ORIGINAL RESEARCH PAPER
Volume - 13 | Issue - 08 | August - 2024 | PRINT ISSN No. 2277 - 8179 | DOI : 10.36106/ijsr
INTERNATIONAL JOURNAL OF SCIENTIFIC RESEARCH

CONGENITAL PULMONARY AIRWAY MALFORMATION :- A RARE PRESENTATION IN A 3YR OLD

Paediatrics

Dr Athul PJ
Dr Chetan Tandel
Dr Najah Abdu Rahman
Dr Shamshad Khan

ABSTRACT

Congenital pulmonary airway malformation (CPAM) is a rare congenital abnormality with unknown exact aetiology or clear genetic association. It is characterised by a failure of bronchial development and localised glandular overgrowth. Typically, it is diagnosed on prenatal ultrasound, only infrequently in children, and even less commonly in adults. We present a case of a 3-year-old boy, with no previous lung diseases who presented with fever for 1 week and fast breathing for 2 days suggestive of pulmonary infection. Chest xray suggestive of right sided consolidation and CT scan showed thin multiple cystic lesion with internal septations in basal segments of right with surrounding consolidation in superior segment of right lower lobe and posterior segment of right upper lobe suggestive of type 2 CPAM with secondary infective pneumonia. He was started on intravenous antibiotics. Blood culture suggestive of methicillin-resistant Staphylococcus aureus. He improved with antibiotic treatment. He was discharged with 6-week course of antibiotics and follow-up afterward.

KEYWORDS

BACKGROUND:-

Congenital pulmonary airway malformation (CPAM) is a rare congenital abnormality that presents in 0.004% of all pregnancies, and constitutes ~25% of all congenital pulmonary anomalies.[1–3] It is characterised by failure of bronchial development and localised glandular overgrowth with no known aetiology or clear genetic association.[4–7] CPAM occurs equally across races and genders.[8] Typically, it is diagnosed prenatally[7] and only infrequently in children and adults.[1] When it presents in adults, the most common presentation is recurrent and resistant infections, even though some of these lesions are also found incidentally on chest imaging in asymptomatic patients.[4 5 8 9] CT scan, in general, is a reliable method for diagnosis and classification.[10] Stocker's criteria are used to classify CPAM from type 0 through type IV. This is ordered by the affected region of the lung from most proximal to most distal.[11–13] Type I CPAM is the most prevalent and is associated with the most favourable prognosis.[6]

Case presentation:-

A 3 year-old boy, with no medical history, presented to the emergency department with fever for 1 week and fast breathing for around 2 days. Parents denied any previous symptoms of respiratory infections even in neonatal period. He is the only son. No other significant family history of lung diseases.

At the emergency department, he had fever 103.0°F (39.4°C), tachycardia 160–170 bpm, blood pressure 90/53, respiratory rates 76 and oxygen saturation ~92% on room air. Chest examination revealed fast breathing, nasal flaring , inter costal retraction and subcostal retraction , decreased breath sounds in right infrascapular area, infra axillary area and dullness to percussion in the same area.

Investigation:-

Initial Complete blood counts showed white cell counts of 1000/mm^3 with neutropenia and elevated CRP level. On serial monitoring WBC showed increasing and CRP showed decreasing trend .Chest X-ray showed near-complete opacification of the right lower lobe (figure 1), this was followed up by HRCT showing thin multiple cystic lesion with internal septations in basal segments of right with surrounding consolidation in superior segment of right lower lobe and posterior segment of right upper lobe with secondary infective pneumonia (figure 2).

Radiologist suggested the it to be CPAM, which was previously named as congenital cystic adenomatoid malformation (CCAM) and more consistent with type 2 given the appearance of microcysts and the involvement of an entire lobe.

