Pulmonary Sequestration; a Case Series Analysis

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Abstract:	Background: Pulmonary sequestration is a congenital pulmonary anomaly which rarely occurs with an approximate incidence of 1 per 8300 to 35,000 of the population. It has variable presentation ranging from asymptomatic to recurrent lung infection and hemoptysis. Objective: The aim of this study is to look at the variable presentation of the disease, both clinically and intraoperatively. Methods: We conducted a retrospective case series analysis of lung sequestration from 1st January 2019 to 31th May 2024 in Pusat Jantung Sarawak, Malaysia. Results: 7 patients were included in our study. 3 patients were male and 4 were female. Age range was between 16 years to 42 years old. 4 patients presented with haemoptysis; 2 patient had infective symptoms and 1 patient was asymptomatic. All patients had aberrant arterial supply from descending aorta and the drainage was variable. All of the patients underwent lobectomy. Conclusion: Pulmonary sequestration is a rare congenital anomaly which has variable presentation. Preoperatively planning including imaging is paramount and surgery remain the mainstay of treatment .

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Pulmonary Sequestration; a Case Series Analysis.

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Abstract

Background: Pulmonary sequestration is a congenital pulmonary anomaly which rarely occurs with an approximate incidence of 1 per 8300 to 35,000 of the population. It has variable presentation ranging from asymptomatic to recurrent lung infection and hemoptysis.
Objective: The aim of this study is to look at the variable presentation of the disease, both clinically and intraoperatively.
Methods: We conducted a retrospective case series analysis of lung sequestration from 1st January 2019 to 31th May 2024 in Pusat Jantung Sarawak, Malaysia.
Results: 7 patients were included in our study. 3 patients were male and 4 were female. Age range was between 16 years to 42 years old. 4 patients presented with haemoptysis; 2 patient had infective symptoms and 1 patient was asymptomatic. All patients had aberrant arterial supply from descending aorta and the drainage was variable. All of the patients underwent lobectomy.
Conclusion: Pulmonary sequestration is a rare congenital anomaly which has variable presentation. Preoperatively planning including imaging is paramount and surgery remain the mainstay of treatment .

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Ethical Approval/Informed Consent

Ethical approval was not sought for the present study because no identifiable images or information were used.

Funding and Conflict of Interest

The authors received no financial support for this research. There is no conflict of interest.

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None to disclose.

Introduction

Pulmonary sequestration which is also known as bronchopulmonary sequestration is a rare congenital anomaly which consists of a non- functioning lung mass which does not communicate with the tracheobronchial tree (1). Various subtypes exist; however, the main ones are intralobar sequestration and extralobar sequestration; intralobar sequestration (ILS) share the common pleura with the normal lung tissue while extralobar sequestrations (ELS) are separated from the remaining lung tissue by a separate lining of pleura(2). It is important to note that ILS and ELS can be present simultaneously (3),(4). The spectrum of the sequestration includes anomalies of the systemic blood supply and venous drainage. This case series will discuss the various origin of the blood supply.

Methods

We review patients who were diagnosed and treated for pulmonary sequestration from 1st January 2019 to 30th May 2024 in Pusat Jantung Sarawak, Malaysia. The study was done retrospectively; we searched the hospital's medical database containing any mention of pulmonary sequestration. Clinical information was retrospectively gathered from case notes, surgical notes and imaging report. 3-dimension image of was reconstructed using Bee DICOM Viewer. Our analysis include symptoms or presentation, age, origin of blood supply, venous drainage, localization and operative technique.

Ethical Approval

Ethical approval and informed consent was not required for the purpose of this study.

Results

The summary of our results are shown in the table 1. We found 7 patients meeting the criteria. There were 3 males and 4 females. During the surgery, the patients age were between 16 years old to 42 years old, and the average was 27.2 years.

4 out of 7 presented with haemoptysis, 2 patients presented with infective symptoms and 1 patient was asymptomatic which was picked up incidentally during TB screening. Both patients with infective symptoms was treated with prolonged antibiotics. In our centre, Computer Tomography (CT scan) remain the imaging of choice. All of the patients underwent CT scan for confirmation of diagnosis. 6 out of 7 of the patients has ILS of the left lower lobe; only 1 patient has ILS of the right lower lobe. This was diagnosed in the CT scan preoperatively and confirmed intraoperatively. Subsequently, all of the ILS was again confirmed with histopathological examination findings.

