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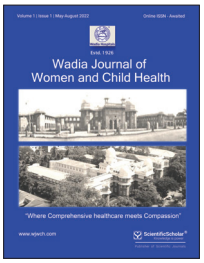

Case Report

An unusual case of extrapulmonary sequestration presenting as vomiting in a neonate

Leena Patwardhan[1], Mausmi Khawale[1], Dhruv Mamtora[1]

[1]Department of Laboratory, Bai Jerbai Wadia Hospital for Children, Mumbai, Maharashtra, India

***Corresponding author:**
Dr. Leena Patwardhan,
Consultant Pathologist,
Bai Jerbai Wadia Hospital
for Children, Mumbai,
Maharashtra, India.
Email: leenapatwardhan68@
gmail.com

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ABSTRACT

Pulmonary sequestration is a rare, mostly sporadic disorder, with an incidence of 0.1–6% of all congenital lung malformations. It is also called as bronchopulmonary foregut malformation (BPFM) or accessory lung. A 16-day-old female child presented with vomiting and inability to take feeds. She was investigated further and differentials included foregut duplication cyst and BPFM. Surgical exploration confirmed the diagnosis of a BPFM. Delay **in** diagnosis may lead to complications such as torsion and sepsis.

Keywords: Extrapulmonary sequestration, Broncho-pulmonary fore gut malformation, Neonate

INTRODUCTION

Pulmonary sequestration is a rare and mostly sporadic congenital malformation with an incidence of 0.1–6% of all congenital lung malformations. There are two distinct forms of pulmonary sequestrations: intralobar and extralobar, depending on whether it has independent pleura and its location of within the lobe of lung or outside lobes of the lung.[1-3]

This article presents a case of extralobar pulmonary sequestration that presented with incessant vomiting and inability to take feeds in a 16-day-old female. The extralobar pulmonary sequestration was in a rare transdiaphragmatic location.

CASE REPORT

A 16-day-old female child was admitted to the hospital with complaints of inability to take feeds and incessant vomiting. She was investigated and her ultrasonography and computed tomography (CT) scans of thorax and abdomen showed a para-esophageal mass at thoraco-abdominal junction. As the mass was causing partial obstruction at the gastroesophageal junction, she was operated upon. Intraoperatively, a small mass was found in the thoraco-abdominal region, across the diaphragm encircling the gastroesophageal junction. The post-operative follow-up was uneventful and the child was discharged on the 10th post-operative day.

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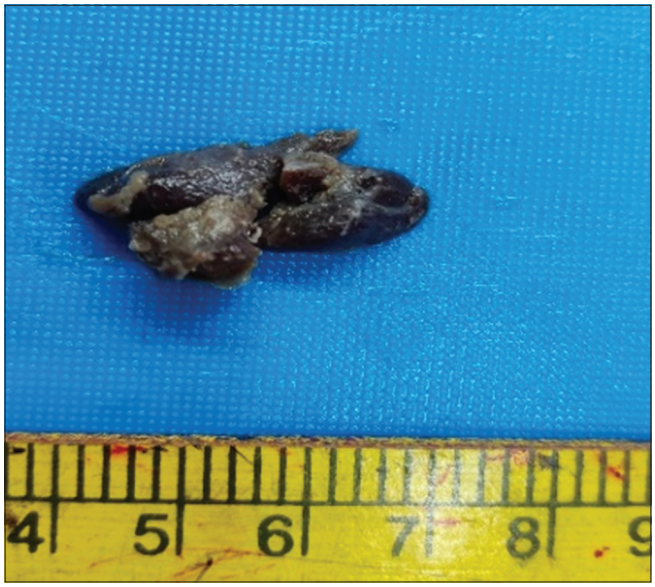

Figure 1: Macroscopic appearance

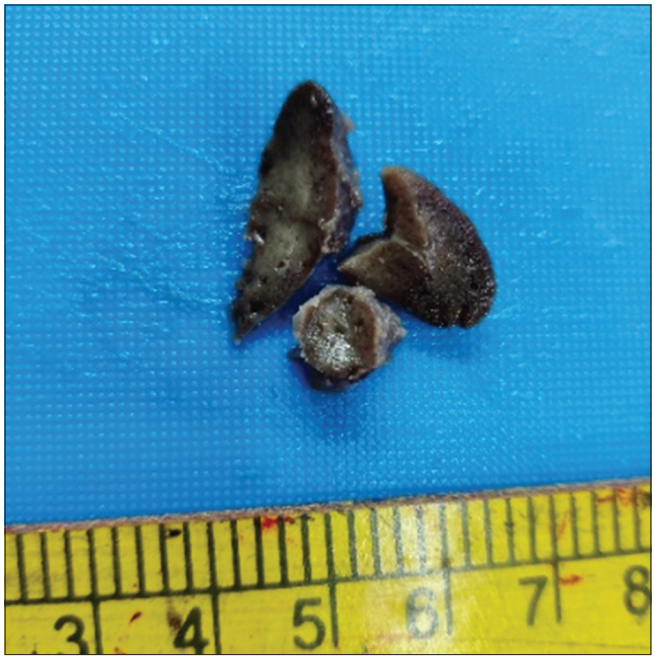

Figure 2: Cut surface showing porous appearance with few cystic spaces

Macroscopic findings

Macroscopic finding was suggestive of a 3.5 × 3 × 3 cm brownish-black soft-tissue mass with a porous appearance and its cut surface showed few cystic spaces [Figures 1 and 2]. Representative sections were taken for examination.

