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Case Report

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Prenatal diagnosis of a large suprasellar arachnoid cyst

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Abstract

Arachnoid cysts represent 1% of all intracranial lesions and appear on the arachnoid membrane (subarachnoid space). The etiology and pathogenesis of arachnoid cysts remain unclear and in their majority they are asymptomatic. Due to the developmental origin of arachnoid cysts, these formations can be identified on prenatal fetal imaging. In this report we presented a case of an enlarged suprasellar arachnoid cyst detected incidentally during a routine fetal sonographic examination at 33 weeks of gestation. The cyst remained stable in size prenatally and postnatally without compressive symptoms and signs and the patient has been set under expectant management with serial clinical evaluation and serial imaging of the cyst.

Key words: Arachnoid cyst, Suprasellar, Intracranial abnormalities, Pregnancy, Ultrasound, MRI

Introduction

Arachnoid cysts are rare central nervous system lesions comprising 1% of all intracranial lesions[1,2]. Arachnoid cysts are classified as primary congenital cysts or secondary acquired cysts. Primary cysts arise from the abnormal leptomeningeal formation in utero, resulting in the abnormal accumulation of cerebrospinal fluid (CSF)[3,4]. Secondary cysts are less common, often following trauma, surgery, infection, or intracranial hemorrhage and usually communicate with the subarachnoid space[1].

While the location of arachnoid cysts may vary, most of these lesions are supratentorial and 50-65% occur in the middle cranial fossa. However, arachnoid cysts may also be found in the cerebellopontine angle, suprasellar and quadrigeminal cisterns, cerebral convexities, and cisterna magna[5,6]. Suprasellar arachnoid cysts represent approximately 5 – 12.5% of those lesions[7,8].

Most cases of arachnoid cysts remain unchanged in size for many years, whereas some may expand causing compressive effect on neurovascular structures; much more rarely, arachnoid cysts may disappear spontaneously[9-12].

Diagnosis is usually made postnatally during childhood. However, due to the developmental origin of arachnoid cysts, these lesions can be detected on

prenatal screening. The development of prenatal ultrasound in combination with the appropriate use of magnetic resonance imaging (MRI) have led to the increased prenatal detection of central nervous system malformations of the fetuses, such as arachnoid cysts. The arachnoid cysts are usually an incidental finding.

The prevalence of arachnoid cysts in adults is estimated at approximately 1.4%, while the prevalence in children at 2.6%. Arachnoid cysts are more common in female adults[5,9,13].

In this report, we present a case of an enlarged suprasellar arachnoid cyst detected incidentally during a routine prenatal sonographic examination at 33 weeks of gestation.

Case Report

A 31-year-old primigravida with unremarkable medical, family and habitual history was scanned routinely at 33 weeks of gestation. The ultrasound scan demonstrated a noncommunicating round anechoic formation in the midline suprasellar or third ventricle region of the fetal brain. The lesion was particularly large with a maximum diameter of 41 mm. This finding was absent at the previous ultrasound scan at 21 weeks of pregnancy, suggesting that the lesion was formed after that time. The scan was repeated after 3 weeks and depicted a mild increase in the size of the cystic lesion. In particular, the lesion was measured as approximately 48 mm in diameter. The ultrasound findings were explained to the family and the mother was referred to our University hospital.

On admission, an ultrasound scan was performed confirming the presence of a round anechoic formation measuring 43 x 24 mm in the midline suprasellar or third ventricle region of the fetal brain and without Doppler flow (Figure 1A, 1B). The lateral ventricles were normal in size. No other abnormalities were identified. Intracranial hemorrhage was excluded due to its form and anechoic appearance.

Antenatal fetal Magnetic Resonance Imaging (MRI) was performed at 36 weeks of pregnancy to evaluate other neural structures and rule out agenesis of corpus callosum. Antenatal MRI revealed a cystic lesion at a suprasellar location measuring 46 x 37 x 31 mm without significant compression effect over midbrain and medulla (Figure 2A, 2B, 2C). The development of corpus callosum was normal. The woman underwent