Video assisted thoracoscopic surgery (VATS) approach was used in 5 cases and posterolateral thoracotomy approach was used for the other 2 cases. All of the cases were treated with lobectomy. During the surgery, all of the feeding vessels are identified; they were ligated either using energy device, ligating clip or Endo GIA stapler.

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The aberrant blood supply came from descending aorta in all of the cases. Venous drainage was more variable; venous drainage via inferior pulmonary vein was seen in 6 cases and 1 case via the hemiazygous.

No	Gender	Age (Years)	Type	Blood Supply	Venous Drainage	Localization	Operative Technique	Presentation/Symptoms
1	Female	42	ILS	DA	Left IPV	Left Lower Lobe	VATS + left lower lobectomy	Hemoptysis
2	Female	34	ILS	DA	Left IPV	Left Lower Lobe	VATS + left lower lobectomy	Hemoptysis
3	Male	16	ILS	DA	Left IPV	Right Lower Lobe	VATS + right lower lobectomy	Pneumonia
4	Male	27	ILS	DA	Left IPV	Left Lower Lobe	VATS + left lower lobectomy	Lung abscess
5	Female	21	ILS	DA	Left IPV	Left Lower Lobe	VATS + left lower lobectomy	Hemoptysis
6	Male	30	ILS	x2 DA	Left IPV	Left Lower Lobe	Left Thoractomy + Left Lower Lobectomy	Hemoptysis
7	Female	21	ILS	DA	HemiAzygous	Left Lower Lobe	Left Thoractomy + Left Lower Lobectomy	Asymptomatic/Incidental

Table 1; ILS: intralobar sequestration, DA: Descending Aorta, IPV: Inferior Pulmonary Vein

3D reconstruction are shown with the arrow pointing to the feeding vessel. Case 1 CT scan was not available.

Discussion

Pulmonary sequestration (PS) was first described by Huber in 1777; as an aberrant blood supply to lung. It is defined as a functionless lung parenchyma without connection or with abnormal connection to the tracheobronchial system and has an abnormal blood supply (5). PS is a rare congenital disease in the spectrum of the bronchopulmonary foregut malformation. PS comprises between 0.1% to 6.6% of all congenital lung malformations [(6),(7),(5),(8)]. The incidence is 1 per 8300 to 35,000 of the population (6). ILS is more common than ELS which makes up 75% of all PS cases; roughly in a 3:1 ratio (9).

Despite its rarity and low incidence rate, PS is the second most common congenital lung anomaly (8). However, the low incidence rates are likely reflecting underestimation of the overall number of affected individuals. Small lesions may remain asymptomatic; therefore, these cases often would not present for medical attention and thus remains undiagnosed (10). The quality of prenatal imaging also plays a fundamental role and is therefore likely to under-detect anomalies (11). ILS are more common than ELS; although ILS may present at any age, unless detected during an antenatal ultrasound, they rarely cause problems before the age of 2 years old as opposed to ELS where patients typically present in their first 6 months of life (8).

PS is considered to be a childhood disease and most cases are diagnosed in the early life (6) and some patients may continue their lives normally asymptomatically or with only mild symptoms (6),(10).

The pathogenesis of PS is poorly understood and has been the subject of great debate. Proposed concepts on it were divided into five groups; vascular traction, vascular insufficiency, co-incidental occurrence, acquired pathology following infection and a common development theory (8). However, the most widely accepted embryological theory is that PS originates early in the pseudo-glandular stage of lung development (around 5 to 17 weeks of gestation) prior to separation of the aortic and pulmonary circulations (1). It is suggested that PS results from the formation of an accessory lung bud caudal to the normal lung buds (8).

Patient with PS commonly presents with nonspecific symptoms; recurrent pulmonary infections such as pneumonia and lung abscesses (5). Symptoms include chest pain, shortness of breath, fever and hemoptysis (6).

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As mentioned previously, patients may be asymptomatic while some may present with incidental finding of PS. This suggest that there is a role for antenatal ultrasound in early diagnosis of PS (9). Although, the limitations of anternatal ultrasound should be taken into consideration such as under-detection risks and the technical skills of different sonographers.

