HYBRID LUNG LESION: AN ACCIDENTAL FINDING

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Introduction

Pulmonary sequestrations and congenital pulmonary airway malformations are a spectrum of lung anomalies that are well known, yet uncommon. When congenital pulmonary airway malformation is associated with a systemic blood supply it is termed as a "Hybrid" lesion. This term was coined by Children's Hospital of Philadelphia (CHOP) in 1998. This abnormal mass of non-functioning lung tissue may be further categorized as extralobar or intralobar. While the extralobar hybrid lesions are well described, literature on the intralobar lesion is scarce.1 We describe a 6 year old boy with a hybrid lesion consisting of an intralobar pulmonary sequestration associated with a type II Congenital Pulmonary Airway Malformation (CPAM). This is the first case of a hybrid lesion in a child reported in Malaysia.

Case Summary

A 6 years old boy presented with fever and cough for 4-5 days was treated for upper respiratory tract infection. The chest xray revealed massive left sided pleural effusion. Subsequently, child developed decompensated septic shock requiring inotrope and was intubated. Ultrasound thorax revealed a large heterogeneous highly vascular lesion in the left lower hemithorax with blood supply arising from the abdominal aorta, superolateral to origin of the coeliac artery. The CECT thorax showed a heterogeneously enhancing mass at the left hemithorax measuring 10.8x11.9x13.1 cm, with numerous fluid-filled cyst within, arterial supply arising from abdominal aorta at the level of the coeliac trunk and venous return into left pulmonary vein. There is also a mediastinal shift with a left sided pleural effusion.

We performed an ultrasound guided biopsy to rule out pleuropulmonary blastoma which revealed no malignant cell, hence a thoracotomy , left lower lobe lobectomy and chest tube insertion were performed. Intraoperatively, left lower lobe was filled with pus and adhered to diaphragm, a feeding vessel from a branch above the coeliac artery was noted piercing through the diaphragmatic tissue. Post-operative complications were haemothorax and recurrent pneumonias with parapneumonic effusions requiring prolonged ventilation. Subsequently, child improved and was discharged well with regular uneventful follow ups. HPE showed various dilated cystic spaces lined by cuboidal to ciliated pseudostratified columnar epithelium with minimal foci of cartilaginous tissue in intervening area suggestive of CPAM most probably type II with associated inflammatory changes and fibrosis.

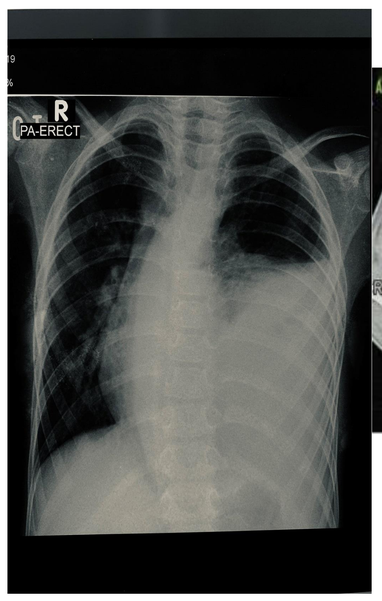

Figure 1. Shows erect chest radiograph during his presentation which shows left massive pleural effusion.

