

Incidental finding - the discovery of a bronchopulmonary foregut malformation through investigations for Crohn's disease

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ABSTRACT

Pulmonary sequestration (PMS) is a rare bronchopulmonary malformation. It has an incidence of between 0.15% to 1.7%. Likewise, cystic adenomatoid malformation (CCAM) is another relatively rare category of a bronchopulmonary malformation with a reported incidence of between 1 in 25,000 to 1 in 35,000. Moreover, a bronchopulmonary malformation with features allied to both of these forms is considered an even rarer entity. In general, bronchopulmonary malformations present with a range of non-specific symptoms. Radiological features can be non-specific yet distinctive when related to clinical features. Ultimately, definitive diagnosis depends upon histological assessment of lung tissue. We present an adult female with radiological features of both pulmonary sequestration and cystic adenomatoid malformation. This was an incidental finding unrelated to the patients presenting complaint. This case highlights the importance of using a structured and systematic approach when interpreting medical imagery.

CASE REPORT

CASE REPORT

A 30-year-old Caucasian female was referred to the Gastroenterology Clinic with intermittent post-prandial left iliac fossa pain. In addition, she also described recently having loose stools and suffering with generalised joint pain. Her past medical history was significant for a longstanding history of loose stools and abdominal pain. This was previously investigated by means of a magnetic resonance imaging (MRI) and colonoscopy. Following this a diagnosis of Crohn's disease was made in 2001. Subsequently, she was successfully medically treated for her Crohn's disease. However, in 2014 following an episode of abdominal pain she had a laparotomy which revealed no abnormalities. Nevertheless, she continued

to follow a strict FODMAP diet in an attempt to alleviate what was considered to be her functional gastrointestinal symptoms. The only additional report from her past medical history included a single incidence of left sided pneumonia which required a hospital admission and raised an unconfirmed diagnosis of bronchiectasis. There were no other overt complaints or written documentations of any other respiratory symptoms.

On examination, she weighed 48.55kg, was not pale or jaundiced. There was no evidence of peripheral lymphadenopathy and her abdomen was soft and slightly tender in both iliac fossae. No mass or organomegaly was identified. At the time the best course of actions was

determined as to explore whether her Crohn's disease was active and the extent of any active Crohn's disease.

Accordingly, investigations ordered included, full blood count, haematinics, liver function test, renal function test and inflammatory markers. However, these investigations were all within normal range. In addition, a MRI was also requested and distribution showed normal enteric structures. However, a 5cm haemorrhagic right ovarian cyst and several very small corpus luteal cysts were incidentally identified; potentially explaining her pelvic discomfort. Moreover, a further unexpected yet interesting observation was made. In particular, an anomalous artery was identified (figure 1). The artery was noticed as originating from the coeliac axis and supplying the lower lobe of the left lung. Subsequently, computed tomography (CT) of the chest with CT angiography was recommended and this established the presence a bronchopulmonary malformation (figures 2, 3A and 3B). In particular, a rudimentary bronchial tree was identified which did not appear to communicate with the main tracheobronchial system. Furthermore, it also appeared to be partially occluded by debris. Additionally, observed within this segment of lung were numerous small cysts ranging from 0.2cm to 2cm in size. These features are recognised radiological features pertaining to both PMS and CCAM. Accordingly, a hybrid type malformation is suspected.

DISCUSSION

Etiology & Demographics:

In 1946 Pryce [1] first coined the term and identified the anatomy of a pulmonary sequestration (PMS). He specified that a PMS comprised a distinctive segment of lung, which could be a bronchopulmonary mass or a cyst, satisfying two conditions. Firstly, that it exists dissociated from the normal tracheobronchial tree and, secondly, that it be in receipt of an arterial blood supply from one or more systemic arteries as opposed to the customary pulmonary vessels [1]. Furthermore, PMS are also categorised as either intralobar or extralobar[1]. The former arises when visceral pleura is shared between normal lung tissue and adjacent abnormal tissue [1–3]. This is in contrast to the latter which occurs when there is a separate independent pleura encasing the abnormal lung viscera [1–3]. This differentiation is understood to be consequential to the particular embryological stage at which the fault in foetal lung and vasculature development arises [1]. Nonetheless, this is a very rare malformation with an incidence believed to be in the range of 0.15% to 1.7% [4]. On the other hand, congenital cystic adenomatoid malformation (CCAM) is another form of bronchopulmonary malformation [5]. It is considered a intraparenchymal lung disease characterised by the absence of normal alveoli and the presence of multiple variably sized cysts present within a segmental area of lung tissue [5,6]. It was first described by Chin and Tang in 1949 and is reported to have an estimated incidence of between 1 in 25,000 to 1 in 35,000 pregnancies [5–7]. CCAM can be classified into one of three categories depending upon the clinical, radiological and pathological assessments of the abnormal lung parenchyma [5,7]. Furthermore, in contrast to pulmonary sequestrations,

CCAMs are typically connected to the normal tracheobronchial tree and draw blood from the normal pulmonary circulations [6].