For patients presented with symptoms such as recurrent chest infections with or without respiratory distress or congestive cardiac failure without an obvious cause, clinicians need to have a high index of suspicion and investigate appropriately with at least an initial chest X-ray (CXR)(8). CXR findings in PS is usually nonspecific which may mimic a lobar consolidation or appear as a soft tissue mass/nodule, or as a mono-cystic or multi-cystic mass (9). In early childhood, ultrasound is recommended as the first diagnostic modality for evaluating a subphrenic mass, which may also delineate the arterial and venous blood supply to the PS (8).

Computed Tomography scan (CT scan) best demonstrates the parenchymal abnormalities associated with PS (12). CT scan will show the complex mass with or without cystic changes which is the typical appearance of PS. It is paramount to identify the anomalous blood supply, as this is crucial to establishing the diagnosis of PS (9). Traditionally, the diagnosis of lung sequestration has been made by arterial angiography (6). Advances in imaging modalities now allows for noninvasive techniques such as CT scan or MR angiography to be widely utilized to demonstrate aberrant vessels; with CT scan providing evaluation of aberrant systemic artery in 80% of the cases (6). CT scan with intravenous contrast and preferably CT angiography (CTA) is the imaging of choice for identifying the arterial supply (9).

Preoperatively, the anomalous blood supply has to be identified; failure to do so can have devastating consequences if the anomalous blood supply is not ligated at the time of surgical resection (3). PS may have a single feeding vessel and occasionally has multiple systemic supplying arteries. Most common arterial blood supply originates from the thoracic aorta (46.1%-86.1%) and abdominal aorta (6.9%-31.9%) (9). Other sites include intercostal artery, diaphragmatic artery, aortic arch, subclavian artery, pulmonary artery, left gastric artery, coronary artery, celiac trunk and renal artery (9). In terms of its venous drainage, more than 95% of PS cases drains to the pulmonary vein (2). Other venous drainage includes azygous vein, hemiazygous vein, inferior venacava or the right atrium (9).

The other reason that the aberrant blood supply needs to be identified is to rule out other differential diagnosis. Patients that present with pulmonary mass will raise the suspicion of lung malignancy. The importance of delineation of aberrant blood supply may assist to confirm the diagnosis of sequestration (6).

In our centre, CT scan is mandatory to delineate the feeding vessel. The arterial feeding supply was emphasized preoperatively. Intraoperatively, this was carefully ligated to prevent injury which may cause devastating consequences such extensive haemorrhage and significant blood loss.

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Mainstay of treatment is surgical resection for both ILS and ELS. Resection of ILS is almost always involving a lobectomy (8). It is suggested that in asymptomatic, incidentally diagnosed ILS, surgical resection is also indicated due to possible infectious complications and occult fungal infection [(7),(13)]

The management of ELS is more controversial. ELS can remain asymptomatic throughout the patients' life, thus strict observation is required (9). In the ELS patient, definitive surgery is required when symptoms develop (9). For both lesions, the identification and control of the anomalous vascular supply is crucial (8).

Open thoracotomy has been recommended for safe isolation and division of the anomalous vascular supply (1). However, in recent years, the use of video thorascopic surgery (VATS) lung resection has gained more usage (7); although this approach should be only performed by experienced surgeons and in situations where minimal adhesions are expected (7),(14). Performing VATS pulmonary sequestration resection is associated with low morbidity and mortality (1).

Conclusion

Pulmonary sequestration is a rare congenital anomaly which can present asymptomatically and incidentally or with recurrent lung infections such as pneumonia or lung abscesses. It is important for clinicians to be aware of this especially when managing such patients. Upon suspicion or diagnosis, it's paramount to identify the systemic arterial feeding vessel pre-operatively using a CT scan. Surgery remains the mainstay of treatment especially in symptomatic PS patients.

Ethical Approval/Informed Consent

Ethical approval was not sought for the present study because no identifiable images or information were used

Funding and Conflict of Interest

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None to disclose.