Microscopy finding

Hematoxylin and Eosin stained slides were examined under 40× original magnification which showed alveolar cystic spaces.

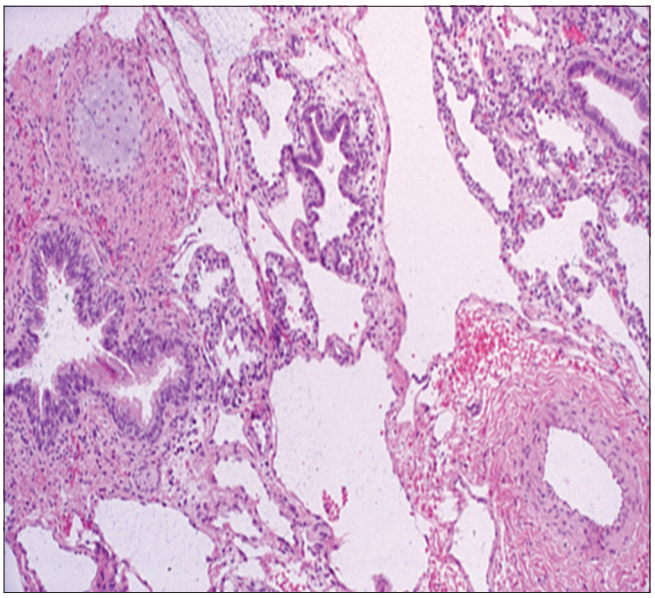

Figure 3: Microscopy, HE staining, 10x Magnification

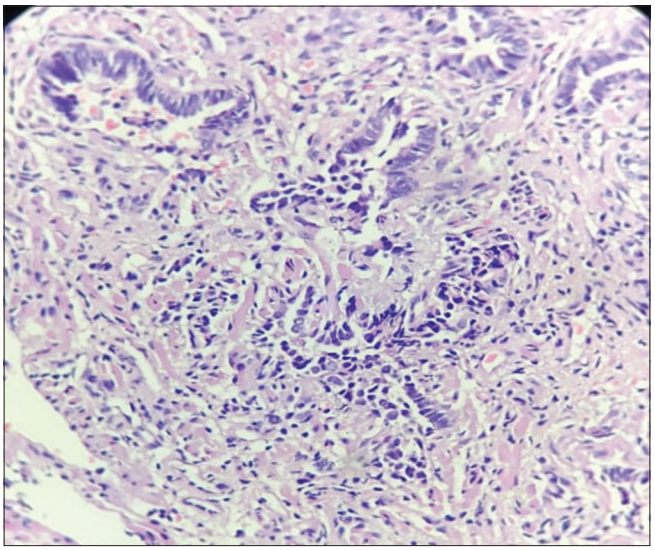

Figure 4: Microscopy, HE staining, 40x Magnification

alveolar cystic spaces lined by flattened epithelium with some showing bronchiolar metaplasia and few cartilage islands. There were few thick-walled congested vessels also seen at the periphery. There was mild chronic inflammation comprising of mature lymphocytes and few plasma cells [Figures 3 and 4]. Overall, the findings were suggestive of an extralobar pulmonary sequestration.

DISCUSSION

Pulmonary sequestration was first described by Pryce in 1946; although similar clinical findings were described by Rokitansky and Rektorik, as early as in 1861; it was not labeled as sequestration then.

Table 1: The differences between intralobar and extralobar sequestration.

	Intralobar Sequestration	Extralobar Sequestration
		• Intrathoracic • Subdiaphragmatic • Transdiaphragmatic
Relation with normal lung bud	Development before formation of pleura So, shares pleura of normal lung	Development after formation of pleura. So, has its own pleura
Association with other anomalies	11%	60%
Incidence	75%	25%
Side affected	66% cases are on left side. Rarely bilateral	90% on left side
Asymptomatic	85%	20%
Treatment is Surgical resection	Sequestrectomy	Lobectomy

Sequestrations (especially Extralobar) are associated with Congenital Pulmonary Airway Malformation (CPAM Type 2) which is then termed as a hybrid lesion. Extralobar sequestration may also be associated with congenital heart disease and congenital diaphragmatic hernia [Table 1]. Extralobar sequestrations do better than intralobar sequestrations.

Twenty-five percent cases are diagnosed prenatally by 18–19 weeks of gestation. Sixty percent of cases are diagnosed by 3 months of age.

Cancer Antigen (CA) 19-9 and Surfactant Protein A (SP-A) antibodies are raised in significant number of cases, hence, used for diagnosis. In our case, these investigations could not be done; as the patient presented in emergency settings.

Possible complications of pulmonary sequestration are hemoptysis, hemorrhagic pleural effusions, development of malignant tumors, and development of secondary infection-pneumonitis. In addition, torsion with hemorrhage and necrosis and sepsis is seen in extra lobar sequestration especially when the diagnosis is delayed - such cases are likely to present as acute surgical emergencies.

When there is extensive hemorrhage and necrosis due to torsion; immunohistochemistry for Thyroid Transcription Factor -1 (TTF-1) and Epithelial Membrane Antigen(EMA) is helpful.

CT angiography and other imaging studies are essential for understanding the vascular relations and pre-operative planning. However, its presence is best confirmed only on histopathology.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent.

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Nil.

Conflicts of interest

There are no conflicts of interest.

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