Hybrid IPS and CCAM are relatively rare bronchopulmonary malformations with no reported incidence [6],[8]. Here we have presented a case report of a young female demonstrating radiologically features of both intralobar pulmonary sequestration (IPS) and CCAM. To the best of our knowledge this is the first case report describing an adult with a IPS, with certain features of a CCAM, in receipt of an anomalous vessel arising from the celiac artery.

If we are to adhere specifically by Pryce's [1] original description of a PMS a hybrid lesion should be viewed as a IPS with accompanying CCAM features. With regards to our case, we incidentally uncovered a left sided bronchopulmonary malformation involving an abnormal artery arising from the coeliac trunk. From a radiological perspective, this malformation has features of both ILS and CCAM, therefore, it is described as a hybrid type malformation. They have previously been recognised and described in the literature to a limited extent. In 1979, Savic's[4] analysis of 100 pathology reports identified that bronchiectatic forms of IPS can exist with polycystic features[4]. Furthermore, in 1997 Cass[6] described treating patients who had IPS with CCAM features. Between 1949 and 1999, 36 cases of pulmonary sequestration associated with CCAM have been described[8]. More recently there have been a number of case reports documenting the descriptions of a PMS with features of CCAM [9–18]. However, the majority of these describe a extralobar pulmonary sequestration (EPS) malformation. We were only able to identify 7 cases pertaining to a IPS with CCAM associated features, [6,11,12] none of which refer to an aberrant artery arising from the celiac artery.

Regarding ILS, it is understood that the posterior basal segments of the lower lobes are the most commonly effected zones [19–21]. In addition, left sided malformations are reported in the literature to be more frequent than right sided [19,21]. In general, the arterial blood supply for ILS is thought to arise from the descending thoracic artery 74% of the time compared to 19% originating from the abdominal aorta [19]. Although one of the earliest reports of an anomalous artery, associated to an ILS, arising from below the level of the diaphragm was described by Pryce in 1946 [1]. Since 1946 we have been able to identify a number of reports which describe a ILS consisting of an anomalous artery derived from the celiac trunk [19,22–32]. Interestingly, the majority of the reports that we have identified refer to a right sided ILS with a celiac artery derived feeder vessel. We were only able to identify 3 reports that describe a left sided ILS with an anomalous celiac artery derived blood supply [29–31]. This would be the fourth report of a left sided ILS with a celiac artery derived bloody supply.

Clinical & Imaging findings:

ILS appears to encompass a spectrum of clinical presentations[1–3]. In particular, presentations can range from

an incidental finding in an otherwise asymptomatic individual to individuals who suffer with chronic bouts of bronchitis and recurrent pneumonia [1-4,21,33]. Particular symptoms which may be observed include chest pain, dyspnoea and haemoptysis [20,25,27,34,35]. Due to a combination of non-specific presenting symptoms, coupled with the rare nature of PMS, a misdiagnosis rate of 87% has been recognised [20]. The majority of cases of ILS are diagnosed in the second decade of life and are rarely diagnosed in those older than 40 [2,33]. Overall, there is no gender predilection [1,2] although with advancement in age there appears to be a preference towards males being diagnosed with ILS [33].

A variety of imaging modalities can be utilised during the assessment of a potential PMS. Doppler ultrasonography (DUS) is an effective imaging modality for the antenatal identification of a PMS [36]. DUS has the capability to recognise an aberrant vessel that may be feeding an abnormal lung mass which is perceived as an abnormal area of echogenicity [36]. However, antenatal diagnosis requires serial imaging which can occasionally inaccurately demonstrate a disappearing lesion which is actually present postnatally [8,37]. In addition, it can be difficult to establish a diagnosis when using DUS as no specific PMS signs exist [7,8]. Chest radiography is a frequently used investigation for patients presenting with respiratory symptoms [20,25,27,35]. It is quick to perform, widely available, relatively cheap, and can direct the requirement for further investigation, consequently it is generally used as a first line investigation [20,25,27,35]. Common signs observed on a chest radiograph include radiopaque areas which may be indicative of nodules, reticular infiltration or masses [20,35]. These radiographic findings are frequently confused with other lung conditions such as pneumonia [35,36]. Hence, due to the non-specific nature of these radiographic signs and lack of capacity for the identification of vasculature, chest radiography is not considered to be specific enough when attempting to diagnose a PMS. CT imaging is commonly used when further assessment of the lung parenchyma is deemed necessary and to establish a definitive diagnosis of a PMS [20,21,29,35]. CT imaging is better able to delineate the anatomy of a suspected PMS [20,36]. Anatomical structures which can be identified on CT include a lung mass, a cyst and a fluid or air filled cavity [20,36]. Moreover, contrast enhanced CT imaging and CTA are considered the modalities of choice for the identification of arterial vasculature [36]. In particular, with CTA vessels are demarcated as they are filled with contrast. This component of imaging, when factored in with the identification of the lung lesion, is fundamental for a definitive PMS diagnosis. Interestingly, pulmonary angiography is considered the gold standard for the diagnosis of a PMS [20]. Nevertheless, CTA and magnetic resonance angiography (MRA) are utilised more often. This is for a number of reasons; firstly, CTA and MRA are less invasive. Secondly, both are able to identify the arterial vasculature to a good level of detail and are considered to give superior information with regards to the lung parenchyma. Finally, they are considered safer alternatives [20].