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References:

1. Sadasivan Nair P, Merry C, White A. Intralobar pulmonary sequestration in an adult: a case report. J Cardiothorac Surg. 2023 Dec 1;18(1).
2. Savic B, Birtel FJ, Tholen W, Funke HD, Knoche R. Lung sequestration: report of seven cases and review of 540 published cases [Internet]. Vol. 34, Thorax. 1979. Available from: http://thorax.bmj.com/
3. Mathew S, Erozan YS. DIAGNOSTIC DILEMMAS Pulmonary Sequestration-A Diagnostic Pitfall: A Case Report. Vol. 16, Diagn. Cytopathol. Wiley-Liss, Inc; 1997.
4. Sade RM, Clouse M, Ellis FH. The Spectrum of Pulmonary Sequestration. Vol. 18, Annals of Thoracic Surgery. 1974. p. 644–58.
5. Gompelmann D, Eberhardt R, Heußel CP, Hoffmann H, Dienemann H, Schuhmann M, et al. Lung sequestration: A rare cause for pulmonary symptoms in adulthood. Respiration. 2011 Oct;82(5):445–50.
6. Petty L, Joseph A, Sanchez J. Case report: Pulmonary sequestration in an adult. Radiol Case Rep. 2018 Feb 1;13(1):21–3.
7. Polaczek M, Baranska I, Szolkowska M, Zych J, Rudzinski P, Szopinski J, et al. Clinical presentation and characteristics of 25 adult cases of pulmonary sequestration. J Thorac Dis. 2017 Mar 1;9(3):762–7.
8. Corbett HJ, Humphrey GME. Pulmonary sequestration. Vol. 5, Paediatric Respiratory Reviews. W.B. Saunders Ltd; 2004. p. 59–68.
9. Qian X, Sun Y, Liu D, Wu X, Wang Z, Tang Y. Case Report Pulmonary sequestration: a case report and literature review [Internet]. Vol. 8, Int J Clin Exp Med. 2015. Available from: www.ijcem.com/
10. Carter R. Pulmonary Sequestration. Vol. 7, Annals of Thoracic Surgery. 1969. p. 68–88.
11. Clements BS, Warner JO. Pulmonary sequestration and related congenital bronchopulmonary-vascular malformations: nomenclature and classification based on anatomical and embryological considerations [Internet]. Vol. 42, Thorax. 1987. Available from: http://thorax.bmj.com/
12. Bronchopulmonary Sequestration: CT Assessment' 375 Thoracic Radiology.
13. Berna P, Lebied ED, Assouad J, Foucault C, Danel C, Riquet M. Pulmonary sequestration and aspergillosis. Vol. 27, European Journal of Cardio-thoracic Surgery. 2005. p. 28–31.
14. Lin CH, Chuang CY, Hsia JY, Lee MC, Shai SE, Yang SS, et al. Pulmonary sequestration-differences in diagnosis and treatment in a single institution. Journal of the Chinese Medical Association. 2013 Jul;76(7):385–9.