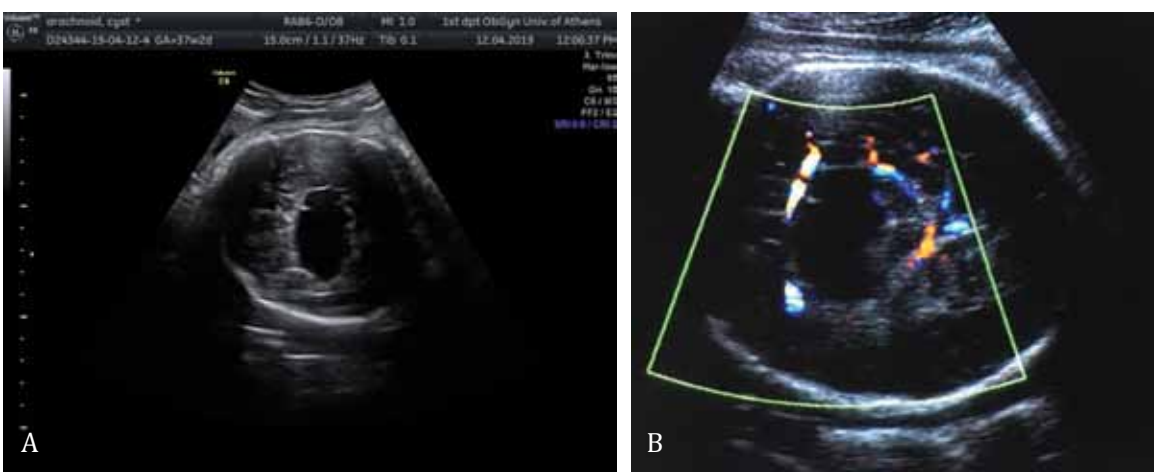

Figure 1. A. Transverse ultrasound plane of the fetus at 36 weeks of gestation with a midline cyst measuring 43 x 24 mm. B. Transverse ultrasound plane of a midline cyst without Doppler flow.

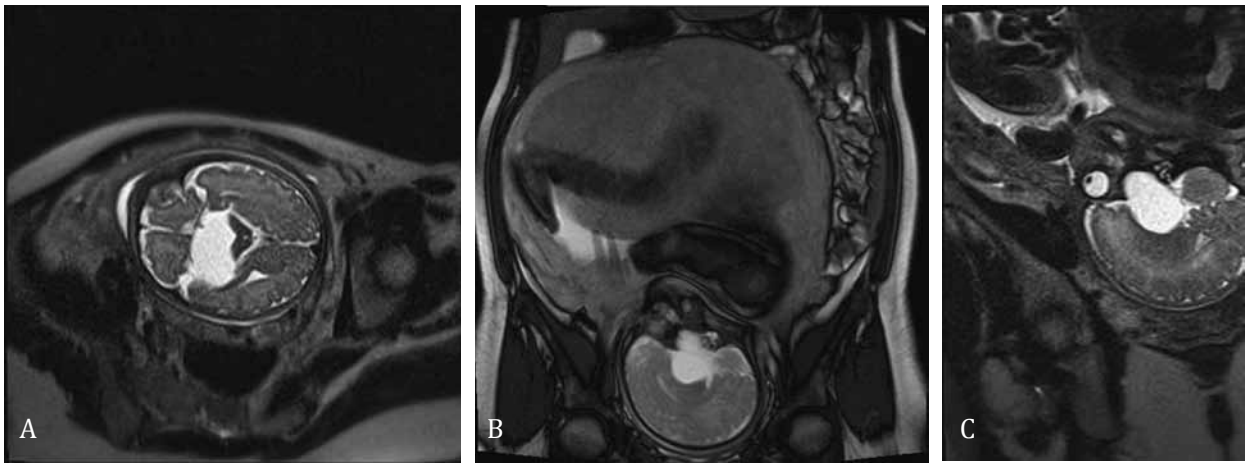

Figure 2. Axial (A) and Sagittal (B,C) fetal MRI demonstrating a midline cystic lesion at 36 weeks of pregnancy.

infectious viral screening, which was found negative. Moreover, the woman underwent karyotyping, which yielded normal results. The family was informed about the possible diagnosis of arachnoid cyst and was given pediatric neurologic and neurosurgical counseling about the neonatal prognosis.

The plan was to follow-up after birth of the neonate.

A 3010gr girl was delivered with elective cesarean section at 38 weeks of pregnancy with Apgar score of 8 points and 10 points at the 1st and 5th minute respectively. The vital signs of the neonate remained stable and the physical examination was found unremarkable. The birth weight, body height, and head circumference were in normal range. The fontanels were flat and soft. No congenital anomaly was noted and neurological examination was normal. The neonate demonstrated good activity and no irritability with normal ocular.

