Preoperative percutaneous catheter drainage for symptomatic macrocystic congenital pulmonary airway malformation: A case report

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ABSTRACT

Introduction: Infants with congenital pulmonary airway malformation (CPAM) are generally asymptomatic and the surgical treatment can be planned using a thoracoscopic technique. We report the case of a newborn with a large type 1 CPAM who presented with severe respiratory distress and was treated with percutaneous transthoracic drainage before open surgery.

Case report: A full-term male patient was born via vaginal delivery. Antenatal imaging had raised suspicion of type 1 CPAM in the right lung. At birth, he was admitted to the neonatal intensive care unit (NICU) due to ventilatory insufficiency and oxygen dependence. A chest X-ray confirmed the antenatal diagnosis of a large type 1 CPAM. On the first days of life (DOL), we inserted a percutaneous transthoracic chest tube to drain the large cyst and initiated high-frequency oscillatory ventilation (HFOV). Although the patient initially showed clinical improvement, his condition subsequently deteriorated. Suspecting tube dislocation, on the seventh DOL a second drainage was placed in the cyst. On the tenth DOL, given the persistent clinical severity, a right upper lobectomy was performed. Postoperatively, the patient was supported by conventional ventilation with a reduced oxygen requirement. On the thirteenth DOL, the infant was successfully extubated. Histological analysis confirmed the diagnosis of type 1 CPAM. After nearly a month, the infant was transferred to the pediatric surgery ward and later discharged. During multidisciplinary follow-up, the patient maintained good general health with no signs of recurrence.

Conclusion: In newborns with large, symptomatic type-1 CPAMs a percutaneous catheter drainage can be used as a temporizing measure before the definitive surgical resection.

1. Introduction

Congenital pulmonary airway malformation (CPAM), formerly known as congenital cystic adenomatoid malformation (CCAM), is a rare developmental anomaly of the lower respiratory tract [1–3]. Although rare, it is the most common congenital lung lesion, with an estimated incidence of 1 in 25,000 to 1 in 35,000 live births. In 90 % of cases, CPAM is typically confined to a single lobe and is often detected through prenatal imaging [4,5].

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There are five types of CPAM, ranging from type 0 to type IV [6]. Type 1 CPAM is the most common, characterized by distinct thin-walled cysts measuring 2–10 cm in diameter. These cysts are usually solitary but may be multiloculated, and the lesions contain well-differentiated tissue. The clinical presentation of type 1 CPAM varies depending on the size of the cysts; affected patients may experience hydrops or respiratory distress during the neonatal period, or they may remain asymptomatic until later in life. In most cases, management and treatment can be planned, though the presence of a large malformation may complicate the clinical scenario, requiring emergent intervention.

We present the case of a large type 1 CPAM that manifested with respiratory distress in the neonatal period and was treated with percutaneous transthoracic drainage as a bridging procedure prior to open surgery.

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2. Case report

A full-term male patient was born at our hospital via vaginal delivery following a pregnancy complicated by gestational diabetes. His birth weight was appropriate for gestational age. Antenatal ultrasound and magnetic resonance imaging (MRI) suggested type 1 CPAM of the right lung with a mediastinal shift to the contralateral side, and the CPAM volume ratio (CVR) was measured at 1.5. At birth, the baby was admitted to the neonatal intensive care unit (NICU) due to ventilatory insufficiency and oxygen dependence, and high-frequency oscillatory ventilation (HFOV) was initiated. A chest X-ray performed upon admission supported the prenatal diagnosis (Fig. 1). On the first day of life (DOL), a percutaneous transthoracic drain was placed into the cyst under ultrasound guidance (Fig. 2) to reduce the lesion's volume and facilitate the transition to conventional mechanical ventilation (MV), which was necessary for transferring the patient to the operating room for lung resection. Although the patient initially showed clinical improvement, his condition worsened, leading to the suspicion of tube dislocation. Consequently, a second drain was placed in the cyst on the seventh DOL. On the tenth DOL, due to persistent clinical severity and difficulties in managing the patient with conventional MV, an emergency open lobectomy was performed. A right posterolateral thoracotomy was made at the fifth intercostal space. Upon exploration, the upper lung lobe was found to be enlarged, with a large cyst located centrally. The cyst appeared reduced in size, likely due to the drainage tube. However, after the tube was removed, the cyst rapidly refilled with air. Additionally, signs of hypoplasia were observed in the middle and lower lobes. The infant underwent resection of the right upper lobe, measuring 8.5 × 7 × 2.5 cm.

