

CASE REPORT

Prenatal diagnosis of pyriform sinus fistula: case report and literature review

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Introduction

Pyriform sinus fistula is a congenital anomaly that originates from the third or fourth branchial pouch remnants. The characteristic findings of pyriform sinus fistula are the location on the left side and adherence to the thyroid gland and thyroid components associated with the cyst [1]. As the anomaly is situated near the respiratory tract, it is potentially dangerous for neonates because of airway compression, which may lead to respiratory distress. Prenatal diagnosis of abnormalities which causes airway compression is important because it has a high impact on fetal adaptation to extrauterine life. However, there have been only two reports of a pyriform sinus fistula that was diagnosed prenatally, cases in which the fetuses presented with a cystic mass in the left neck [2, 3]. In the present case report, we describe a case in which a pharyngeal cyst without external neck mass was detected and diagnosed prenatally as a pyriform sinus fistula on ultrasonography and magnetic resonance imaging (MRI). We also reviewed all cases of prenatal diagnosis of pyriform sinus fistula in the English language literature.

Case Report

A 38-year-old woman, gravida 1, was referred to our hospital for further evaluation of a pharyngeal cyst of the

Key Clinical Message

We describe a case of prenatal diagnosis of a pharyngeal cyst as a pyriform sinus fistula on the findings of ultrasonography and magnetic resonance imaging.

Keywords

Ex utero intrapartum treatment, magnetic resonance imaging, prenatal diagnosis, pyriform sinus fistula, ultrasonography.

fetus at 35 weeks gestation. Prenatal ultrasonography showed that the fetus had a $3 \times 2 \times 2$ cm hypoechoic simple cystic mass in the pharyngeal region, without signs of internal blood flow by Doppler examination (Fig. 1A and B). Amniotic fluid volume was normal. Fetal MRI at 35 weeks gestation indicated the presence of a $3 \times 3 \times 2$ cm simple cystic mass on the left side of the posterior pharyngeal gap, a mass that showed marked extrinsic compression of the airway (Fig. 2A). A prenatal diagnosis of pyriform sinus fistula was made on the basis of these findings. There was no family history of congenital malformation.

The prevention of asphyxia at birth was of such importance that the mode of delivery was discussed extensively, with combined input from obstetrics, neonatology, anesthesiology, and pediatric surgery. The ex utero intrapartum treatment (EXIT) procedure was adopted to manage airway access at birth. The cesarean section was performed at 37 weeks of gestation under deep maternal and fetal anesthesia. The fetus was delivered via the EXIT procedure, and the endotracheal tube was placed to establish the airway in the first attempt. Direct laryngoscopy showed a mass that demonstrated extrinsic compression of the airway at the level of the larynx. At that time, an opening of the pyriform sinus fistula was not observed. A male neonate weighting 2684 g, with Apgar scores of 5

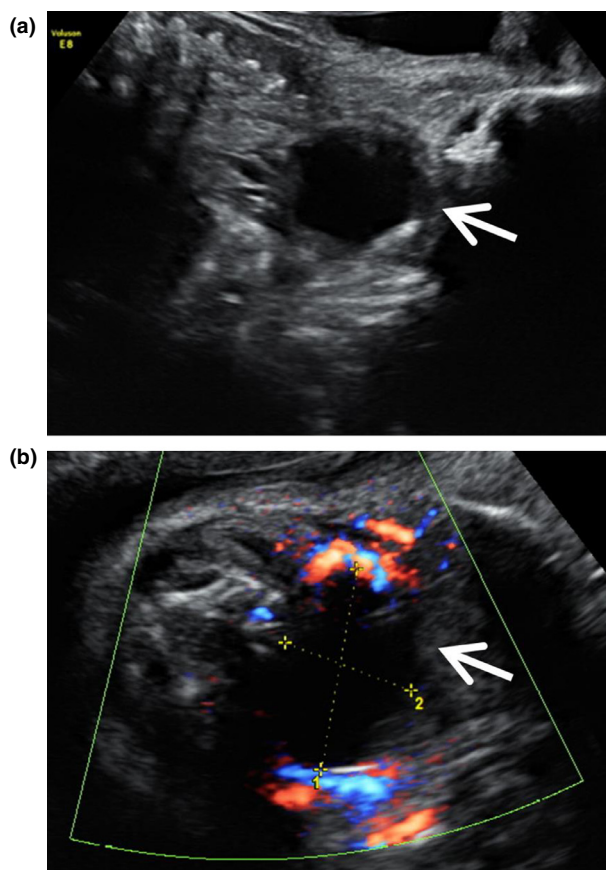


Figure 1. Prenatal ultrasonography showed that the fetus had a $3 \times 2 \times 2$ cm hypoechoic simple cystic mass in the pharyngeal region (A), without signs of internal blood flow by Doppler examination (B).

and 6 at 1 and 5 min, respectively, was delivered with no apparent neck mass or visible cutaneous sinus.

