Spontaneous Rupture of Prenatally Detected Thoracic Neurenteric Cyst: Case Report and Review of Literature

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Abstract

Neurenteric cysts are usually associated with vertebral anomalies of the same segments of vertebrae. Rarely, they can be detected antenatally. The prognosis depends upon the size and site of the lesion. In cases of small cysts with minimal mass effect, the prognosis is good. Larger cysts, however, can cause hydrops fetalis and, if untreated, can lead to intrauterine fetal demise. The present case report describes the successful conservative management of the neurenteric cyst detected prenatally and is accompanied by a summary from literature.

Keywords

- neurenteric cyst
- prenatal diagnosis
- spontaneous rupture
- vertebral defect
- conservative management

Introduction

Neurenteric cyst (NEC) is an uncommon (incidence approximately 0.7–1.3% among spinal cord lesions) developmental anomaly where the ectoderm of the notochord and primitive foregut fails to separate during the 3rd to 6th week of embryonic development, also resulting in associated vertebral anomalies like hemivertebrae. It is largely believed to occur due to vascular anomalies leading to inadequate nutrient supply to the neural folds. Based upon its embryonic origin, NEC presents in intraspinal or extraspinal locations (cervical or thoracic region, mainly in the posterior mediastinum [PM] adjacent to the vertebrae).[1–17]

Case Report

A 29 year old primigravida mother, nonconsanguineous marriage, was diagnosed with isolated fetal cervicothoracic hemivertebrae by prenatal ultrasonography (US) at 21 weeks of gestation. Limb movements were normal. The couple was counseled regarding the good prognosis of isolated hemivertebrae. At 25 weeks, a small anechoic, avascular, spherical mass ($17 \times 12 \times 8$ mm) was seen in the left PM (►**Figs. 1** and **2**) without any mediastinal shift. Fetal magnetic resonance imaging (MRI) (►**Fig. 3**) suggested that the dorsal hemivertebrae at the D1-D6 level with a posterior mediastinal mass suggestive of an NEC. Monthly follow up scans were done to look for cyst size and adjacent visceral compression until 36 weeks (►**Table 1**). The cyst gradually increased to $39 \times 21 \times 20$ mm with a mediastinal shift. There was no hydrops. At 36 weeks, the cyst had reduced in size and had irregular, thick hypoechoic margins. Autocorrection of the mediastinal shift with left unilateral minimal pleural effusion occurred, likely due to rupture of the cyst (►**Table 1**). At 36 weeks, labor was induced for maternal preeclampsia and a 2.42 kg female baby was delivered (►**Figs. 4** and **5**).

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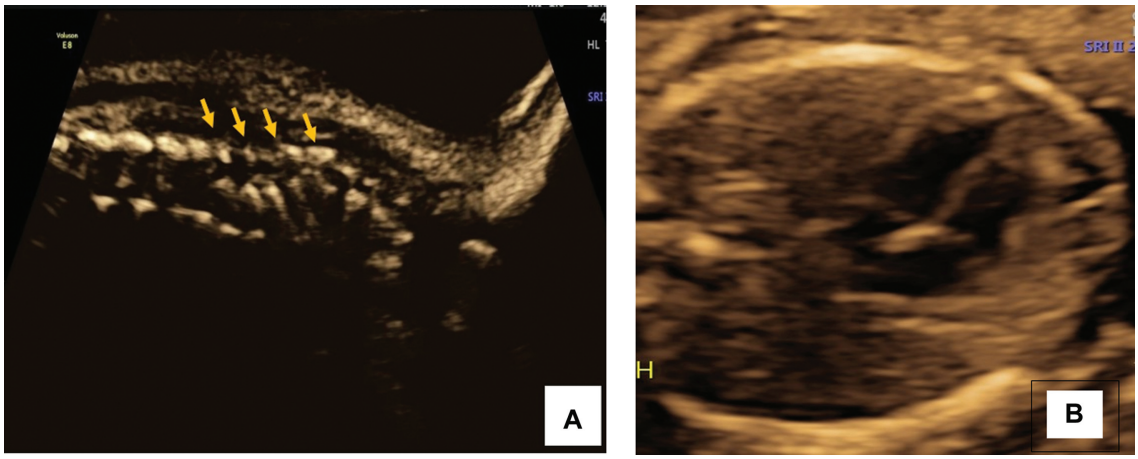

Fig. 1 (A) Cervicothoracic spine with hemivertebrae (marked with yellow arrows). (B) Normal four-chamber view of the heart at Targeted Imaging for Fetal Anomalies (TIFFA).

