

Posterior mediastinal gastroenteric cyst in neonate

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ABSTRACT

Posterior mediastinal enteric cysts are infrequently reported. They are mostly asymptomatic. 1. The incidence of gastroenteric cysts presenting during immediate neonatal period is rare. Alimentary tract duplications are other rare congenital anomalies and are commonly seen in relation to the ileum. However, the high incidence of associated thoracic or cervical vertebral anomalies with foregut cysts provide an early clue to the diagnosis. 2. A detailed timely antenatal scan can increase the awareness regarding such rare condition and help in diagnosis and better outcome. The purpose of this case report is to highlight the clinical diagnosis and management of a neonate with posterior mediastinal gastroenteric cyst.

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Mediastinal cysts in neonates and young children are infrequently reported. Enteric cysts in the posterior mediastinum are mostly asymptomatic.¹ the incidence of gastroenteric cysts presenting during immediate neonatal period is rare. However the high incidence of associated thoracic or cervical vertebral anomalies with foregut cysts provide an early clue to the diagnosis.²

Case Report. A full term male newborn infant admitted in our neonatal intensive care unit (NICU) at maternity and children's hospital, Madina, Kingdom of Saudi Arabia, with history of respiratory distress and cyanosis developed soon after birth. Apgar scores were 8 and 10 at 1 and 5 minutes. Mother was 29-year-old, and she had regular antenatal check ups. Antenatal ultrasound scan at 35-weeks gestation reported a cystic mass in right hemithorax. The birth weight was 3.45 kgs, length and head circumference were above 50th percentile for gestational age. Pulse rate 158 beats per minute and blood pressure 78/43 mm Hg.

Patient had no dysmorphic features and had been pink at birth, but subsequently he developed respiratory distress and cyanosis. Chest examination revealed impaired breath sounds on right side. Peripheral pulses were equally palpable and synchronous with normal heart sounds. Abdominal examination revealed no organomegaly, and nasogastric tube could easily pass to the stomach. Patient had normal external male genitalia. Patient passed urine and normal meconium stools within next 12 hours of age. Initial arterial blood gas showed pH 7.22, partial pressure of carbon dioxide (PCO₂) 47.2 mm Hg, partial pressure of oxygen (PO₂) 49.9 mm Hg, HCO₃ 19 mmol/L, and oxygen saturation 76.5%. Patient was intubated and ventilated because of respiratory distress and hypoxemia, subsequently he became pink and blood gases were satisfactory. Hematological profile and serum chemistry results were all within normal range. Chest x-ray revealed rounded opacity in right lung field with mediastinal shift to the left, and thoracolumbar spine x-ray did not reveal any

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vertebral abnormalities. Chest ultrasound scans reported thick walled cystic mass in the posterior region of right hemithorax and suspected pleural effusion with collapsed right lung. However, CT (computerized tomography) scan of chest reported encysted pleural effusion, an oval cystic lesion in the paradorsal area of lower lobe of right lung. Three possible entities were included in our differential diagnosis; namely, bronchogenic cyst, cystic adenomatoid malformation and neurenteric cyst (**Figures 1 & 2**). During the hospital course patient was initially resuscitated and clinically stabilized and on the 6th day of life, he was subjected to surgical intervention. Right lateral thoracotomy incision revealed the following observations. the chest wall and pleura were thick and edematous, and thick inflamed necrotic mass measuring 3 x 7 cm extending posteriorly towards vertebral bodies in the mid chest but free from esophagus. The mass ruptured during surgical dissection and foul smelling dark fluid was discharged. Finally, the mass was completely removed and chest wall was sutured with the chest drain in situ.

Histopathological examination of the resected mass revealed features of mucosal ulceration, and microscopically the wall of the cyst lined by gastric mucosa, glands and underlying longitudinal, circular and oblique muscle layers, which was consistent histopathologically with gastric mucosa (**Figure 3**). Post operative period was uneventful and patient was extubated on 6th post operative day and discharged in good general condition at 16th day of life to be followed up in the out patient clinics.