The neonate was admitted immediately to the neonatal intensive care unit. On admission, ultrasonography showed a well-circumscribed hypoechoic mass lying adjacent to the left lobe of the thyroid. Six hours after birth, MRI indicated the presence of a $4 \times 3 \times 1$ cm simple cystic mass in the left side of the posterior pharyngeal gap. This cyst compressed the airway mostly at the level of the oropharynx, but did not show complete airway obstruction (Fig. 2B). Although the endotracheal tube was removed the next day, the neonate showed signs of respiratory distress, which was managed by continuous positive airway pressure (CPAP).

On the ninth day after birth, computed tomographic scan showed an expanded cyst filled with air, indicating communication with the pharyngeal cavity, which marked extrinsic compression of the upper airway. The infant was taken to the operating room. The diagnosis was confirmed by preoperative laryngoscopy and catheterization of the fistula. A left cervical transverse incision was

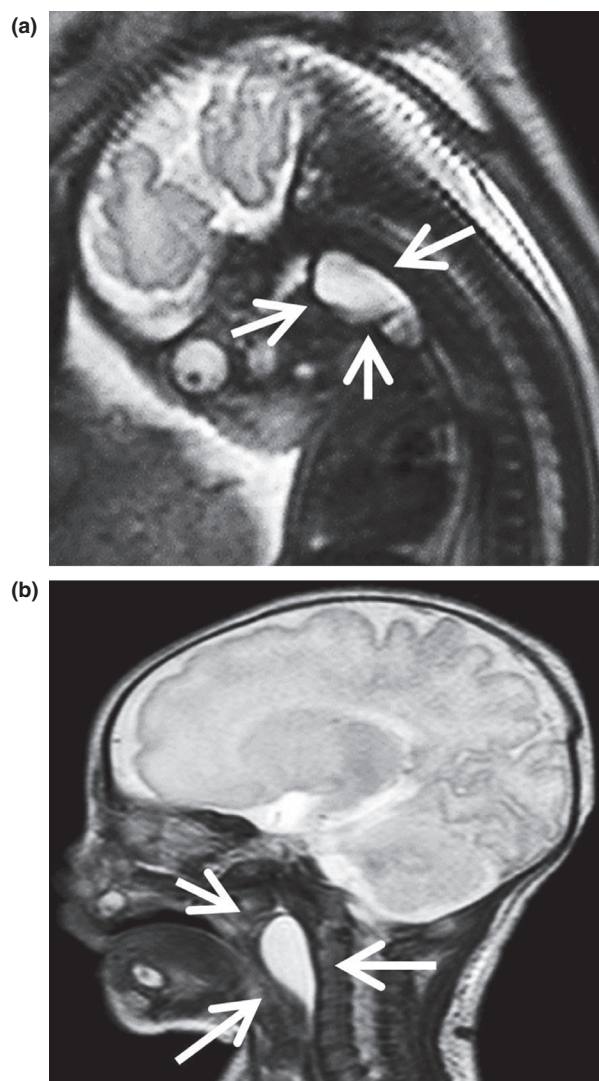


Figure 2. (A) Prenatal magnetic resonance imaging showed a simple cystic mass (arrows) in the posterior pharyngeal gap, a mass that showed marked extrinsic compression of the airway. (B) Magnetic resonance imaging showed a simple cystic mass in the left side of the posterior pharyngeal gap, a mass that showed compression of the airway mostly at the level of the oropharynx.

performed, and the pyriform sinus cyst and entire fistulous tract were excised. Histopathological examination showed that the cyst was lined with ciliated epithelium with adherent thyroid tissue. The postoperative course was uneventful. The infant was discharged 10 days after the operation having completely recovered.

Discussion

Pyriform sinus fistula is a congenital anomaly, which reported to be accounting for 3–10% of pharyngeal

lesions [4]. The clinical manifestations of a pyriform sinus fistula depend on its location and size, as well as whether it is infected. In neonates, common symptoms of pyriform sinus fistula are a left neck mass and respiratory distress. However, cervical pain and recurrent suppurative thyroiditis, which is a common presentation in late childhood, are extremely rare in the neonatal period [5].

In this article, we report a case of prenatal diagnosis of a pharyngeal cyst as a pyriform sinus fistula on ultrasonography and MRI, which did not present as an external neck mass as commonly observed in the cases of neonatal pyriform sinus fistula. Differential diagnosis of a pharyngeal cyst includes a second branchial cleft cyst, pyriform sinus fistula, or vascular or lymphatic malformation [6]. In the present case, the cyst was unilocular and located on the left side of the posterior pharyngeal gap and was compressing the airway at the level of the larynx. Therefore, we diagnosed it as a pyriform sinus fistula prenatally, and the diagnosis was confirmed by

preoperative laryngoscopy and catheterization of the fistula.