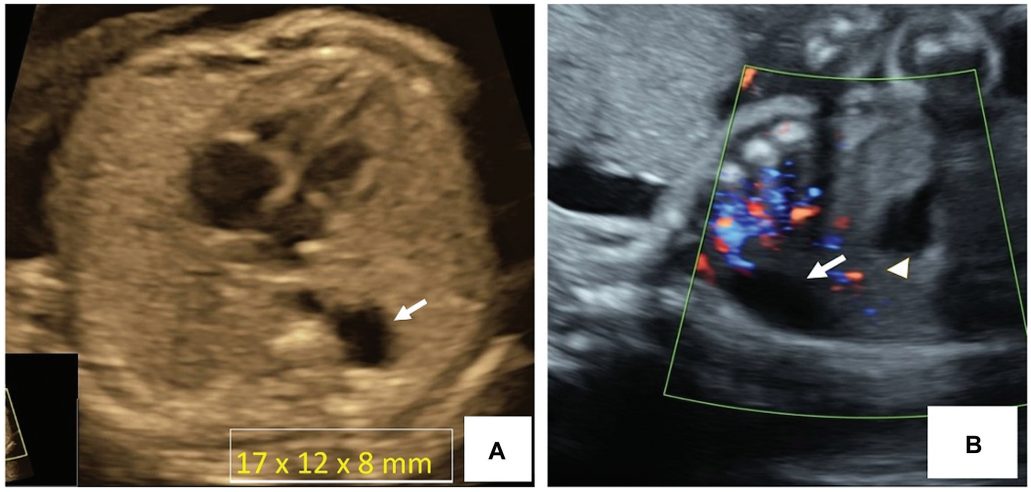

Fig. 2 (A) Small anechoic avascular cyst (marked with white arrow) measuring 17 × 12 × 8mm in the posterior mediastinum posterior to the descending aorta in the axial section of the fetal thorax. (B) Sagittal section of the fetal thorax showing avascular cyst. White arrowhead*: stomach.

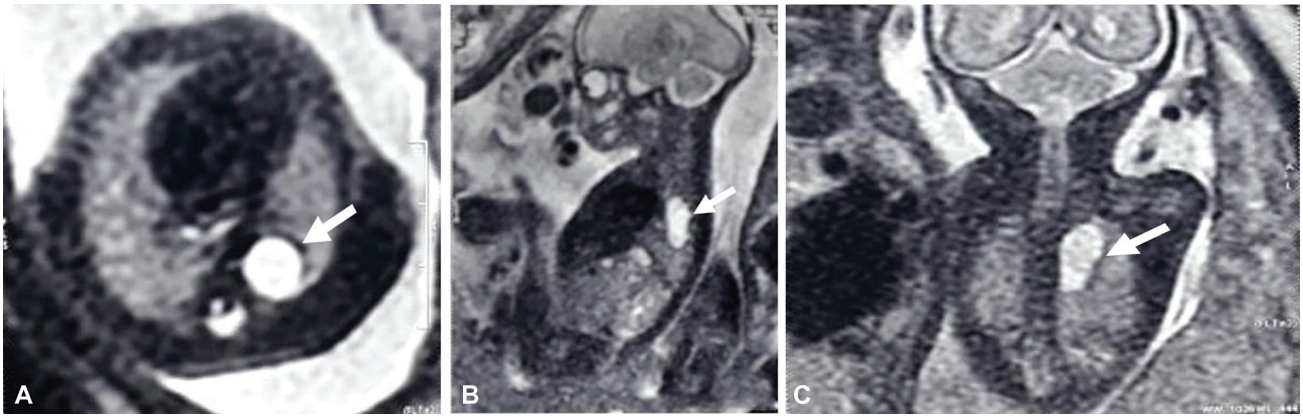

Fig. 3 Fetal magnetic resonance imaging (MRI). (A) A small anechoic cyst (white arrow) in the posterior mediastinum in the axial section of the fetal thorax measuring 22 ×13 × 12 mm. (B) Sagittal section of the fetal thorax showing avascular cyst. (C) Coronal section of the fetal thorax shows the proximity of the cyst to the hemivertebral segment.

