DOI: 103126/JNPS.V4113

Case Report

Was It Pneumothorax? A Case Report of a Neonate with Congenital Pulmonary Airway Malformation from Rural Nepal

Pradip Kumar Paudel[1], Pragya Devkota[2], Bibek Devkota[3]

[1]Department of Paediatrics, Province Hospital, Surkhet, Karnali Province, Nepal
[2]Department of Radiology, Province Hospital, Surkhet, Karnali Province, Nepal
[3]Department of Anesthesia and Critical Care, Province Hospital, Surkhet, Karnali Province, Nepal

Article History
Received On : 27 Aug, 2021
Accepted On : 08 Jan, 2023

Funding sources: None

Conflict of Interest: None

Keywords: CPAM; ICD; neonate; pneumothorax

Online Access

DOI:
https://doi.org/10.3126/jnps.v42i2.39397

***Corresponding Author**
Pradip Kumar Paudel,
Department of Paediatrics,
Province Hospital,
Surkhet, Karnali Province, Nepal
Email: drpradippaudel@gmail.com

Abstract

Formerly called congenital cystic adenomatoid malformation (CCAM), congenital pulmonary airway malformation (CPAM) is a rare but most common congenital lung malformation that may manifest as an asymptomatic lesion at birth or in later life. The diagnosis may be difficult and is easily confused for pneumothorax often resulting incorrect ICD insertion on an emergency basis, which causes complications and delays accurate diagnosis. We report a case of a neonate, who presented with increasing respiratory distress at birth and hyperlucency in a chest radiograph initially misdiagnosed as pneumothorax and ICD inserted, later correctly diagnosed as CPAM type I with help of a CT chest. This case report is presented to highlight that CPAM is a rare cause of respiratory distress in newborns at birth.

Introduction

Formerly called congenital cystic adenomatoid malformation (CCAM), congenital pulmonary airway malformation (CPAM) is a rare lung malformation. It is the most common congenital lung malformation that may be asymptomatic or present with respiratory distress at birth. It may sometimes present in later life with respiratory issues. The aetiology is implicated in a variety of genes which results in dysplastic malformation of the tracheobronchial tree. It is resulted due to disruptive morphogenesis of the tracheobronchial tree at about the 35th day of life.[1,4] They occur usually as a unilobar and unilateral lesion. The reported incidence is between one in 8,000 to 35,000 with no predilection for the side of the lung, sex, or race.[2] Respiratory distress is the commonest presentation of symptomatic CPAM in the neonatal period and hyperlucency on x-ray can be easily confused with pneumothorax.[7,8] Here we report a rare case of CPAM in a neonate with a fatal outcome, which was treated with intercostal tube drainage (ICD) misdiagnosed as pneumothorax.

Case Report

A 28 - day - old female neonate was referred to our centre from a rural health post with a history of tachypnea and fever for five days. There was no history of cough, cyanosis, forced feeding or choking. The primigravida mother had done antenatal checkup at healthpost but no antenatal scan was done before delivery. The female baby was delivered vaginally at term with a 3.1 kg birth weight. The immediate neonatal period was uneventful. The mother noticed fast breathing and feeding difficulty at day 16 of life which progressively worsened. At presentation, she had marked respiratory distress with a respiratory rate of 86 per minute with intercostal and subcostal retractions. She

Copyrights & Licensing © 2022 by author(s). This is an Open Access article distributed under Creative Commons Attribution License (CC BY NC)

presented with a fever with a temperature of 101.3 F, HR 160 per minute, and SpO_2 76% with cyanosis. SpO_2 improved to 94% with oxygen. The trachea and the apex beat were shifted to the right side. There was a hyper-resonant percussion note, decreased breath sound on the left side, and scattered crackles on the right. The rest of the examination was normal.

The baby was shifted to NICU with a diagnosis of pneumonia. X-ray chest showed hyperlucency of the left lung and shifting of the mediastinum to right (Figure 1). The patient was given oxygen via headbox. A provisional diagnosis of pneumothorax was made and an intercostal drainage tube (ICD) was inserted. The baby was then kept on a mechanical ventilator as the condition did not improve. A repeat X-ray done afterwards revealed the same finding as the previous X-ray (Figure 2). Considering an alternative diagnosis, a CT chest was done. CT showed type I CPAM in the left lower lobe with significant contralateral mediastinal shift and hypoplasia of the right lung with iatrogenic pneumothorax (Figure 3). The baby was planned to refer to a higher center but the patient party were reluctant as they didn't believe the baby will improve. The baby succumbed on 4th- day in NICU.