Postoperatively, the patient was hemodynamically stable, had two chest tubes in place, and was maintained on conventional ventilation with a FiO2 of 0.55. In the following days, the chest drains were successfully removed. However, on the eighth postoperative day (PO), the infant developed tachypnea, hypercapnia, and an increased oxygen requirement. A right pneumothorax was diagnosed, and a chest tube was reinserted for seven days, resulting in a modest reduction of the visible air collection on the chest X-ray, consistent with a prolonged air leak post-lobectomy. On the thirtieth DOL, following a weaning protocol from invasive ventilatory support,
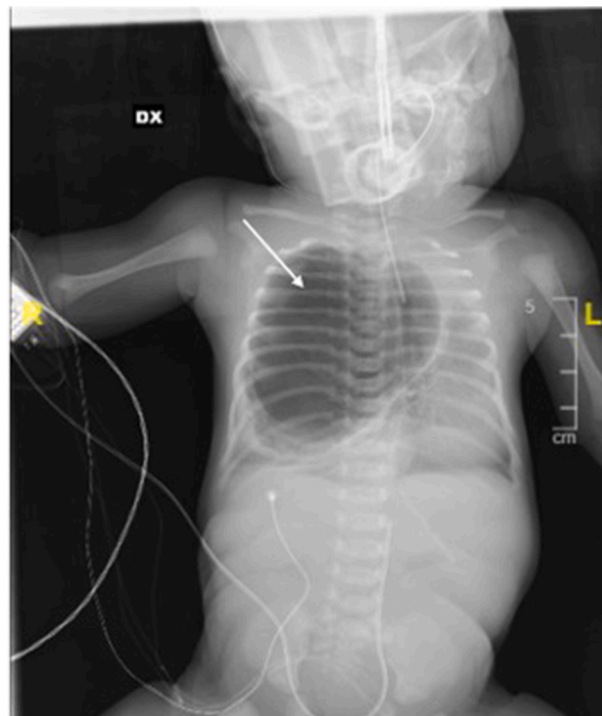

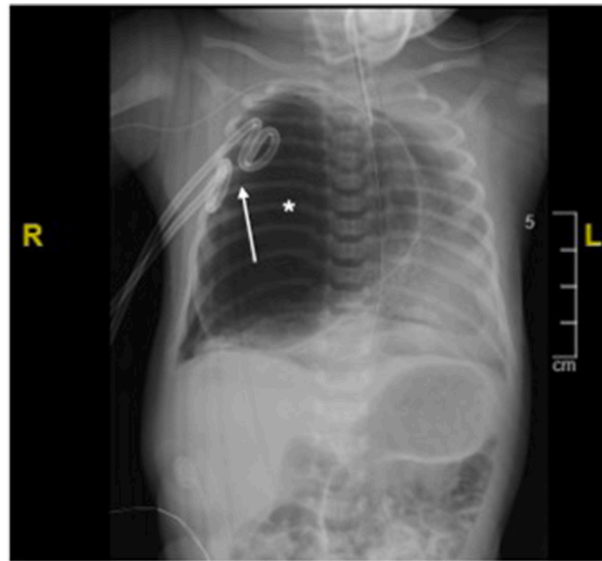**Fig. 1.** X-ray at birth (the cyst is indicated by the white arrow).

Fig. 2. X-ray after the placement of the second chest tube (indicated by the white arrow). The cyst is marked by the white asterisk.

the infant was successfully extubated. Histological examination confirmed the diagnosis of type 1 CPAM with superimposed acute abscess-forming inflammation.

At six weeks of age, the infant was transferred to the pediatric surgery ward and subsequently discharged with a multidisciplinary follow-up plan. Currently, the patient is in good general condition with no signs of recurrence.

3. Discussion

We present the case of a newborn with a large type 1 CPAM that caused severe respiratory insufficiency at birth, requiring invasive respiratory support and prompt surgical intervention.

CPAMs are congenital malformations originating from the lower respiratory tract. Stocker classified CPAMs into types 0 through IV, based on their embryological origins and histological characteristics. Type 0 arises from the tracheobronchial region, type I from the bronchial/bronchiolar region, type II from the bronchiolar region, type III from the bronchiolar/alveolar duct region, and type IV from the distal acinar region. Type 0 is the rarest, accounting for 2 % of all cases, while type 1 is the most common, comprising 60–70 % of cases [7].