At the day of delivery, the neonate was transferred to a neurosurgical department for a specialized evaluation and treatment. The clinical neurosurgical evaluation and the following postnatal electroencephalogram of the neonate were unremarkable. A postnatal MRI was performed on the third day of life confirming the presence of the suprasellar arachnoid cyst measuring 53 x 38 x 32 mm with a mild compression effect on brain stem. The ventricular system was normal in size showing no dilatation. A fundoscopic exam was performed for the evaluation of cranial nerves showing no abnormal findings. Furthermore, the neonate underwent a cranial ultrasound on the sixth day of life reconfirming the existence of the arachnoid cyst measuring 2,39 x 4,41 x 2,85 cm with a mild midline shift. Doppler assessment did not reveal blood flow within the cyst. The ventricular system remained normal in size.

Since the neonate was free of signs and symptoms of raised intra-cranial pressure, any urgent intervention was deferred. The neonate was discharged from neurosurgical care unit on the eighth day in stable condition. The patient was decided to be followed up with serial clinical evaluation and imaging assessment (cranial ultrasound and MRI scan) without any intervention.

At 7 months of age, this child remains in good condition with normal neuromotor development, while the surveillance imaging, including 2 MRIs at 2,5 and 5 months of life respectively, has not demonstrated any increase of the size of the cyst.

Discussion

Arachnoid cysts are rare space-occupying malformations of central nervous system representing 1% of all intracranial lesions[1,2]. Primary arachnoid cysts are congenital benign anomalies arising from the abnormal embryogenesis of the arachnoid leading to irregular splitting or duplication of arachnoid membrane[14]. As a result, accumulations of cerebrospinal fluid (CSF) are formed between the dura and the neural substance along the cerebrospinal axis in relation to the arachnoid membrane. These malformations consist of layers of arachnoid cells in combination with collagen fibers. Primary cysts do not communicate with the subarachnoid space or ventricular system[15-17]. Arachnoid cysts may also be secondary acquired anomalies following trauma, surgery, infection, or intracranial hemorrhage and usually communicate with the subarachnoid space[1].

The formation of arachnoid cysts can occur at various locations along the arachnoid membrane. They are usually recognized as a single-chamber formation in the midline or asymmetrically. These formations are usually characterized by irregular outlines. Most of arachnoid cysts are supratentorial and 50-65% occur in the middle cranial fossa. These lesions may also be found in the cerebellopontine angle, suprasellar and quadrigeminal cisterns, cerebral convexities, and cisterna magna[5,6]. Suprasellar arachnoid cysts account for approximately 5 – 12.5% of those lesions[7,8].

The size of the cyst may remain unchanged in utero, as in the case presented in this report, or expand gradually with the developing fetus, leading to secondary ventriculomegaly.

The absence of long-term follow-up data of untreated patients leads to incomplete knowledge of the natural history of arachnoid cysts. Most cases of arachnoid cysts remain unchanged in size for years, whereas some may increase gradually causing compressive effect on neurovascular structures; much more rarely, arachnoid cysts may disappear spontaneously[9-12].

Arachnoid cysts are mainly diagnosed in childhood after the onset of manifestations[18]. The frequency of these lesions has increased due to the widespread use of routine scanning for a variety of reasons. Al-Holou et al demonstrated that the incidence of arachnoid cysts was estimated at 2,6% in 11738 children under 18 years of age who underwent brain MR imaging[9].

While this lesion can present as a mass lesion, the vast majority of cases is generally asymptomatic and the cysts can be found incidentally during imaging that is usually performed for other reasons[5]. However, the enlargement of these lesions may occasionally present with compression symptoms. In particular, the compressive effect of these masses on adjacent structures may lead to neurological signs and symptoms, such as headaches, dizziness, nausea, vomiting, mental status changes, ataxia, seizures, and hearing loss[19].

