CASE REPORT

Perforated neuroenteric cyst masquerading as congenital pulmonary airway malformation

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SUMMARY

A 3-month-old child was presented with haemoptysis with respiratory distress. Imaging was suggestive of a cavitary lesion in the lung with surrounding consolidation. Diagnosis of a primary lung pathology like congenital pulmonary airway malformation was considered. Based on clinical suspicion and prior experience, a Tc-99m pertechnetate radionuclide study was performed, which clinched the diagnosis of foregut duplication cyst. Intraoperative findings confirmed the presence of a neuroenteric cyst. The child remains asymptomatic on follow-up awaiting neurosurgical intervention for the intraspinal component of the cyst.

BACKGROUND

Foregut duplication cysts are rare entities, but the diagnosis is often evident on imaging, which reveals a mediastinal cyst with gut signature on walls. This is often associated with bony abnormalities of the spine. Rarely this cyst becomes complicated due to rupture or haemorrhage, which may lead to secondary changes in the lung. In such cases the cyst may mimic congenital pulmonary airway malformation (CPAM) on radiological investigation. This report describes successful management in a case where differentiation between these two entities was evident on imaging.

CASE PRESENTATION

A 3-month-old girl was presented to paediatric emergency with complaints of respiratory distress for the past 1.5 months. The child had been hospitalised twice elsewhere and had been managed conservatively with intravenous antibiotics. The child also had an episode of blood expectoration 3 days back and was subsequently referred to our institute. On examination, the child was febrile with tachycardia and tachypnoea. On chest auscultation, the air entry was decreased in the left upper region with presence of crepitations.

INVESTIGATIONS

Chest radiograph and contrast-enhanced CT of the chest both revealed a cavitary lesion in the left upper lobe with surrounding consolidation (figure 1). A diagnosis of CPAM with pneumonia was made, and the child was admitted for stabilisation and intravenous antibiotics under paediatrics unit, and surgical consultation was sought.

In view of the history of expectoration of blood, thick-walled cyst and close proximity to posterior

mediastinum, the diagnosis of duplication cyst was still suspected. A Tc-99m pertechnetate radionuclide scan was performed to look for ectopic gastric mucosa. The scan showed avid uptake in the upper thoracic cyst suggestive of a foregut duplication cyst (figure 2).

TREATMENT

The patient underwent left posterolateral thoracotomy, which revealed a 3 cm by 3 cm intact cystic lesion in the left paravertebral area at the level of T2–T3 vertebrae, just lateral to the aorta. There were adhesions to the surrounding structures. Narrow communication of sac with the vertebrae was present at the same level. The mucosa of the cyst was excised flush with the vertebrae. The tract was closed with interrupted sutures. On histopathology, the specimen was lined by gastric mucosa with underlying two-layered smooth muscle bundle compatible with neuroenteric cyst type B.

OUTCOME AND FOLLOW-UP

Postoperative period was uneventful. The child was advised an MRI of the cervicodorsal spine, which revealed an intraspinal cyst at the level of the sixth and seventh cervical vertebrae (figure 3). The child was advised neurosurgery consultation at her native place as per the parent's request.

DISCUSSION

Alimentary tract duplications have a reported incidence of 1 in 4500 by autopsy series¹; still, it is a condition rarely encountered in clinical practice. Thoracic enteric duplication cyst accounts for 20% of all duplication cysts.² Neuroenteric cyst is a thoracic enteric duplication that extends to the spine and is associated with thoracic or cervical vertebral abnormalities. Therefore, the diagnosis can be suspected on a plain chest radiograph, which reveals a posterior mediastinal cyst associated with vertebral defects, which can be further buttressed by performing CT or MRI. These cysts have characteristic enteric lining that may contain ectopic gastric mucosa in up to half of the cases.³ Tc-99m pertechnetate radionuclide imaging is a useful modality in the preoperative localisation of ectopic gastric mucosa, with a sensitivity approaching 81%.⁴ Ectopic gastric mucosa is physiologically active and releases acid which may erode the wall of the cyst, resulting in haemorrhage or rupture of the cyst.

The index case is unique in two ways. The child was presented with expectoration of blood,

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Figure 1 (A) Chest radiograph frontal view showing a cavity (white arrow) in the left upper zone with surrounding consolidation (black arrow). (B) Contrast-enhanced CT scan of the chest coronal plane showing a thick-walled cavity (black arrow) in the left upper lobe with surrounding consolidation and another thick-walled fluid-filled cavity in the paravertebral region (white arrow).

which was associated with cough along with a cystic lesion in the thorax. These features are suggestive of a primary lung pathology. However, due to suspected bony abnormality, posterior mediastinal location of the lesion and presence of bleeding made us to suspect a diagnosis of neuroenteric cyst. A similar case seen in our department about a decade ago in which the diagnosis was made postoperatively made us wiser.⁵ A Tc-99m pertechnetate scan was ordered to look for ectopic gastric

mucosa, which clinched the diagnosis of enteric cyst, clarifying the radiological misdiagnosis. The rupture of the cyst resulted in secondary consolidation or cavitation in the adjoining lung, which appeared as primary lung pathology on imaging studies.

Multiple reports have documented duplication cyst or neuroenteric cyst as the cause of haemoptysis.^{5–7} In spite of significant haemoptysis in these cases and documentation of blood in the bronchus on bronchoscopy, none of these patients demonstrated a communication with the bronchus or the

Figure 2 Tc-99m pertechnetate scan revealed tracer accumulation in the thoracic region, suggesting ectopic functioning gastric mucosa in the thoracic duplication cyst (black arrow).

Figure 3 T2-weighted MRI of thoracic spine in sagittal plane showing a well-defined intraspinal cystic lesion (arrow) anterior to the spinal cord at the level of C6–C7 and associated fusion of C6 to T1 vertebral bodies with non-visualisation of intervertebral disc space.

oesophagus. In the index case also, other than adhesions with the surrounding structures, no air leak or oesophageal communication was noticed after excision of the cyst. Probably the fistulisation between the airway and the cyst is at a microscopic level and is not visible to the naked eye.

Once the diagnosis is established, the treatment is straightforward and complete removal of the cyst is needed. If the cyst wall is adherent to the adjoining vital structures, then complete removal of the cyst mucosa will suffice.

Learning points

- ▶ Neuroenteric cyst is a rare condition and usually presents with symptoms related to mass effect.
- ▶ Rarely the cyst may involve the lung and result in haemoptysis and cavitation in the lung, which may look like a primary lung pathology.
- ▶ 99Tc pertechnetate scan can clinch the diagnosis in these doubtful cases.

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