Table 1 Cyst measurements and ultrasound findings through the course of gestation in the current case

Gestational age	USG finding	Cyst measurement	Location	Corresponding figure no.
25 wk	Small anechoic, avascular, spherical structure	17 × 12 × 8 mm	Left PM, posterior to descending aorta and just adjacent to the spinal defect, without any mediastinal shift	►**Fig. 2**
29 wk	Without any mediastinal shift, compression effect or hydrops	31 × 12 × 10 mm		
32 wk	Mild mediastinal shift but without any signs of hydrops	34 × 14 × 13 mm		►**Fig. 4**
35 wk		39 × 31 × 20 mm		
36 wk	Autocorrection of the mediastinal shift with left-sided unilateral minimal pleural effusion with irregular, thick hypoechoic margins, likely suggestive of antenatal rupture of the cyst	25 × 11 × 11 mm		►**Fig. 5**
Postnatal USG and chest X-ray	Collapsed cyst	25 × 11 × 11 mm		►**Fig. 6**

Abbreviations: PM, posterior mediastinum; USG, ultrasonography.

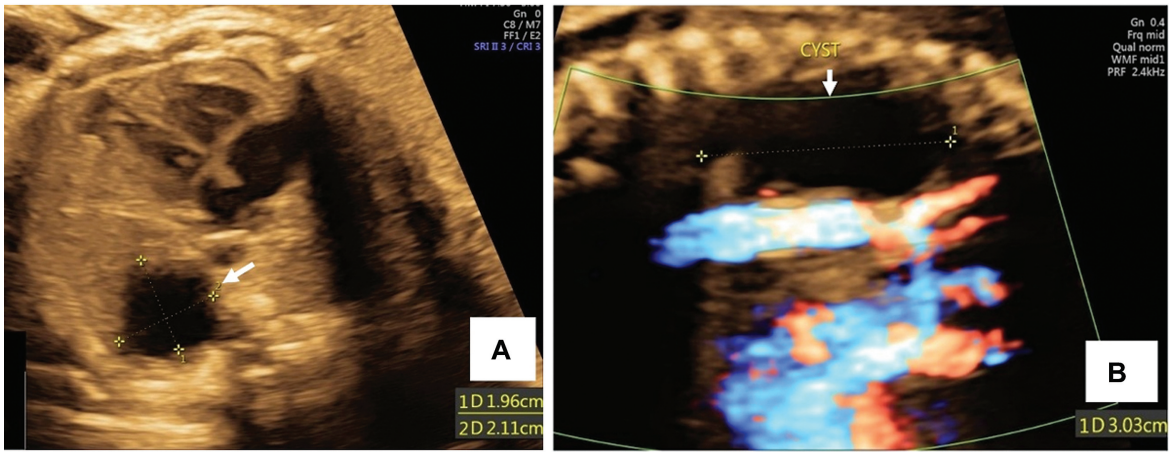

Fig. 4 (**A**) Axial section of the fetal thorax showing enlargement of the size (white arrow) with midline shift (pseudodextrocardia). (**B**) Sagittal section of the thorax demonstrating avascularity of the cyst.

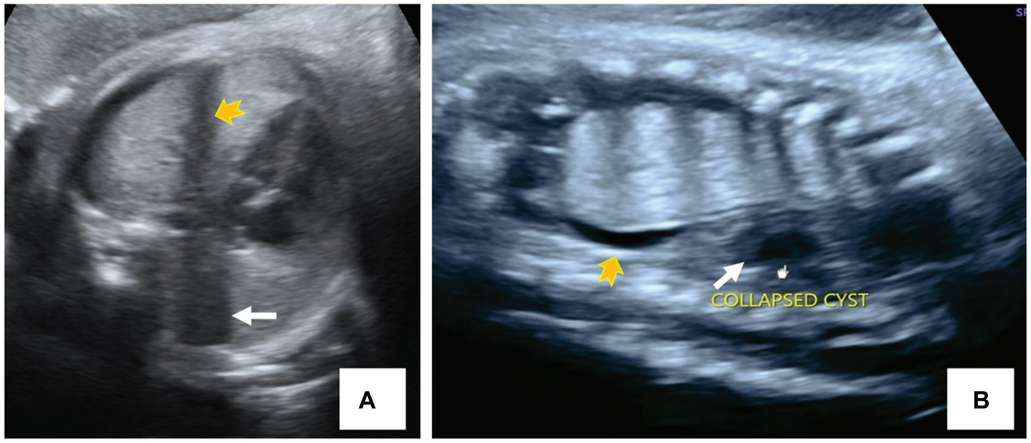

Fig. 5 (**A**) Axial section and (**B**) sagittal section showing collapsed cyst (white arrow) with pleural effusion (yellow notched arrow).