The anatomic location of suprasellar arachnoid cysts may lead to various subsequent symptoms. Therefore, obstruction of the foramen of Monro or displacement of the aqueduct by the cyst could lead to ventriculomegaly. Most cases with suprasellar cysts are characterized by signs of intracranial hypertension following the development of obstructive hydrocephalus, which occurs in almost 90% of patients with suprasellar cysts[20]. The location of suprasellar cysts may also be associated with compressive effect on structures, such as thalamus, hypothalamus-pituitary system and optochiasma leading to related clinical symptoms, such as seizures, endocrine dysfuction and visual field defects[20-22]. Endocrine manifestations are related with hormone deficiencies, such as thyrotropin and growth hormone deficiencies, stimulation of the hypothalamic-pituitary-gonadal axis and tall stature[22]. Moreover, endocrine dysfuction does not

seem to regress, even after surgery and decrease of the cyst size[23].

Due to the developmental origin of arachnoid cysts, these formations can be identified on prenatal fetal imaging. Arachnoid cysts are depicted as hypoechoic lesions and may be detected as early as 20 weeks of gestation. However, Bretelle et al reported the recognition of a posterior fossa arachnoid cyst at 13 weeks of gestation[24]. Larger studies have demonstrated that the prevalence of these cysts on imaging is estimated at approximately 2%[5]. Other lesions with the same hypoechoic appearance include glioependymal cysts, craniopharyngiomas, benign cystic gliomas, Rathke cleft cysts and colloid cysts of the third ventricle and they should be considered as possible differential diagnoses[1,7,25-29].

Arachnoid cysts have been associated with several malformations, such as agenesis of corpus callosum, absent septum pellucidum, deficient cerebellar lobulation, cervical syringomyelia, sacrococcygeal tumor, tetralogy of Fallot, arteriovenous malformation, Chiari type 1 malformation, and neurofibromatosis type 1[15,30-32].

Prenatal MRI is necessary for the diagnosis and the evaluation of other neural structures such as corpus callosum. Arachnoid cysts are depicted as no-contrast-enhanced lesions following cerebrospinal fluid intensity in all MRI sequences[33]. High diagnostic accuracy is vital for the appropriate counseling, therapeutic management and follow-up plan.

The prognostic outcome of an arachnoid cyst depends on several parameters. The co-existence of other central nervous system anomalies, such as agenesis of the corpus callosum, the increasing size of the cyst with advancing gestational age and the presence of associated obstructive hydrocephalus are parameters determining the prognosis and the follow-up of an arachnoid cyst[7]. Concerning our case, serial sonographic examinations allowed us to assess changes in the size of the cyst and ventricles, whereas prenatal MRI allowed us to recognize the exact location of the lesion excluding other co-existing anomalies.

Arachnoid cysts can occur as isolated defects or be associated with chromosomal abnormalities[3]. Therefore, prenatal diagnosis of an arachnoid cyst should be followed by cytogenetic investigation. In the present report, no evidence of abnormal karyotype was identified in association with the arachnoid cyst.

The management of arachnoid cysts depends on the patient's symptoms and radiological findings. Surgical treatment should be considered for patients presenting a mass effect of the cyst or raised intracranial pressure. However, arachnoid cysts remaining stable in size without compressive symptoms and signs could be set under expectant management with serial clinical evaluation of the patient and serial imaging of the cyst, as in the present case. There are reports of spontaneous resolution of arachnoid cysts both in children and adults[18].

Symptomatic cysts should be treated[34]. Arachnoid cysts blocking the normal pathway of cerebrospinal fluid and leading to symptomatic hydrocephalus should be treated surgically. Other complications of arachnoid cysts include hemorrhage within the cyst or into the subdural chamber or even rupture of the cyst. Surgical treatment includes craniotomy, open cyst fenestration, stereotactic cyst aspiration, endoscopic cyst fenestration and shunt placement[20,21,35]. The significant morbidity of open fenestration and the increased incidence of failures and infections in cystoperitoneal shunting obliged neurosurgeons to seek for alternative surgical options. Endoscopic fenestration seems to be a better surgical option as it does not require the invasiveness of open craniotomy and avoids the complications of shunting[36]. Endoscopic fenestration can be a treatment option for suprasellar cysts because of their location in the anterior portion of the third ventricle.

Conclusion

This case of suprasellar arachnoid cyst incidentally detected with prenatal sonography was managed in consultation with obstetricians and pediatric neurosurgeons. Prenatal MRI was performed as a supplementary prenatal method in order to confirm the suprasellar location of the cyst and evaluate any compression effect on adjacent neural structures. Antenatal serial sonographic scans and MRI are necessary to monitor the size of the cyst and additional complications. As of this writing, the patient had normal development without symptoms; nevertheless, long-term follow-up was recommended.

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