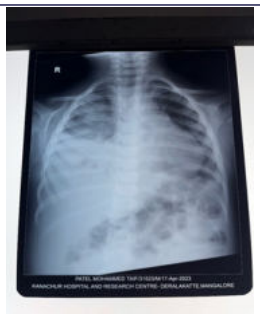

Figure 1

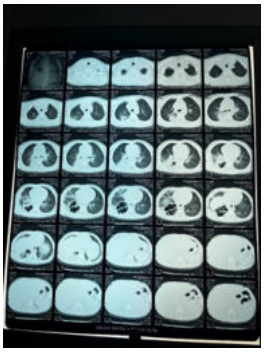

Figure 2

Treatment :-

He was admitted to the intensive care unit with the diagnosis of right lower lobe pneumonia and sepsis and was started on broad spectrum intravenous antibiotics, including vancomycin, ceftriaxone, amikacin as culture came positive for MRSA .Later on CT scan confirmed the diagnosis of CPAM.

As child started developing continuous fever spikes 4 days later , after initial control of fever and clinical improvement. Child was started with Clindamycin and child become afebrile 24 hr after the initiation of clindamycin.

Outcome :

In the following few days, he started to improve further clinically and his white cell counts decrease from 21k to 10 k and CRP decreased to 20 from 54. He was transferred to ward side. Vancomycin was stopped

after 14 days . Paediatric surgeon was involved and was advised for thoracostomy and lobectomy following 6 weeks treatment of patient with oral clindamycin. Follow up CECT scan of the chest planned before surgery.

DISCUSSION :

CPAM, which was previously named as congenital cystic adenomatoid malformation (CCAM), is a rare congenital abnormality that presents in 0.004% of all pregnancies,[1 2] and constitutes ~25% of all congenital pulmonary anomalies.[3] Its aetiology is poorly understood.[4 5] It is characterised by a failure of bronchial development and localised glandular overgrowth.[6 7] Its prevalence appears to be equal across race and gender,[8] and it lacks a clear genetic association.[4] Typically, it is diagnosed on prenatal ultrasound;[7] however, in the literature, it is also infrequently diagnosed in children and even less commonly in adults.[1]

When this anomaly is diagnosed after birth, there are a number of likely complications. The most common presentation in adults is recurrent and resistant infections. However, some of these lesions are also found incidentally on chest imaging in asymptomatic patients.[4 5 8 9] Other common presentations include pneumothorax, dyspnoea and haemoptysis.[5 8 9] Late presentation is typically diagnosed, and fairly reliably classified, by CT scan of the chest.[5 10] In adults, biopsy is often necessary to rule out malignancy.[1 9]

By Stocker's criteria, CPAM has been categorised from type 0 through type IV—ordered by the affected region of the lung from most proximal to most distal.[11] Type 0 involves the primary bronchi or trachea. This type is not frequently discussed in the literature, as those affected do not survive gestation.[12] Type I is the most prevalent, accounts for 60–70% of cases, and is associated with large (up to 10 cm) cystic lesions of the bronchi.[11] These patients have the most favourable prognosis.[6] Type II is characterised by bronchiolar involvement with many small (<2 cm) cysts.[11 12] This type carries a risk of a variety of other developmental abnormalities—most commonly other intrathoracic, abdominal or skeletal malformations.[11] Type III involves the distal bronchiolar and alveolar regions, and is associated with large lung masses with subcentimetre cysts.[2 11 12] Finally, type IV is associated with the most distal acinar region of the lung, with large cysts similar to type I.[11 12] Histologically, these cysts are made up of the same cell types as the region from which they were derived, from pseudostratified ciliated columnar in type I, transitioning to more cuboidal epithelium in types II and III, and finally squamous cells in type IV.[11 12] There has also been a variety of cross-over patterns described in the literature, among the CPAM classifications[10] and other congenital malformations, with bronchial atresia and bronchopulmonary sequestration being highly associated with CPAM.[7 10 13]