Treatment & Prognosis:

When a diagnosis has been established, active management is usually considered the best option [22]-[32]. Surgical resection, in particular an open lobectomy, is the mainstay of treatment for IPS and offers a good prognosis [20,21,35]. It is considered for all patients that are symptomatic and suitable for surgery [36]. The earlier surgery is performed the better the prognosis for two main reasons. IPS is associated with chronic pulmonary infection which from a septic point of view may be fatal [21]. In addition, longstanding pulmonary inflammation can destroy the lung parenchyma [35], both sacrificing lung function and ultimately making surgery more challenging due to the loss of surgical margins, risking a larger resection [21]. In those who are asymptomatic, the benefits of surgery have been disputed [21]. However, due to the risk of recurrent infection and subsequent need for a larger resection surgery is generally encouraged, nevertheless, conservative management may also be considered as an option in those who are asymptomatic [21]. Novel therapeutic options which have been explored include the endovascular occlusion of the anomalous vessel and resection via minimally invasive techniques such as video assisted thoracoscopic surgery [29,36].

Differential Diagnosis:

With regards to intralobar pulmonary sequestration the following differential diagnoses are considered: bronchiectasis, bronchogenic cysts, empyema, lung abscess, pneumonia and pulmonary neoplasm. Due to the relative commonness of these diagnosis interpretation and description of imaging has been more commonly established. In the majority of cases a chest radiograph is performed in the first instance as it is quick and provides important initial details. CT and MRI are subsequently performed as second line investigations if more detail is warranted.

Bronchiectasis

The chronic airway dilatation and mucus secretion which occurs in bronchiectasis results in alteration in the lung parenchyma which can be observed on chest radiography and CT imaging [20][35][36]. When considering chest radiography, tram-track opacities which can be observed are produced by thickened bronchial walls, increased bronchovascular markings with ill-defined pulmonary vasculature and ring shadows that are created by viewing terminal bronchi front-on can direct towards a diagnosis of bronchiectasis. Nevertheless, CT is considered the superior imaging modality. Three classical imaging features observed include the tram-track sign, signet sign and honeycomb appearance.

Bronchogenic cysts

These congenital lesions are observed on chest radiography as variably sized yet well circumscribed areas of increased opacity [20]. Furthermore, CT imaging can be used and demonstrates spherical or tubule masses of homogenous or variable attenuation [35]. Moreover, MRI imaging may also be utilised. In particular, areas corresponding to bronchogenic

cysts on T1 weighted MRI demonstrates variable signal intensity, whereas, on T2 weighted MRI high signal intensity regions are observed [38].

Pulmonary empyema

Pus which gathers in the pleural space is normally a consequence of a background pathology process. Chest radiography and CT are routinely requested when there is any suspicion of pus in the pleural space [33]. The former demonstrates a unilateral-asymmetric biconvex fluid level which can be difficult to distinguish from a pleural effusion. While the latter identifies fluid collections as increased density in the pleural space which can sometimes arise with locules of gas. In addition, a thickened pleura may be observed and the split pleura sign can be appreciated.

Lung abscess

Chest radiography and CT imaging are typically utilised to investigate this circumscribed collection of pus within lung tissue [33]. Chest radiography typically demonstrates a cavity in the lung which contains a fluid level. CT imaging also identifies the cavity and the air-fluid level, as well as, demarcating the wall of the cavity.

Pneumonia

Inflammation of the alveoli produces characteristic radiological features and signs which are identified with chest radiography and CT imaging [21]. Less frequently MRI is used [38]. In particular, with a chest radiograph areas of homogenous opacification are typically seen in a lobar pattern. Furthermore, when utilising CT imaging ground glass opacity is observed correspondingly in a lobar pattern. However, if the inflammation is extensive non-lobar patterned segments may be involved. Less commonly MRI is used; on a T2 weighted scan areas of high signal are observed from the lung areas that are infected.

Pulmonary neoplasm

Chest radiography and CT imaging are normally used when investigating suspected pulmonary positioned neoplasms [36]. When utilising chest radiography, no specific findings as such are observed. However, radiological features such as hilar or perihilar opacification with mediastinal widening and central or peripheral nodules/zones of increased opacification may be indicative of a pulmonary neoplasm. With CT imaging, greater detail can be determined with reference to viewing of enlarged lymph nodes, infiltration of neoplasm into adjacent structures, lung atelectasis, cavitation and haemorrhage or necrosis of lung tissue associated with a neoplasm.