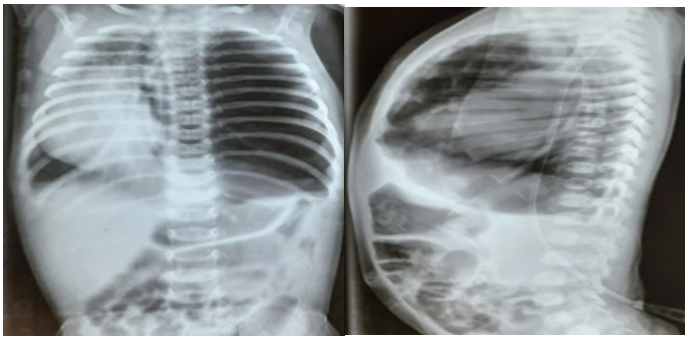

Figure 1. Initial chest radiograph showing large hyperlucency in left hemithorax, crossing the midline and extending to involve partly right hemithorax. The mediastinum including the trachea and cardia has been shifted to the right.

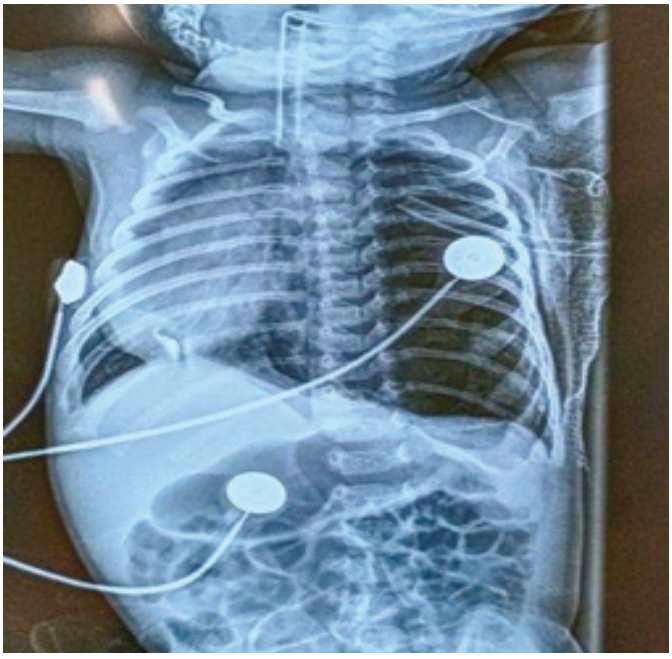

Figure 2. Follow-up chest radiograph after placement of intercostal drainage (ICD) tube at 5th intercostal space on the left side. Note that there is no interval resolution of hyperlucency in the left hemithorax, suggesting that the condition could be due to pulmonary pathology.

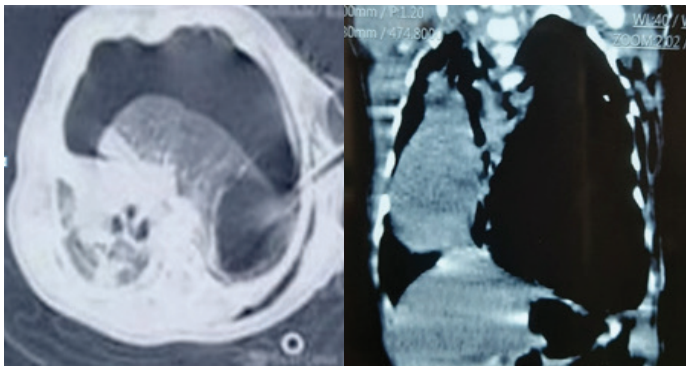

Figure 3. Axial and Coronal section on CT chest showing cystic lesion in left lower lobe with significant contralateral mediastinal shift, herniation of ipsilateral lung with hypoplasia of contralateral lung along with iatrogenic left-sided pneumothorax and subcutaneous emphysema. The tip of the chest tube drain is seen within the cyst.

Discussion

Our patient had CPAM, which was symptomatic till the late neonatal period and ultimately proved fatal without receiving definitive management for various reasons. Respiratory distress is the commonest presentation of symptomatic CPAM in the neonatal period as in our patient.

CPAM is classified into five categories as per Stocker's classification. This classification is based upon the size and epithelial lining of the cyst and is denoted as type 0 to type 4.[3] Type 1 is most common among all subtypes and consists of cysts that measure from 0.5 to 10.0 cm and are mostly limited to one lobe. Radiographically, it consists of one or more large air- or air / fluid-filled cysts in an expanded lobe that compresses the other lobes and shifts the mediastinum flattening the diaphragm.[2,3] The prognosis for type 1 lesions is excellent but poor for types 2 and 3.