REFERENCES :-

1. McDonough RJ, Niven AS, Havenstrite KA. Congenital pulmonary airway malformation: a case report and review of the literature. *Respir Care* 2012;57:302–6. 10.4187/respcare.00727 [PubMed] [CrossRef] [Google Scholar]
2. Morelli L, Piscioli I, Licci S et al.. Pulmonary congenital cystic adenomatoid malformation, type I, presenting as a single cyst of the middle lobe in an adult: case report. *Diagn Pathol* 2007;2:17 10.1186/1746-1596-2-17 [PMC free article] [PubMed] [CrossRef] [Google Scholar]
3. Oh BJ, Lee JS, Kim JS et al.. Congenital cystic adenomatoid malformation of the lung in adults: clinical and CT evaluation of seven patients. *Respirology* 2006;11:496–501. 10.1111/j.1440-1843.2006.00878.x [PubMed] [CrossRef] [Google Scholar]
4. Feng A, Cai H, Sun Q et al.. Congenital cystic adenomatoid malformation of lung in adults: 2 rare cases report and review of the literature. *Diagn Pathol* 2012;7:37 10.1186/1746-1596-7-37 [PMC free article] [PubMed] [CrossRef] [Google Scholar]
5. Luján M, Bosque M, Mirapeix RM et al.. Late-onset congenital cystic adenomatoid malformation of the lung. Embryology, clinical symptomatology, diagnostic procedures, therapeutic approach and clinical follow-up. *Respiration* 2002;69:148–54. [PubMed] [Google Scholar]
6. Shanmugam G, MacArthur K, Pollock JC. Congenital lung malformations—antenatal and postnatal evaluation and management. *Eur J Cardiothorac Surg* 2005;27:45–52. 10.1016/j.ejcts.2004.10.015 [PubMed] [CrossRef] [Google Scholar]
7. Ioachimescu OC, Mehta AC. From cystic pulmonary airway malformation, to bronchioloalveolar carcinoma and adenocarcinoma of the lung. *Eur Respir J* 2005;26:1181–7. 10.1183/09031936.05.00011705 [PubMed] [CrossRef] [Google Scholar]
8. Hellmuth D, Glerant JC, Sevestre H. Pulmonary adenomatoid malformation presenting as unilobar cysts in an adult. *Respir Med* 1998;92:1364–7. 10.1016/S0954-6111(98)90143-3 [PubMed] [CrossRef] [Google Scholar]
9. Zylak CJ, Eyler WR, Spizarny DL. Developmental lung anomalies in the adult: radiologic-pathologic correlation. *Radiographics* 2002;22:S25–43. 10.1148/radiographics.22.suppl_1.g02oc26s25 [PubMed] [CrossRef] [Google Scholar]
10. Kim WS, Lee KS, Kim IO et al.. Congenital cystic adenomatoid malformation of the lung: CT-pathologic correlation. *AJR Am J Roentgenol* 1997;168:47–53. 10.2214/ajr.168.1.8976918 [PubMed] [CrossRef] [Google Scholar]
11. Malinova DV, Kolova PL, Radev RS. Congenital cystic adenomatoid malformation of the lung: a case report and review of the literature. *J Biomed Clin Res* 2015;8:160–3. [Google Scholar]
12. Yikilmaz A, Lee EY. CT imaging of mass-like nonvascular pulmonary lesions in children. *Pediatr Radiol* 2007;37:1253–63. 10.1007/s00247-007-0637-4 [PubMed] [CrossRef] [Google Scholar]
13. Riedlinger WF, Vargas SO, Jennings RW et al.. Bronchial atresia is common to extralobar sequestration, intralobar sequestration, congenital cystic adenomatoid malformation, and lobar emphysema. *Pediatr Dev Pathol* 2006;9:361–73. 10.2350/06-01-0023.1 [PubMed] [CrossRef] [Google Scholar]
14. Avitabile AM, Greco MA, Hulnick DH. Congenital cystic adenomatoid malformation of the lung in adults. *Am J Surg Pathol* 1984;8:193–202. 10.1097/00000478-198403000-00005 [PubMed] [CrossRef] [Google Scholar]