Outcome and follow-up

Resection of the congenital pulmonary malformation was considered as it was deemed to be the likely cause of this patient's bronchiectasis features. Furthermore, it was hypothesised that the aberrant artery may be causing a steal like syndrome at the coeliac trunk which could potentially be causing her abdominal pain. However, she was subsequently managed conservatively after a respiratory follow up as firstly her lung function test was reported as sufficient, secondly, she did not encounter any further respiratory infections and finally

her abdominal pain appeared to resolve with the use of regular analgesia.

TEACHING POINT

A hybrid pulmonary sequestration (PMS) and congenital cystic adenomatoid malformation (CCAM) is a bronchopulmonary malformation that can be proficiently identified on a contrast enhanced CT scan. From this imaging modality the essential radiological description that is required for diagnosis includes the appreciation of an atypical artery supplying an abnormal territory of lung parenchyma that is noted to contain multiple cystic projections alongside a rudimentary bronchial tree.

REFERENCES

1. Pryce DM. Lower accessory pulmonary artery with intralobar sequestration of lung; a report of seven cases. *J. Pathol. Bacteriol.* 1946;58:457–67. PMID: 20283082
2. Pryce DM, Holmes Sellors T, Blair LG. Intralobar sequestration of lung associated with an abnormal pulmonary artery. *Br. J. Surg.* 1947;35:18–29. PMID: 20266070
3. Sade RM, Clouse M, Ellis FH. The Spectrum of Pulmonary Sequestration. *Ann. Thorac. Surg. The Society of Thoracic Surgeons*; 1974;18:644–58. PMID: 4611367
4. Savic B, Birtel FJ, Tholen W, Funke HD, Knoche R. Lung sequestration: report of seven cases and review of 540 published cases. *Thorax.* 1979;34:96–101. PMID: 442005
5. Stocker JT, Madewell JE, Drake RM. Congenital cystic adenomatoid malformation of the lung. Classification and morphologic spectrum. *Hum. Pathol.* 1977;8:155–71. PMID: 856714
6. Cass DL, Crombleholme TM, Howell LJ, Stafford PW, Ruchelli ED, Adzick NS. Cystic lung lesions with systemic arterial blood supply: A hybrid of congenital cystic adenomatoid malformation and bronchopulmonary sequestration. *J. Pediatr. Surg.* 1997;32:986–90. PMID: 9247218
7. Laberge JM, Flageole H, Pugash D, Khalife S, Blair G, Filiatrault D, et al. Outcome of the prenatally diagnosed congenital cystic adenomatoid lung malformation: A Canadian experience. *Fetal Diagn. Ther.* 2001;16:178–86. PMID: 11316935
8. Samuel M BD. Management of antenatally diagnosed pulmonary sequestration associated with congenital cystic adenomatoid malformation. *Thorax.* 1999;54:701. PMID: 10413723
9. Kuechle JB, Pasha S, Sadat K, Tran J, Hatipoglu B, Bui JT, et al. Pulmonary Sequestration With CCAM Type II Appearing as Adrenal Tumor Protruding Through a Bochdalek Hernia. *Int. J. Surg. Pathol.* 2011;19:203–6. PMID: 18701512