Table 2 Review of literature: summary of cases reported by various authors

Authors	GA	Size of the cyst	Location	Vertebral anomalies	Hydrops	Intervention prenatally	Complications	Delivery	Surgery	Communication	Postnatal complications	Outcome
Wilkinson et al[7] (1999)	28 wk	5 cm	RT PM	UT HMV	Yes	FTC (twice)	Recurrence	At 31 wk 2 d (preterm labor) 1.4 kg baby	D3- Percutaneous aspiration of the cyst D4- RPLT CRC	Spinal canal with 2 cm stalk	RDS difficulty in extubation	Good
Macaulay et al[23] (1997)	23 wk	6.1 × 2.7 × 4.6 cm	RT PM	Cleft vertebrae cervical	Yes	FTC + TAS	–	38 wk 5 d	D21- Emergency CRC	No	None	Good
Gadodia et al[8] (2010)	28 wk	4 × 5 cm	PM and anterior to SC	Yes	No	None	No	37 wk	D15- CRC (both cysts) by RPLT	Spinal canal	None	Good
Çay et al[9] (2018)	31 wk	2.7 × 3.2 × 2.7 cm	RT PM	HMV, butterfly vertebrae	No	None	Increase in size + mild pleural effusion	37 wk	D7 CRC	Closely adherent to spinal column	None	Good
Perera and Milne[24] (1997)	34 wk	4.6 × 2.4 cm	RT HT PM	HMV T3-T4	No	None	Increase in size	Term	D2 CRC	No	None	Good
Daher et al[16] (1995)	32 wk	2 cm	RT HT PM	No defect (fibrous connection)	No	None	No	35 wk 3 kg male	D2 RPLT CRC	To first thoracic vertebra with fibrous tract	RDS	Good
Rizalar et al[25] (1995)	32 w	4 × 5 × 5 cm	RT HT PM	HMV, anterior spina bifida	No	None	No	38 wk	D1 RPLT CRC	Esophagus, RT diaphragm	RDS	Good
Gulrajani et al[10] (1993)	38 wk	3 × 2 cm	Anterior to the vertebral defect	Anterior kyphosis C7-T1	No	None	No	Term	8 mo, RPLT CRC	Spinal canal	Increase in size after 3 months Postoperative CSF pleural effusion requiring multiple thoracocentesis	Good
Bejjani et al[11] (2022)	Prenatally	3.5 × 2 × 2 cm (PM) 1.4 × 0.7 × 1 cm (spinal)	PM and anterior to SC	HMV, ventral cleft C7-T1	No	None	No	Term	5 mo RPLT CRC of mediastinal cyst followed by partial resection of intraspinal cyst 3 weeks later	Spinal cord	Meningitis	Good
Fernandes et al[26] (1991)	22 wk	Large	RT HT PM	Anterior vertebral body defect	Not reported	None	Not reported	34 wk, 2.2 kg	D7 RPLT CRC	Jejunum and spinal canal at T1 level	No	Good
Uludağ et al[22] (2001)	34 wk	3.14 × 4.39 cm	RT HT PM	HMV, cervicothoracic	No	None	No	36 wk, 2.6 kg male	D4 RPLT and CRC	No	No	Good
Bernasconi et al[12] (2007)	38 wk	4 cm in HT and 2 cm in spinal canal	RT HT PM	Mid-thoracic vertebral defect	No	None	No	Term	3 wk RPLT and CRC	Spinal canal	No	Good till 3 years
Kimya et al (2007)[13]	21 wk	2.5 × 1.1 cm	RT HT PM	Split vertebra T5-T7	No	None	No	Termination	NA	Epidural space	NA	Termination
Setty et al[27] (2005)	Not available	Not available	RT HT PM	HMV scoliosis	No	None	No	Term	1 mo- RPLT and CRC	No	RDS postnatally for 2 weeks	Good
Worrel et al[14] (2021)	22 wk	Suspected CDH with stomach in thorax	LT CDH with stomach and pancreas in thorax	Anterior fusion defect C2-C6 HMV C7	No	None	Fetal distress	31 wk 3 d	NA	Stomach with anterior cervical vertebra	Postnatal RDS	NND on D1
Present	25 wk	17 × 12 × 8 mm	LT HT PM	HMV T1-T6	No	No	Rupture of the cyst with pleural effusion	36 wk	2 and half month	No	None	Good

Abbreviations: CDH, congenital diaphragmatic hernia; CRC, complete resection of cyst; CSF, cerebrospinal fluid; FTC, USG-guided percutaneous fetal thoracentesis; GA, gestational age; HMV, hemivertebrae; HT, hemithorax; LT, left; NA, not available; NND, neonatal death; PM, posterior mediastinum; RDS, respiratory distress syndrome; RPLT, right posterolateral thoracotomy; RT, right; SC, spinal cord; TAS, thoracoamniotic shunt; UT, upper thoracic.