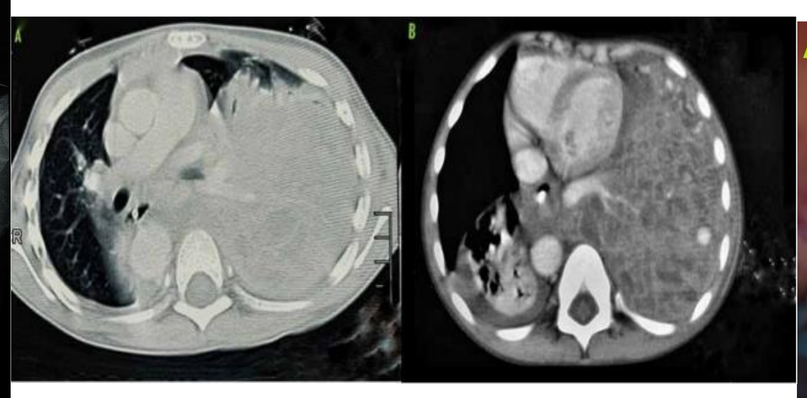

Figure 2 A, B Shows CT images of the hybrid lung lesion with the feeding vessel

Discussion

The intralobar pulmonary sequestration associated with type II CPAM is a congenital lung malformation described as a non-functioning lung tissue invested within the same pleura as normal lung, with an aberrant arterial supply from the systemic vasculature and drainage into the pulmonary or systemic veins. The non-functioning lung tissue displays features of type II CPAM with multi cystic areas of over proliferation and dilatation of terminal respiratory bronchioles, lacking normal alveoli.[2,3] Literature suggest these lesions to be commonly symptomatic during the neonatal or infancy period and are rarely present during childhood.[4,5,6,7,8,9] .However, Pierre Goussard et al[10] and Fatima Naumeri et al[11], reported hybrid lesions in older children, with similar presentation to our case. Therefore, differential diagnosis of hybrid lung lesion in children with recurrent pneumonia should be considered.

Savic et al from 1862-1978 in his extensive review commented 86% of patients had hybrid lesions in the left lower lobe, as seen in this child[12]. Common radiological studies used are chest x-ray, ultrasound and CECT Thorax. Involution of hybrid lesions with significant systemic arterial supply is highly unlikely.

To prevent post op complications and morbidities such as recurrent pneumonias, haemoptysis and malignant transformations, surgical resections ought to be performed electively or once child is optimized. Besides, an earlier intervention helps reduce parental anxiety.[7] Choosing between thoracotomy and thoracoscopic surgery is widely controversial, however a meta-analysis by A.Nasi,j.bass[13] proved no difference pertaining complications and duration of surgery but a thoracoscopic approach offers better cosmesis with lesser post-operative pain and shortens hospital stay.

In this child, the diagnosis of hybrid lesion was confirmed as the intraoperative findings of the left posterolateral thoracotomy correlated with the CECT and HPE reports.

Conclusion

The diagnosis of a hybrid lesions should be in the list of differentials when treating children with recurrent episodes of pneumonia. Radiological features of these hybrid, Early surgical resection is advised to reduce the risk of morbidity and parental anxiety.

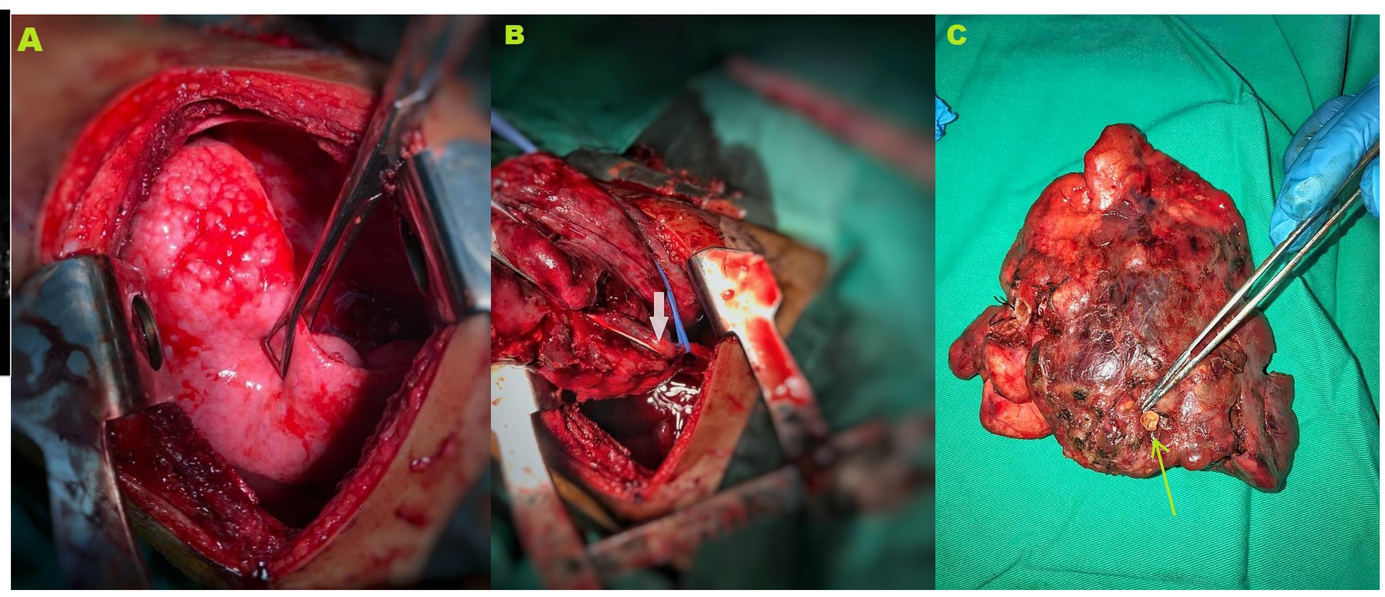

Figure 3A. Shows the affected lung.
Figure 3B. Arrow shows the feeding vessel to the hybrid lesion.
Figure 3C. Shows the gross sample of the excised hybrid lesion with the arrow showing the feeding vessel.