10. Care AM, Selvaratnam R, Srirangalingam U, Mclean E, Goulden P. A rare cause of acute chest pain in a young adult Case progression. *Clin. Med. (Northfield. Il)*. 2011;11:265–7. PMID: 21902082
11. Herman TE, Siegel MJ. Congenital intralobar pulmonary sequestration with cystic adenomatoid malformation. *J. Perinatol*. 2009;29:175–7. PMID: 19177049
12. Carsin A, Mely L, Chrestian MA, Devred P, De Lagausie P, Guys JM, et al. Association of three different congenital malformations in a same pulmonary lobe in a 5-year-old girl. *Pediatr. Pulmonol*. 2010;45:832–5. PMID: 20652995
13. Gupta K, Sundaram V, Das A, Kumar P. Extralobar sequestration associated with congenital pulmonary airway malformation (CPAM), type I: an autopsy report. *Fetal Pediatr. Pathol*. 2011;30:167–72. PMID: 21355683
14. Cheney LB, Patel B, Lam A, Arbuckle S, Morris J, Holland AJA. Extralobar pulmonary sequestration in association with congenital cystic adenomatoid malformation: An unusual abdominal mass. *ANZ J. Surg*. 2011;81:556–8. PMID: 22295391
15. Olivieri C, Nanni L, Busato G, Rindi G, Marano R, Pintus C. Intradiaphragmatic hybrid lesion in an infant: Case report. *J. Pediatr. Surg. Elsevier B.V.*; 2012;47:e25–8. PMID: 22901939
16. Mackenzie TC, Guttenberg ME, Nisenbaum HL, Johnson MP, Adzick NS. A fetal lung lesion consisting of bronchogenic cyst, bronchopulmonary sequestration, and congenital cystic adenomatoid malformation: The missing link? *Fetal Diagn. Ther*. 2001;16:193–5. PMID: 11399876
17. Al-Mudaffer M, Brenner C, McDermott M, White M, Wood AE, McMahan CJ. Successful Surgical Resection of Intrapericardial Extralobar Pulmonary Sequestration With Congenital Pulmonary Adenomatoid Malformation Type II. *Ann. Thorac. Surg*. 2006;82:327–9. PMID: 16798246
18. McLean SE, Pfeifer JD, Siegel MJ, Jensen ER, Schuler PM, Hirsch R, et al. Congenital cystic adenomatoid malformation connected to an extralobar pulmonary sequestration in the contralateral chest: Common origin? *J. Pediatr. Surg*. 2004;39:13–7. PMID: 15300558
19. Erden ES, Yetim TD, Balci A, Akcay AB, Hakverdi S, Demirkose M. Intralobar pulmonary sequestration with arterial supply from two different origins: A case report. *Ann. Thorac. Cardiovasc. Surg*. 2012;18:560–3. PMID: 22572225
20. Tashtoush B. Pulmonary Sequestration: A 29 Patient Case Series and Review. *J. Clin. Diagnostic Res*. 2015;10–3. PMID: 26816878
21. Wani SA, Mufti GN, Bhat NA, Baba AA. Case Report Pulmonary Sequestration: Early Diagnosis and Management. Hindawi Publishing Corporation; 2015;2015. PMID: 26273485
22. Tsubouchi H, Matsumoto N, Yanagi S, Ashitani J-I, Nakazato M. Successful treatment of chronic lower respiratory tract infection by macrolide administration in a patient with intralobar pulmonary sequestration and primary ciliary dyskinesia. *Respir. Med. case reports. Elsevier Ltd*; 2015;15:62–5. PMID: 26236606
23. Shibano T, Endo S, Tetsuka K, Saito T, Kanai N, Yashiro T. Aberrant aneurysm of adult-type pulmonary sequestration: report of a case. *J Thorac Dis*. 2015;7:E33-6. PMID: 25922746
24. Gürsu AH, Boyvat F, Varan B, Erdoğan I. Embolization of pulmonary sequestration with Onyx: An unusual application. *Turk Kardiyol. Dern. Ars*. 2014;42:174–7. PMID: 24643150
25. Yoshitake S, Hayashi H, Osada H, Kawahara M. Emergency laparotomy helped the resection of an intralobar pulmonary sequestration with haemorrhagic shock. *Eur. J. Cardiothorac. Surg*. 2013;43:190–2. PMID: 22764148
26. Ayloo SM, Addeo P, Buchs NC, Giulianotti PC. Intralobar pulmonary sequestration with an aberrant artery arising from the celiac trunk. *Eur. J. Cardio-thoracic Surg*. 2011;39:418. PMID: 20667746
27. Kim WH, Jeong SH, Ha KW, Lee WS, Kim DC, Chon GR. Bronchopulmonary Sequestration with Dual Arterial Supply from Celiac Artery and Thoracic Aorta. *Tuberc. Respir. Dis. (Seoul)*. 2010;68:101.
28. Izzillo R, El Hajjam M, Qanadli S, Guichaux F, Mignon F, Caubarrere I, et al. [Intralobar sequestration of the lung in an adult (type 1 of pryce). Treatment by coil embolization]. *J. Radiol*. 2000;81:996–9. PMID: 10992102
29. Ahn SJ, Kim EY, Kim JH, Byun SS, Kim HS, Choi HY, et al. Successful endovascular treatment of bilateral intralobar pulmonary sequestration with a bridging isthmus in a child. *Pediatr. Pulmonol*. 2014;49:126–9. PMID: 24500896
30. Karakas SP, Morrison S, Soldes O. Unilateral Coexisting Intralobar and Extralobar Pulmonary Sequestrations. *Radiol. Case Reports. The Authors.*; 2008;3:171. PMID: 27303537
31. Gabriele OF. Arterial supply to the lung via the celiac axis. *American Journal of Roentgenology*. 1970 Jul;109(3):522-7.
32. Lee ML, Lue HC, Chiu IS, Chiu HY, Tsao LY, Cheng CY, et al. A systematic classification of the congenital bronchopulmonary vascular malformations: Dysmorphogeneses of the primitive foregut system and the primitive aortic arch system. *Yonsei Med. J*. 2008;49:90–102. PMID: 2615272
33. Petersen G, Martin U, Singhal A, Criner GJ. Intralobar sequestration in the middle-aged and elderly adult: Recognition and radiographic evaluation. *J. Thorac. Cardiovasc. Surg*. 2003;126:2086–90. PMID: 14688733
34. Van Langenhove G, Convens C, Seynaeve P, Van Den Heuvel P, Van Den Branden F, Stockman D, et al. Intralobar

pulmonary sequestration supplied by the right coronary artery. *Catheter. Cardiovasc. Interv.* 1999;47:218–20. PMID: 10376510

35. Sotto Mayor J, Rocha D, Esperanca S, Oliveira e Silva A. Intralobar pulmonary sequestration: diagnostic expertise. *BMJ Case Rep.* 2015;2015:bcr2015212384-bcr2015212384. PMID: 26584907