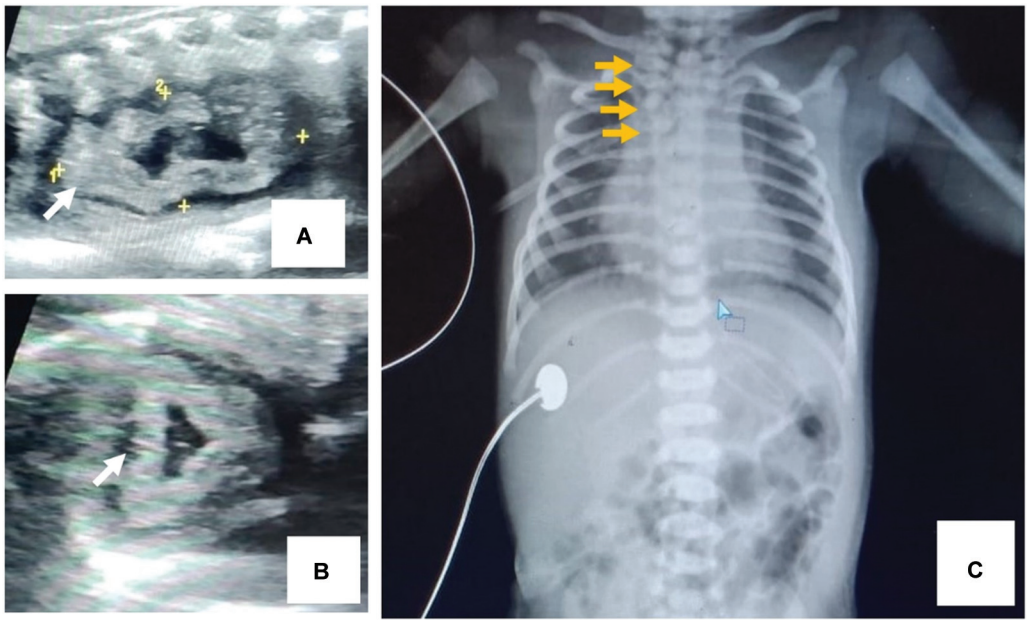

Fig. 6 (A) Neonatal chest ultrasound. (A) Sagittal section and (B) axial section showing collapsed cyst (white arrow) measuring 25.3 × 11.3 mm. (C) Neonatal infantogram showing clear lung spaces and hemivertebrae in thoracic segment (yellow arrows).

Postnatal examination confirmed the antenatal findings with spontaneously resolved pleural effusion (►**Fig. 6**). The child had been asymptomatic since birth. At 2 months of age, she underwent elective resection of the cyst through a right posterolateral thoracotomy and no active communication was found with the spinal canal. The postoperative period was uneventful. Histopathology of the specimen showed cyst wall fragments with ulcerated lining, submucosa, and muscularis mucosa resembling bowel. She has attained normal milestones since.

Discussion

Among thoracic NEC, most case reports (►**Table 2**) have documented cysts in the right PM, but in the current case, it was in the left PM. Similar to the current case, these usually gets detected beyond the late second trimester of pregnancy. Combined use of US and fetal MRI to better assess intraspinal or intracranial smaller cysts, spinal defects, and communication with the spinal column can help improve diagnosis.[18–27]

Rare genetic associations between NECs and Klippel-Feil syndrome and VACTERL association are documented, often with multiple fetal defects. However, genetic testing is not warranted for isolated cases due to low recurrence risk and limited genetic association.[28–31]

Serial ultrasound monitoring of the cyst size and compression effect is essential. If enlarged and complicated by hydrops, fetal procedures like thoracentesis, thoracoamniotic shunting, or in utero thoracic mass excision are available. However, these are associated with complications like reaccumulation of fluid, infection, and increased morbidity (►**Table 2**). The current case had minimal compression effect without hydrops. A conservative approach was, therefore, preferred.[7]

Expectant management is challenging due to the late presentation, evolving complications and associated anomalies. Timely delivery and multidisciplinary care that involves an obstetrician, fetal medicine specialist, neonatologist, pediatric orthopedic surgeon and pediatric surgeon are essential. Long term follow up is required for respiratory complications until postnatal resection and beyond. Even after a successful complete resection, postoperative complications like infections and cerebrospinal fluid leak or pleural effusion can occur. In partially resected cases, recurrence or enlargement of the cyst may be anticipated and timely treated.[7–12,16,22–27,32]

Conclusion

Serial ultrasound in NEC cases guides treatment based on size and location. Anticipating recurrence and proactive surgical excision, despite cyst collapse, highlights the need for vigilant management to ensure optimal outcomes.

Conflict of Interest
None declared.

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