References

1. The Children's Hospital of Philadelphia. (2019, July 12). Hybrid lesions. Children's Hospital of Philadelphia. Retrieved February 20, 2022, from https://www.chop.edu/ conditions-diseases/hybrid-lesions
2. Jonathan Durell, Kokila Lakhoo,Congenital cystic lesions of the lung,Early Human Development,Volume 90, Issue 12,2014,Pages 935-939,ISSN 0378-3782, https://doi.org/10.1016/j.earlhumdev.2014.09.014. (https://www.sciencedirect.com/science/ article/pii/ S0378378214002394)
3. Darren J. Fowler, Steve J. Gould, The pathology of congenital lung lesions,Seminars in Pediatric Surgery,Volume 24, Issue 4,2015,Pages 176-182,ISSN 1055-8586, https://doi.org/10.1053/j.sempedsurg.2015.02.002. (https://www.sciencedirect.com/science/ article/pii/S1055858615000281)
4. Herman, T., Siegel, M. Congenital intralobar pulmonary sequestration with cystic adenomatoid malformation. J Perinatol 29, 175-177 (2009). https://doi.org/10.1038/ jp.2008.90
5. Orpen, N., Goodman, R., Bowker, C. et al. Intralobar pulmonary sequestration with congenital cystic adematous malformation and rhabdomyomatous dysplasia. Ped Surgery Int 19, 610-611 (2003). https://doi.org/10.1007/ s00383-003-1040-3
6. Darrell L Cass, Timothy M Crombleholme, Lori J Howell, Perry W Stafford, Eduardo D Ruchelli, N.Scott Adzick, Cystic lung lesions with systemic arterial blood supply: A hybrid of congenital cystic adenomatoid malformation and bronchopulmonary sequestration,Journal of Pediatric Surgery, Volume 32, Issue 7,1997,Pages 986-990, ISSN 0022-3468, https://doi.org/10.1016/S0022- 3468(97)90383-3. (https://www.sciencedirect.com/ science/article/pii/S0022346897903833)
7. Samuel M, Burge DM. Management of antenatally diagnosed pulmonary sequestration associated with congenital cystic adenomatoid malformation. Thorax. 1999;54(8):701-706. doi:10.1136/thx.54.8.701
8. Zeidan, S., Hery, G., Lacroix, F. et al. Intralobar sequestration associated with cystic adenomatoid malformation: diagnostic and thoracoscopic pitfalls. Surg Endosc 23, 1750- 1753 (2009). https://doi.org/10.1007/ s00464-008-0183-7
9. S c h u s t e r B , H o l l a n d - C u n z S G , Z imm e rm a n n PIntradiaphragmatic hybrid lesion: surgical decisionmaking and value of minimal invasive surgeryCase Reports 2017;2017:bcr-2016-218527.
10. Goussard P, Andronikou S, Mfingwana L, et alHybrid lesion in a child presenting with cough, fever and haemoptysisBMJ Case Reports CP 2020;13:e238796.
11. Naumeri, Fatima & Sajjad, Muhammad. (2018). Hybrid Lesion: Extralobar Sequestration With Cystic Adenomatoid Malformation - Misdiagnosed As Pulmonary Tuberculosis. Journal of the College of Physicians and Surgeons Pakistan. 28. S204-S206. 10.29271/jcpsp.2018.09.S204.
12. Savić B, Birtel FJ, Knoche R, Tholen W, Schild H. Pulmonary sequestration. Ergeb Inn Med Kinderheilkd. 1979;43:57- 92. doi: 10.1007/978-3-642-67379-5_2. PMID: 394961.
13. Nasr A, Bass J. Thoracoscopic vs open resection of congenital lung lesions: a meta- analysis.JPediatr Surg. 2012 May;47(5):857-61. doi: 10.1016/j.jpedsurg.2012.01.036. PMID: 22595561.