36. Naffaa L, Tank J, Ali S, Ong C. Bronchopulmonary sequestration in a 60 year old man. *J. Radiol. Case Rep.* 2014;8:32–9. PMID: 25426223

37. Sfakianaki AK, Copel J a. Congenital cystic lesions of the lung: congenital cystic adenomatoid malformation and bronchopulmonary sequestration. *Rev. Obstet. Gynecol.* 2012;5:85–93. PMID: 3410507

38. Biederer J, Mirsadraee S, Beer M, Molinari F, Hintze C, Bauman G, et al. MRI of the lung (3/3)-current applications and future perspectives. *Insights Imaging.* 2012;3:373–86. PMID: 3481076

FIGURES

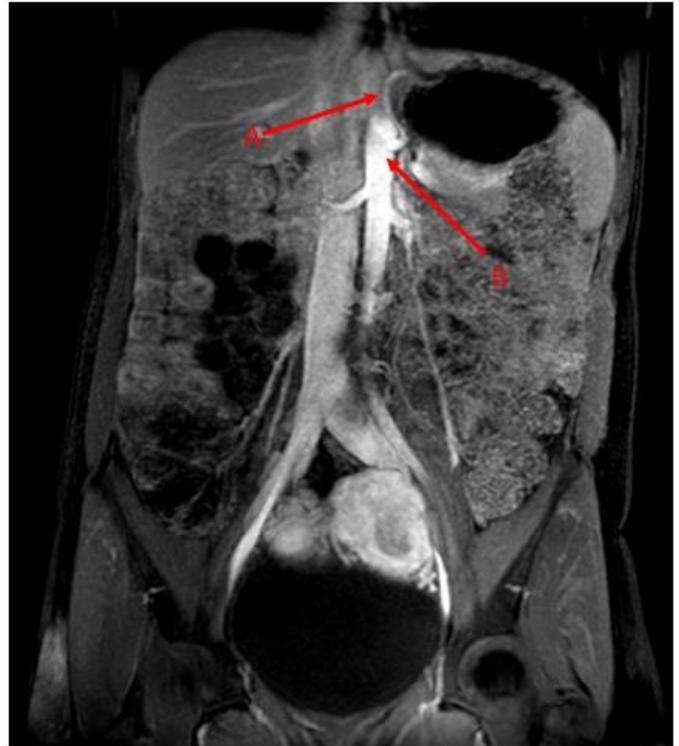


Figure 1: 30-year-old caucasian female with a intralobar pulmonary sequestration displaying features of congenital cystic adenomatoid malformation.

Findings: Post-contrast Magnetic Resonance Image (MRI) presenting the aberrant artery (A). The artery can be observed arising from the coeliac axis (B). It tracks cranially via the oesophageal hiatus and appears to supply the lower lobe of the left lung.

Imaging technique: Coronal T1 High Resolution Isotropic Volume Excitation (THRIVE) MRI performed 60 seconds after 10mls of intravenous gadolinium chelate (Dotarem, Guerbet Laboratories) using a 3 Tesla Philips Achieva unit and the manufacturer's body coil.



Figure 2: 30-year-old caucasian female with a intralobar pulmonary sequestration displaying features of congenital cystic adenomatoid malformation.

Findings: A Computed Tomographic Angiography (CTA) three-dimensional reconstruction demonstrating the origin and course of the aberrant artery (A). The vessel (A) can be seen originating from the celiac trunk (B) and passing cranially through the oesophageal hiatus. The beginning of a bifurcation may be observed at the distal end of the artery (A) visible in the above image. Imaging technique: Axial Computed Tomography (CT) (Toshiba Aquilion One Vision), variable mAs, 100kVp, 0.5mm slice thickness, 1mm reconstruction interval, Adaptive Iterative Dose Reduction 3D reconstruction performed 30 seconds following 100mls of intravenous Omnipaque 300 (GE healthcare). Images reconstructed using Agfa Impax Volume Viewing application)