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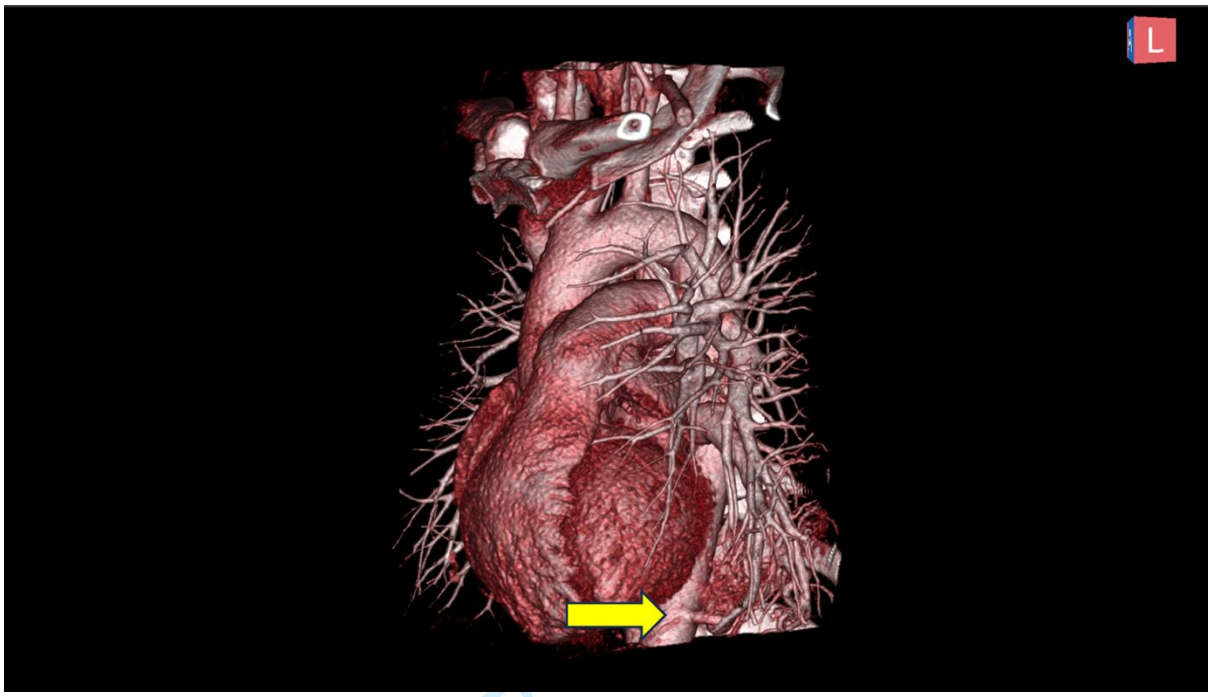
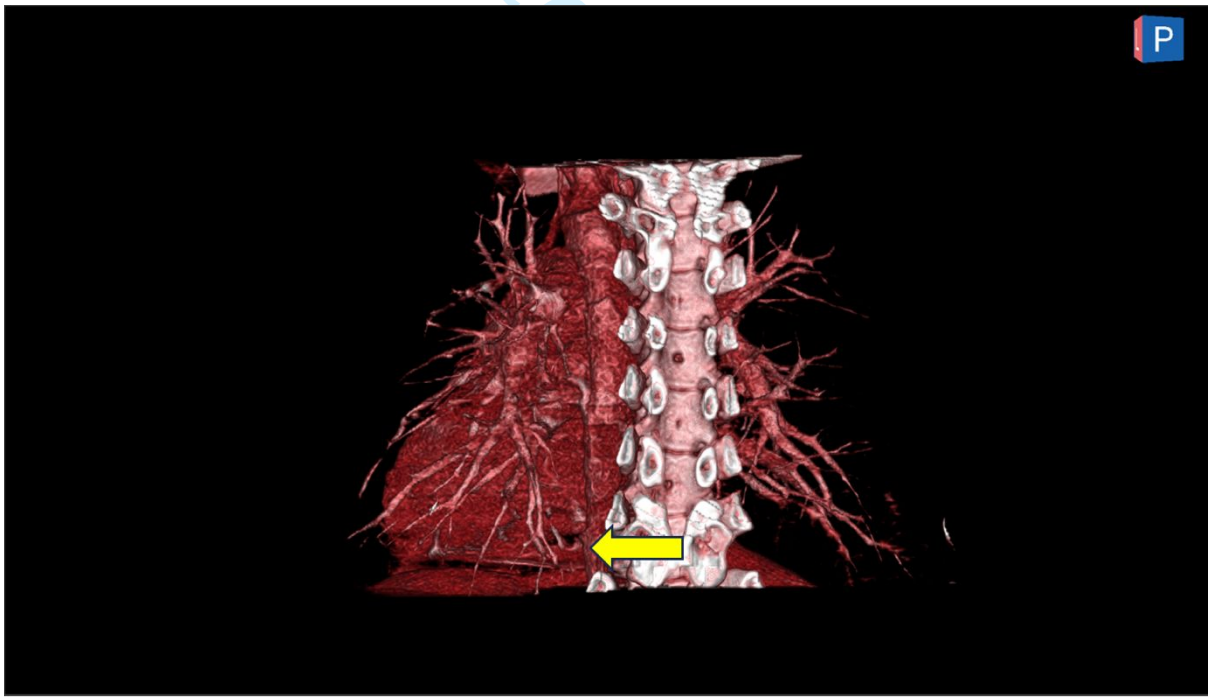