The diagnosis of CPAM can be suspected on clinical examination and chest X-ray, but a CT chest is required to differentiate from others. Though antenatal ultrasonography can identify CPAM lesions[5] who are asymptomatic at birth, an antenatal scan wasn't performed in our case due to the unavailability of facilities in a rural setting. The differential diagnosis is all conditions that appear as hyperlucency on chest radiographs like pneumothorax, congenital lobar emphysema, bronchogenic cyst, pulmonary agenesis, foreign body aspiration, endobronchial tumor, congenital diaphragmatic hernia (CDH) etc.[1,6] The differentiation at times may be difficult if only clinical and x-ray findings are used and are easily confused for pneumothorax resulting incorrect ICD insertion on an emergency basis as in our case and other case reports.[7,10] This not only results in worsening of respiratory distress but may also lead to increased rates of complications such as pneumothorax, hydropneumothorax, subcutaneous emphysema, infections and delayed diagnosis.[7,9,10]

The definitive treatment of CPAM is surgical excision due to the risk of malignant transformation and recurrent pulmonary infection. Asymptomatic CPAM can be observed with serial imaging. Early

surgery between three to six months of age is technically easier and has the benefit of reduced infection risk and decreased need for respiratory support and allows more time for compensatory lung growth.[9] Due to the rural setting, we faced difficulty transferring sick neonates to higher centers (16 - 20 hours via ambulance) where surgery was possible, and this contributed to the fatal outcome.

Conclusions

CPAM is a rare congenital disorder presenting with progressive respiratory distress in the neonatal period. Paediatricians should think about alternative diagnoses if no improvement of the distress and non-expansion of the lung is seen even after ICD placement. One shouldn't hesitate to ask for a CT chest to diagnose other causes, such as CPAM or congenital lobar emphysema. Timely diagnosis will prevent unwarranted procedures and delayed diagnosis, and allow a prompt referral to facilities with thoracic surgery.

References

1. Blatter JA, Finder JD. Cystic adenomatoid malformation. In: Kliegman RM, Stanton BF, St Geme III JW, Schor NF, Berman RE, editors. Nelson Textbook of Pediatrics. 20th ed. Philadelphia: Elsevier Saunders; 2016. p. 2057-9.
2. Rosado-de-Christenson ML, Stocker JT. Congenital cystic adenomatoid malformation. Radiographics: a review publication of the Radiological Society of North America, Inc. 1991;11(5):865-86. DOI: 10.1148/radiographics.11.5.1947321
3. Stocker JT. Cystic lung disease in infants and children. Fetal Pediatr Pathol. 2009; 28: 155-184 [PMID: 19842869] DOI: 10.1080/15513810902984095
4. Annunziata F, Bush A, Borgia F, Raimondi F, Montella S, Poeta M, et al. Congenital Lung Malformations: Unresolved Issues and Unanswered Questions. Front Pediatr. 2019; 7:239. DOI:10.3389/fped.2019.00239
5. Durell J, Lakhoo K. Congenital cystic lesions of the lung. Early Hum Dev. 2014 Dec;90(12):935-9. DOI: 10.1016/j.earlhumdev.2014.09.014. Epub 2014 Oct 25. PMID: 25448785.
6. Wasilewska E, Lee EY, Eisenberg RL. Unilateral hyperlucent lung in children. AJR Am J Roentgenol. 2012 May;198(5): W400-14. DOI: 10.2214/AJR.11.7028. PMID: 22528920.
7. Anand M, Rahul Singh R, Bhaskaran A Congenital pulmonary airway malformation (Cpam): A diagnostic dilemma! JSM Clin Case Rep 2017; 5(3): 1135.
8. Disu EA, Kehinde OA, Anga AL, Ubuane PO, Itiola A. Akinola IJ, et al. Congenital pulmonary airway malformation, A case report of a rare cause of neonatal respiratory distress and review of the literature. Niger J Clin Pract 2019; 22:1621-5. DOI:10.4103/njcp.njcp_20_19
9. Calvert JK, Lakhoo K. Antenatally suspected congenital cystic adenomatoid malformation of the lung: postnatal investigation and timing of surgery. J Pediatr Surg. 2007 Feb;42(2):411-4. PMID: 17270559. DOI:10.3389/fped.2019.00239
10. Lee CY, Osman SS, Noor HM, Isa NSA. A missed late presentation of a congenital pulmonary airway malformation as a large infected bulla. Sultan Qaboos Univ Med J. 2018;18(4): e541-e4. DOI:10.18295/squmj.2018.18.04.020