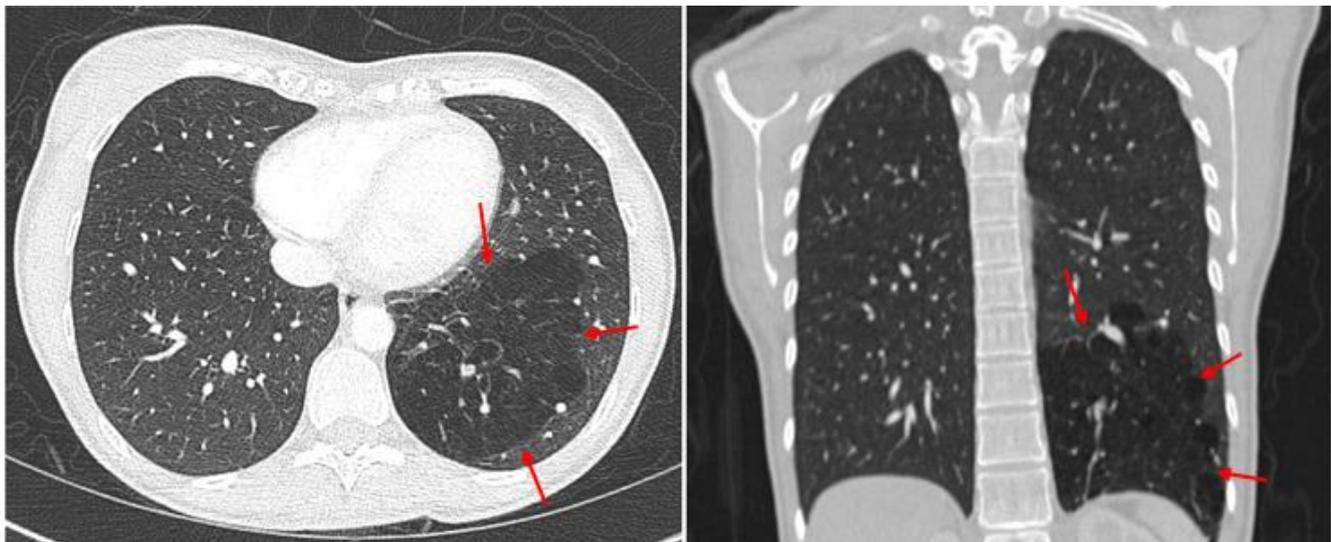


Figure 3: 30-year-old caucasian female with a intralobar pulmonary sequestration displaying features of congenital cystic adenomatoid malformation.

Findings: CT images in the axial (A) and coronal planes (B) illustrating the lung segment which the aberrant artery is supplying. The margins of the malformation can be appreciated as highlighted by the red arrows. Approximately half of the left lower lobe is involved. A rudimentary bronchial tree is also present which does not appear to communicate with the main tracheobronchial system. There are also several small cysts which can be observed within the irregular lung segment. These findings would suggest a intralobar pulmonary sequestration with cystic features of a CCAM.

Imaging technique: Axial and coronal CT (Toshiba Aquilion One Vision), variable mAs, 100kVp, 0.5mm slice thickness, performed following 100mls of intravenous Omnipaque 300 (GE healthcare).

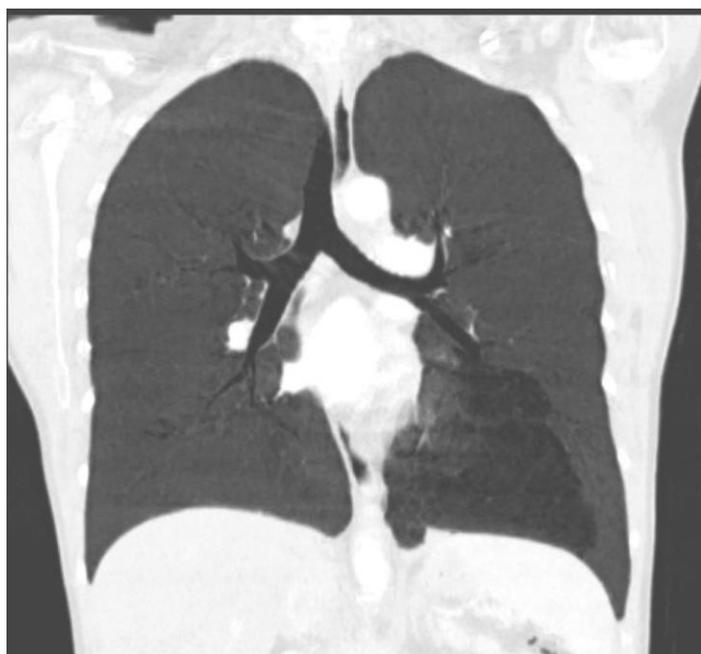


Figure 4 (left): 30-year-old caucasian female with a intralobar pulmonary sequestration displaying features of congenital cystic adenomatoid malformation.

Findings: The bronchial tree system can be observed for both the right and left lung. Within the segment of lung parenchyma that the aberrant artery is supplying a rudimentary bronchial tree is observed which does not seem to communicate with the main tracheobronchial system. These findings, along with the cystic features described in the lung segment are highly descriptive of a intralobar pulmonary sequestration with features of a CCAM.

Imaging technique: Minimal Intensity Projection CT images in the coronal plane, image acquired using CT (Toshiba Aquilion One Vision), variable mAs, 100kVp, 10 mm slice thickness, performed following 100mls of intravenous Omnipaque 300 (GE healthcare).

Etiology	Theory: genetic mutation or insult leading to a fault in fetal lung and vasculature development
Incidence	0.15% to 1.7%
Gender ratio	50:50, however, with increased age there is a preference towards the male gender for presentation
Age predilection	Incidental finding present from birth
Risk factors	Unknown
Treatment	Most people who are symptomatic will require a lobectomy. However, in those who are asymptomatic conservative management may be preferred.
Prognosis	Good with appropriate management

Table 1: Summary table for pulmonary sequestration.