Discussion. Mediastinal cysts in neonates and young children are infrequently reported. Enteric cysts in the posterior mediastinum are mostly asymptomatic. If the cysts are communicating with the lumen of respiratory tract, then they may present with respiratory distress. These communicating cysts may get infected, and present with features of sepsis and respiratory complications.³ Posterior mediastinal cysts have been reported in 7% of all posterior mediastinal masses in 4 series of 572 patients.¹ Enteric cyst or enterogenous or esophageal cysts have been rarely reported. Enteric cysts (or enterogenous cysts) are congenital anomalies of the developing foregut and include esophageal duplication cysts and neurenteric cysts.⁴ A neurenteric cyst is a term used to describe an enteric cyst associated with vertebral or neural anomalies secondary to failure of the foregut to lose its connection to the notochord. A fibrous tract attaching the cyst to the spine may be seen. In our patient, although a thick fibrous cord like structure was extending posteriorly but the vertebral bodies and esophagus were free. Neurenteric cysts



Figure 1 - Plain chest x-ray showing opacity of the right hemithorax.

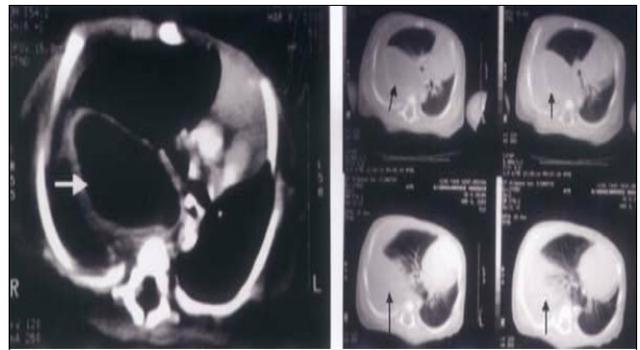


Figure 2 - Chest computerized tomography scan revealing right posterolateral cystic shadow.

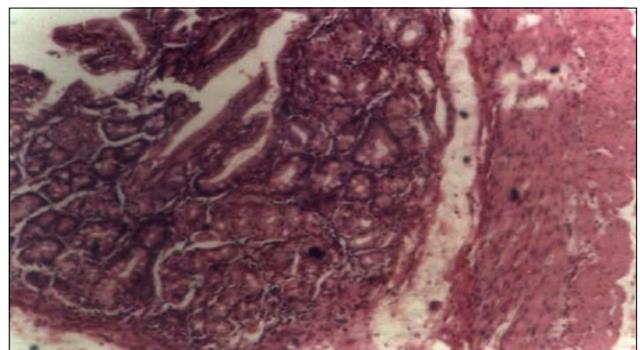


Figure 3 - Histopathology appearance of the mass.

frequently contain both neural and enteric tissue, including gastric mucosa. Approximately 90% of neurenteric cysts occur in the posterior mediastinum, usually superior to the carina, on the right, and separated from the esophagus. Associated vertebral abnormalities are found in 50% of cases such as scoliosis, anterior spina bifida, hemivertebrae, and butterfly vertebrae.⁵ The wall of an esophageal duplication cyst contains alimentary epithelium and 50-60% of them contain gastric mucosa, or pancreatic tissue.⁵ Mediastinal enteric cyst is a distinct entity from esophageal duplications, but it is usually included within esophageal duplication cysts because of its location. They are usually found in the right posterior mediastinum in retrocardiac position and often extending into the right hemithorax. Clinical presentation may include respiratory distress due to mass effect, as it was true in our patient who developed distress soon after birth. Mediastinal enteric cyst's lumens are partially or completely lined by gastric or intestinal mucosa surrounded by smooth muscle. They arise from the failure of coalescence of vacuoles early in the development of the foregut.

To the best of our knowledge, there have been very few reports of enterogenic cysts in posterior mediastinum in neonates and young children, and possible this is the first case report of posterior mediastinal gastroenteric cyst presented in early neonatal period in the Kingdom of Saudi Arabia.^{4,6-8} Preoperatively it is difficult to distinguish the type of cyst. Chest x-ray usually revealed sharply defined cysts with rounded soft tissue masses typically located in the middle or posterior mediastinum. The cyst wall is typically thicker than that associated with bronchogenic cyst. Radioisotopic study with Technetium 99 scan may be useful in the diagnosis of enteric cysts. Computerized tomography scan is a useful imaging modality in defining the exact anatomic location of the cyst. Surgical excision of the cyst is the mainstay of treatment along with comprehensive supportive care.⁹ Prenatal sonographic detection of a posterior mediastinal cyst is feasible¹⁰ and suspected cases of lung mass should preferably be delivered in hospitals where

neonatal intensive care facilities are available and the newborn infant can be further investigated appropriately with combined neonatal and pediatric surgical team approach that will help in timely intervention which hopefully will smooth out the ultimate outcome.¹¹

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