Case 2; single arterial supply from DA at level of T10



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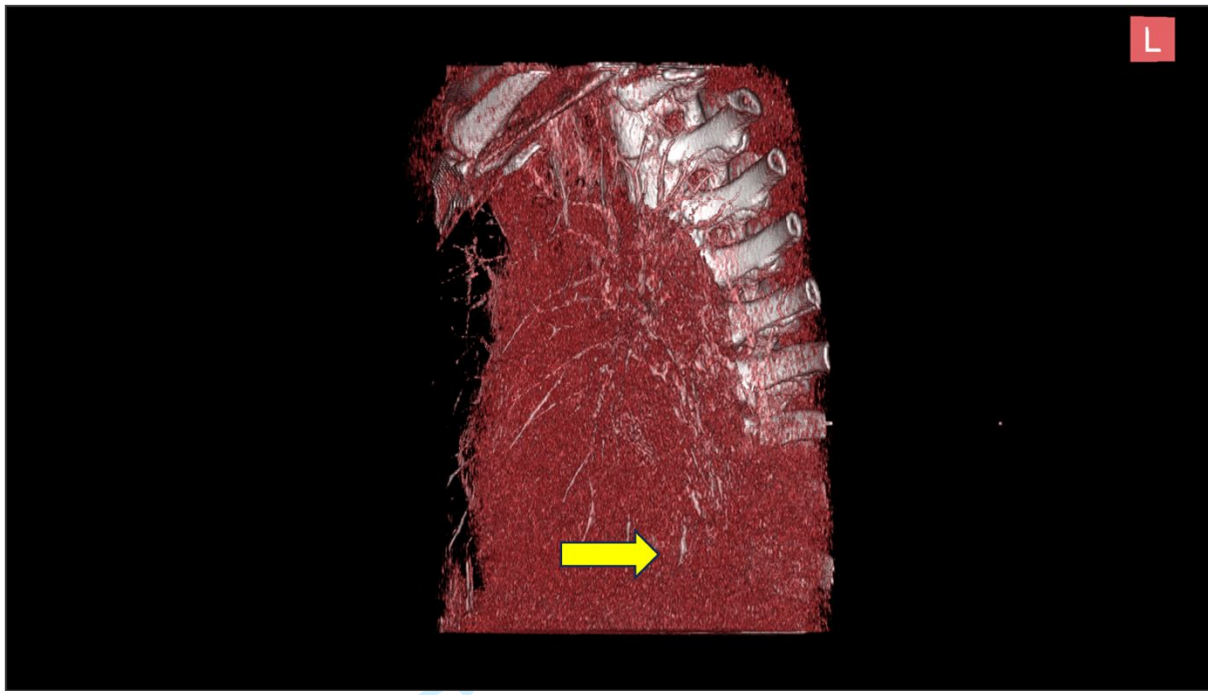