Etiology	Theory: genetic mutation or insult leading to a fault in fetal lung development
Incidence	1 in 25,000 to 1 in 35,000 pregnancies
Gender ratio	No reported gender preference
Age predilection	Present from birth
Risk factors	Unknown
Treatment	Surgical resection is definitive treatment; however, conservative management may be sought if the patient is asymptomatic
Prognosis	Good with appropriate management

Table 2: Summary table for congenital cystic adenomatoid malformation (CCAM).

	<u>X-ray findings</u>	<u>CT findings</u>	<u>MRI findings</u>
Pneumonia	<ul style="list-style-type: none"> • Homogenous opacification in a lobar pattern 	<ul style="list-style-type: none"> • Focal ground-glass opacity in a lobar or segmental pattern 	<ul style="list-style-type: none"> • Not routinely used • On T2 Weighted (T2W) images a high signal is observed from the infected lung tissue
Lung abscess	<ul style="list-style-type: none"> • Cavity containing an air-fluid level 	<ul style="list-style-type: none"> • Cavity that has a variable size and shape. • May contain fluid or have an air-fluid level • Can have a thick wall and irregular luminal surface 	<ul style="list-style-type: none"> • Not routinely used
Bronchiectasis	<ul style="list-style-type: none"> • Tram-track opacities • Increased bronchovascular markings • Terminal bronchi can appear as ring shadows • Pulmonary vasculature can look ill-defined 	<ul style="list-style-type: none"> • Classical signs include the tram-track sign and signet ring sign. • A honeycomb appearance is also observed that represents large cystic areas. 	<ul style="list-style-type: none"> • High signal from bronchial wall or bronchial lumen on T2W. • Enhanced bronchial wall on fat suppressed T1 Weighted (T1W)
Empyema	<ul style="list-style-type: none"> • Unilateral/asymmetric pleural fluid with a biconvex shape • Loculated effusion may also be seen 	<ul style="list-style-type: none"> • Fluid accumulation observed in the pleural space • Split pleura sign: thickening of the parietal and visceral pleura which are observed separated due to collection of pleural fluid 	<ul style="list-style-type: none"> • Not routinely used
Bronchogenic cysts	<ul style="list-style-type: none"> • Appear as well circumscribed zones of increased opacity that are usually round but variable in size 	<ul style="list-style-type: none"> • Well demarcated spherical or tubule masses of homogenous or variable attenuation 	<ul style="list-style-type: none"> • T1W displays lung lesions with variable signal intensity • T2W display high signal intensity lung lesions
Pulmonary neoplasm	<ul style="list-style-type: none"> • No specific findings. • Hilar or perihilar opacification with mediastinal widening. • Central or peripheral nodules/areas of increased opacification 	<ul style="list-style-type: none"> • Enlarged lymph nodes • Infiltration and disruption in the anatomy of adjacent structures • Atelectasis • Cavitation • Hemorrhage of lung tissue • Necrosis of lung tissue 	<ul style="list-style-type: none"> • Not routinely used
Pulmonary sequestration (PS)	<ul style="list-style-type: none"> • Increased areas of opacification observed in the affected segments of lung • Air-filled lesions can be identified with the presence of multiple cysts 	<ul style="list-style-type: none"> • Intralobar and extralobar pulmonary sequestration (PS) can be identified via <ul style="list-style-type: none"> ○ Identification of a rudimentary bronchial tree ○ Observation of an analogous blood supply 	<ul style="list-style-type: none"> • T1W and T2W produce high signal against the sequestered segment relative to the normal lung tissue. • Contrast enhanced Magnetic Resonance Angiography shows the blood supply to PS
Cystic Adenomatoid Malformation (CCAM)	<ul style="list-style-type: none"> • Multicystic lesions: <ul style="list-style-type: none"> ○ Air-filled ○ Variable sized • Lesion may appear with an air-fluid level or appear solid • Presentence of mediastinal shift or depression 	<ul style="list-style-type: none"> • Small to large cysts can be identified <ul style="list-style-type: none"> ○ Circumferential areas of low attenuation ○ Air – fluid level may also be present • Zone of consolidation. • Presence of mediastinal shift 	<ul style="list-style-type: none"> • Not routinely used

Table 3: Differential diagnosis table for bronchopulmonary malformations.

ABBREVIATIONS

CCAM = Congenital Cystic Adenomatoid Malformation
CT = Computed Tomography
EPS = Extralobar Pulmonary Sequestration
FODMAP = Fermentable Oligosaccharides, Disaccharides, Monosaccharides and Polyols
IPS = Intralobar Pulmonary Sequestration
MRI = Magnetic Resonance Imaging
PMS = Pulmonary Sequestration

KEYWORDS

bronchopulmonary foregut malformation; pulmonary sequestration; congenital cystic adenomatoid malformation; intralobar pulmonary sequestration; extralobar pulmonary sequestration

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