The diagnosis of CPAM is increasingly made through prenatal imaging and is confirmed at birth by radiographic examinations. While a minority of lesions detected by fetal ultrasonography may resolve during gestation, most lesions present at birth will persist [2]. The main differential diagnoses for CPAM include pulmonary sequestration, congenital bronchogenic cyst, congenital lobar emphysema, pleuropulmonary blastoma, congenital cystic bronchiectasis, and congenital diaphragmatic hernia. The molecular mechanisms underlying CPAM formation remain unknown but may involve an imbalance between cell proliferation and apoptosis during organogenesis [8–12]. The clinical presentation of CPAM varies, and postnatal management depends on whether the patient is symptomatic. Approximately three-quarters of patients with a prenatal diagnosis of CPAM are asymptomatic at birth [13–16]. However, some of these infants may develop complications later in life, such as infections, pneumothorax, or hemoptysis. The risk of malignancy is particularly significant in type 1 and type 4 CPAMs [17]. The uncertainty surrounding the risks of infection and malignancy contributes to ongoing debate regarding the management of these patients [12]. The remaining 25 % of patients with CPAM are symptomatic at birth [13–15] and may require neonatal surgery if they experience severe respiratory distress. These infants often present with macrocystic CPAM, which exerts mass effects on the heart and lungs [18], as seen in our case. In other situations, it is preferable to postpone surgery until after the neonatal period [18,19]. Surgical intervention is curative and generally associated with few complications. In our clinical case, the persistent air leak observed post-lobectomy is a known complication documented in the literature [20]. Lobectomy is generally preferred over wedge resection due to the technical challenges in identifying dissection planes with certain lesions and the increased morbidity associated with partial resections [2,12,21]. A meta-analysis comparing thoracoscopy and open resection for CPAM found no significant differences in overall complications or surgery duration between the two approaches [22]. However, the minimally invasive approach was associated with a shorter hospital stay and fewer days requiring a chest tube. Emergency surgical intervention in the first few DOLis rare, as supported by literature data. For example, a French study [14] involving 89 infants with prenatal-diagnosed congenital pulmonary malformations (the majority being CPAMs) found that 25 % had abnormal breathing, 13 % experienced respiratory distress requiring oxygen supplementation or ventilatory support, and only 3 % underwent surgery before day 30 [14]. Severe respiratory distress at birth was best predicted by factors such as polyhydramnios, ascites, a CVR >0.84, or mediastinal shift [14]. In our case, the newborn exhibited both a mediastinal shift and a CVR >0.84. In a Dutch study of 80 patients with prenatal-diagnosed pulmonary malformations, 35 were confirmed to have CPAM after birth. Of these, 17 % re-

quired respiratory support (most often non-invasive) on the first DOLand underwent surgery within the first 28 days. Respiratory support at birth and surgery within 28 DOLwere needed in all cases of macrocystic CPAM or CVR >1.46 diagnosed prenatally [23]. Rare cases in the literature describe successful outcomes with cyst drainage followed by elective surgery [24–26]. However, in our case, drainage alone was insufficient to stabilize the patient and schedule the operation. Nonetheless, drainage did allow for temporary improvement in ventilatory parameters, enabling delayed open surgery. For most infants with CPAM and respiratory distress, surgical excision during the neonatal period is curative, and the prognosis is excellent [2,27–29]. This favorable outcome is likely due to compensatory growth and development of the remaining lung parenchyma [30]. The generally good prognosis associated with CPAM reflects the predominance of type 1 lesions. In our case, the patient had a favorable respiratory outcome, as confirmed by objective respiratory testing before discharge and normal follow-up results with pulmonologists.

4. Conclusion

For newborns with large, symptomatic type 1 CPAMs, percutaneous catheter drainage can serve as a temporizing measure before definitive surgical resection.

5. Patient consent

"Consent to publish the case report was not obtained. This report does not contain any personal information that could lead to the identification of the patient."

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Authorship

All authors attest that they meet the current ICMJE criteria for Authorship.

CRediT authorship contribution statement

Ilaria Acquaviva: Writing – review & editing, Writing – original draft, Formal analysis, Data curation, Conceptualization. **Edoardo Bindi:** Writing – review & editing, Conceptualization. **Giovanni Cobellis:** Validation, Supervision.

Declaration of competing interest

The authors declare that they have no known competing financial interests or personal relationships that could have appeared to influence the work reported in this paper.

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