts represent approximately 1% of all intracranial space-occupying lesions. Primary (congenital) arachnoid cysts are benign accumulations of clear fluid between the dura and the brain substance throughout the cerebrospinal axis in relation to the arachnoid membrane and do not communicate with the subarachnoid space or ventricular system. Secondary (acquired) arachnoid cysts result from hemorrhage, trauma, and infection and typically communicate with the subarachnoid space. The most frequent locations of arachnoid cysts are the surface of the brain at the level of the main brain fissures, such as the Sylvian, Rolandic, and interhemispheric fissures, the sella turcica, the anterior cranial fossa, and the middle cranial fossa. Because arachnoid cysts are accepted to be developmental in origin, it should be possible to identify them on fetal imaging. Prenatal sonography can show an arachnoid cyst as a hypoechoic lesion as early as 20 weeks’ gestation. However, Bretelle et al reported the first-trimester diagnosis of a posterior fossa arachnoid cyst at 13 weeks’ gestation by transvaginal sonography.

The differential diagnosis for other lesions that appear hypoechoic or space occupying in this region includes gliopendymal cysts, cranio-
pharyngiomas, benign cystic gliomas, Rathke cleft cyts, and colloid cysts of the third ventricle.5–9 Correct diagnosis has very important implications for counseling, especially with respect to continuation of pregnancy and treatment options. Prenatal MRI helps confirm the diagnosis and exclude other possible central nervous system anomalies, especially additional findings of corpus callosum agenesis/abnormalities and cortical gyral abnormalities.7,8 Suprasellar arachnoid cysts may progressively enlarge and cause ventriculomegaly in utero and therefore require serial sonographic examinations.9

There are 3 important points when evaluating an arachnoid cyst and establishing follow-up and the prognostic outcome: (1) determine the presence of other anomalies, especially midline brain developments, such as agenesis of the corpus callosum; (2) assess the size of the ventricular system by measuring the atrium, and search for associated obstructive hydrocephalus, as in our case; and (3) evaluate whether the size of the cyst changes with advancing gestational age.8 In our case, serial sonography allowed us to evaluate the progression of the cyst and ventricles, and prenatal MRI allowed us to identify the accurate location of the cyst and diagnose it early by excluding other sellar lesions. Prenatal diagnosis allowed us to have early discussions with the parents about management designed to minimize damage, including termination of pregnancy.

Fetal arachnoid cysts can be associated with chromosomal abnormalities. Therefore, prenatal diagnosis of an arachnoid cyst, especially in association with structural abnormalities, should prompt a cytogenetic investigation. Various associated chromosomal abnormalities have been reported.11 DiRocco12 reported that among 478 pediatric patients with intracranial arachnoid cysts, one-third had arachnoid cysts on the Sylvian fissure, and only 15% had cysts in the suprasellar region, as in our case. Suprasellar arachnoid cysts are rare, but the anatomic location may result in various subsequent symptoms. If the cyst obstructs the foramen of Monro or displaces the aqueduct, ventriculomegaly may occur, and the head size may increase. Most suprasellar arachnoid cysts are diagnosed after they have caused symptoms in childhood,13,14 and very few cases have been diagnosed prenatally before the onset of manifestations (Table 1).10,15–18

Most patients with suprasellar arachnoid cysts have signs of intracranial hypertension due to hydrocephalus, as in our case. Obstructive hydrocephalus is the most common cause of initial symptoms and occurs in almost 90% of patients with suprasellar arachnoid cysts.19 Other clinical features include compression of adjacent structures, such as the brain stem, thalamus, hypothalamus-pituitary system, and optochiasma by the cyst, and clinical manifestations, such as seizures, endocrine dysfunction, and visual field defects.19,20

Figure 6. T1-weighted axial MRI scans at 5 months of age. A, Note the cystoventriculostomy. B, The low-signal midline cyst in the midbrain is decreased in size but still visible.
Suprasellar arachnoid cysts may cause growth hormone and thyrotropin deficiencies, stimulation of the hypothalamic-pituitary-gonadal axis, a tall stature, and excessive weight gain. Adan et al. reported that of 30 suprasellar arachnoid cysts studied, 60% were associated with endocrine disorders. Additionally, the disorders never regressed postoperatively, even when the decrease in the cyst volume was satisfactory. Mohn et al. reported that most pediatric arachnoid cysts had a growth hormone deficiency and required hormone replacement therapy, even after surgery. There is a high possibility that in our case, the child will also have major complications.