MRI appears to be better than ultrasonography in the prenatal evaluation of airway obstruction, which is most likely due to the image contrast of the MRI [7]. In the present case, the fetal neck was found to be flexed on MRI, closely associating the jaw and chest wall. Therefore, the extent of airway compression by the pharyngeal cyst was predicted to be more severe than what was found upon evaluation by MRI after birth. The present case indicated that the prenatal evaluation of airway obstruction on MRI can be affected by the extent of flexion of the neck and may cause difficulty in accurately predicting airway obstruction.

To the best of our knowledge, there have been only two reports on the diagnosis of prenatal pyriform sinus fistula [2, 3]. The previous case reports and the current case are summarized in Table 1. The two previous reports were made based on a prenatal diagnosis due to the find-

Table 1. Summary of case reports of prenatal diagnosis of pyriform sinus fistula.

Authors [ref]	Year	Gestational age at diagnosis	Finding of ultrasonography at diagnosis	Finding of magnetic resonance imaging at diagnosis	Delivery	Clinical presentation after birth	Treatment
Chin <i>et al.</i> [2]	2000	29	A 14 × 14 × 11 mm cystic lesion in the anterior triangle of the left side of the neck	NA	Spontaneous vaginal delivery at full term	No evidence of a neck mass or respiratory embarrassment at birth Respiratory distress and a 3 × 3 cm fluctuant mass in the anterior triangle on the left side of the neck a week after birth ¹	Total excision via open neck surgery
Yanai <i>et al.</i> [3]	2004	23	A 7 × 8 × 6 cm hypoechoic cystic mass in the left neck	A large cystic mass in the left neck	Vaginal delivery at 39 weeks gestation	A 7 × 8 cm mass under the sternocleidomastoid muscle on the left side of the neck at birth Respiratory difficulty that required endotracheal intubation 6 hours after birth	Total excision via open neck surgery
Present case	2013	35	A 3 × 2 × 2 cm simple cystic mass in the pharyngeal lesion	A simple cystic mass 3 × 3 × 2 cm on the left side of posterior pharyngeal gap	Cesarean delivery and ex utero intrapartum treatment procedure at 37 weeks gestation	No evidence of a neck mass or skin change Endotracheal intubation via ex utero intrapartum treatment procedure, and respiratory difficulty, which was managed by continuous positive airway pressure after extubation on day 1 after birth until the surgery	Total excision via open neck surgery

NA, not applicable.

¹Data of the timing of surgery were not available.

ing of a cystic mass of the left side of the neck. This report is believed to be the first case report of prenatal diagnosis of a pharyngeal cyst as a pyriform sinus fistula. Nathan et al. reported a case of a neonate with stridor due to a pyriform sinus fistula, which was diagnosed on postnatal MRI, without external neck mass [8]. Therefore, the present case indicated the importance of noticing that pyriform sinus fistulas can be prenatally identified by the presence of a pharyngeal cyst.

The neonates in the previous two reports were born by vaginal deliveries. As endotracheal intubation was required to establish the airway in the previously reported case with a large cystic mass [3], respiratory distress may worsen in the early neonatal period. The EXIT procedure can be used to obtain a fetal airway while uteroplacental gas exchange is preserved. Allen et al. reported a case of pregnancy complicated by a fetal oropharyngeal cyst and successfully securing the fetal airway using the EXIT procedure [9]. In the present case, the EXIT procedure was applied to manage airway access at birth, and endotracheal intubation via the EXIT procedure was useful until the airway was evaluated and secured. Prenatal diagnosis allows us to utilize the EXIT procedure or resuscitation of the newborn. We consider it is important to determine the indications for the EXIT procedure based on multidisciplinary discussion.

The previous case reports and the current case were excised the entire pyriform sinus fistulous via open neck surgery. The aim of the treatment is the complete excision of the pyriform sinus cyst and fistula [10]. Nicoucar et al. reported on 45 neonatal cases of pyriform sinus fistula [11]. In this report, 13 (31%) of the 45 neonates received incision and drainage and 34 (76%) open neck surgery. Incision and drainage showed a high failure rate. Amano et al. reported that the cyst generally expanded as the baby swallowed saliva or milk, and its respiratory distress worsened in the early neonatal period [5]. Furthermore, an infected pyriform sinus complicates surgery because identification and dissection of pyriform sinus fistula is quite difficult [12]. Therefore, early diagnosis can help surgeons in planning the surgical intervention to treat a pyriform sinus fistula.

In conclusion, a pyriform sinus fistula in the prenatal period is rare but should be considered in the differential diagnosis of pharyngeal cyst or lateral cervical cyst. Prenatal diagnosis can help facilitate perinatal management by a multidisciplinary approach.

Conflict of Interest

None declared.

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