Surgical treatment options remain controversial and most commonly include craniotomy, open fenestration of the cyst, stereotactic cyst aspiration, endoscopic cyst fenestration, and shunt placement. However, it is a difficult procedure, even if the case is younger than 1 year and with such a location.

In conclusion, our case of a suprasellar arachnoid cyst incidentally identified with prenatal sonography was managed carefully in consultation with obstetricians, pediatric neurologists, and neurosurgeons. To our knowledge, 3-dimensional sonographic findings and related images in such a case have not been reported previously. Antenatal MRI was helpful as a supplementary method in the prenatal diagnosis, especially to confirm the suprasellar region. Serial sonographic scans are necessary to monitor the progress of the cyst and additional complications. As of this writing, the patient had near-normal development at 1 year of age with no symptoms, but long-term follow-up was recommended.

References


Table 1. Cases With Prenatal Diagnosis of Suprasellar Arachnoid Cysts

<table>
<thead>
<tr>
<th>Reference</th>
<th>GW at Diagnosis</th>
<th>Sonographic Findings</th>
<th>MRI Findings (GW)</th>
<th>Birth, Findings, Treatment, Perinatal Outcome</th>
</tr>
</thead>
<tbody>
<tr>
<td>Diakoumakis et al.</td>
<td>32</td>
<td>3.5-cm suprasellar cyst developing later hydrocephalus</td>
<td>None</td>
<td>Vaginal delivery at 37 wk, 4.3-cm suprasellar arachnoid cyst with hydrocephalus, solitary seizure at 3 d, cystoperitoneal shunt at 19 d</td>
</tr>
<tr>
<td>Langer et al.</td>
<td>32</td>
<td>6-cm supratentorial arachnoid cyst in the midline with ventricular dilatation</td>
<td>None</td>
<td>Cesarean delivery at 40 wk, suprasellar arachnoid cyst, mild ventricular dilatation, cerebrospinal and ventriculoperitoneal shunts at 8 d, normal development at 20 postnatal d</td>
</tr>
<tr>
<td>Golash et al.</td>
<td>17</td>
<td>2.5-cm midline cystic lesion, increasing to 3.2 cm later in pregnancy</td>
<td>Slight dilatation of the occipital horns of the lateral ventricles (28)</td>
<td>Elective cesarean delivery at 38 wk, ventriculomegaly and macrocephaly, endoscopic cystoventriculostomy and cystocisternostomy with success, normal development at 2 y</td>
</tr>
<tr>
<td>Nakamura et al.</td>
<td>28</td>
<td>Macrocephaly, enlarged ventricles, and midline cystic lesion</td>
<td>Supratentorial and infratentorial fossa cyst compressing the brain stem (28)</td>
<td>Vaginal delivery at 35 wk, implementation of a cyst-peritoneal shunt on day 18, endoscopic cystoventriculostomy at 2 mo with success, a 1-mo developmental delay at 4 mo</td>
</tr>
<tr>
<td>Fujimura et al.</td>
<td>25</td>
<td>3-cm midline cystic lesion, no ventriculomegaly</td>
<td>Confirmation of suprasellar arachnoid cyst (28)</td>
<td>Vaginal delivery at 37 wk, endoscopic cystoventriculostomy at 5 mo, normal development at 3 y</td>
</tr>
<tr>
<td>This case</td>
<td>24</td>
<td>2.5-cm midline cyst with ventriculomegaly, increasing to 5.4 cm with ventriculomegaly and hydrocephaly</td>
<td>Confirmation of suprasellar arachnoid cyst and ventriculomegaly with no additional malformation (26)</td>
<td>Cesarean delivery at 37 wk, ventriculomegaly and macrocephaly, endoscopic cystoventriculostomy with partial success, slight developmental delay at 6 mo</td>
</tr>
</tbody>
</table>

GW indicates gestational week.