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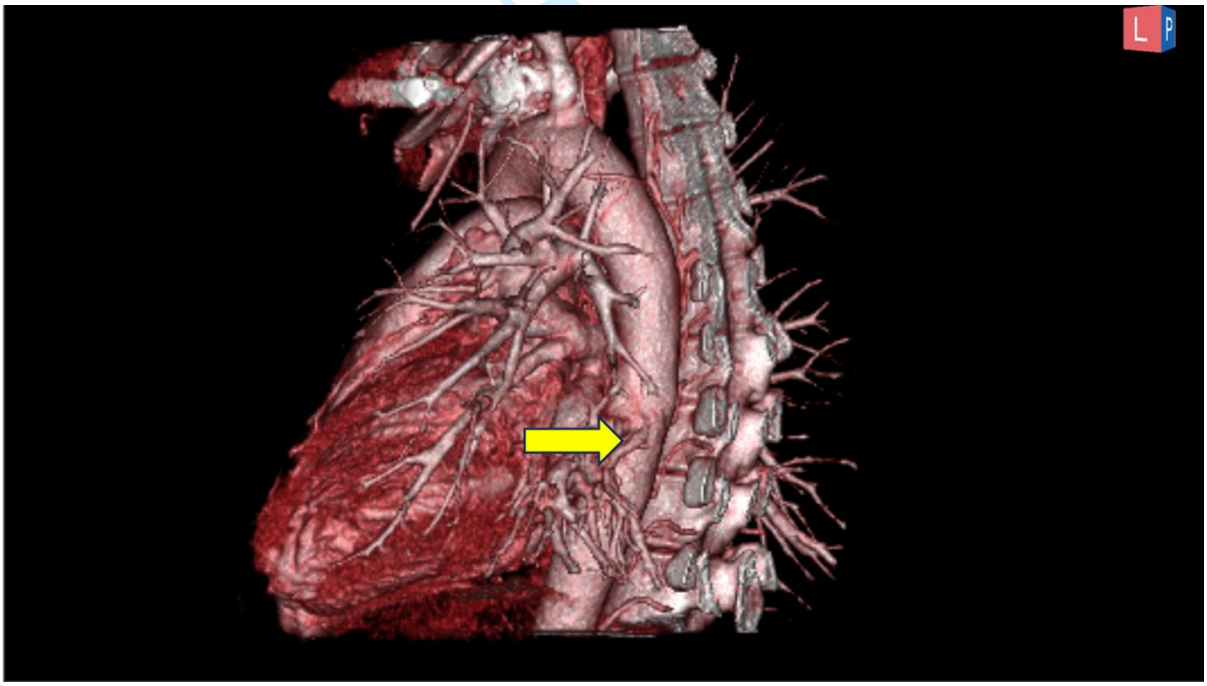

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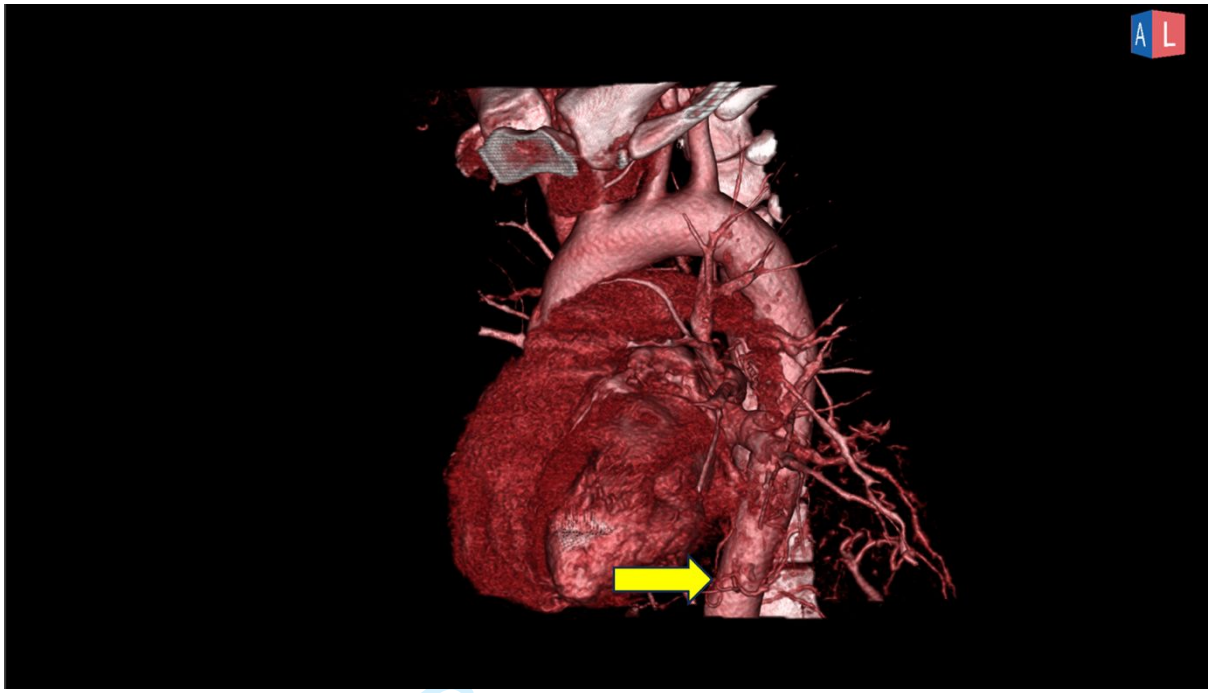
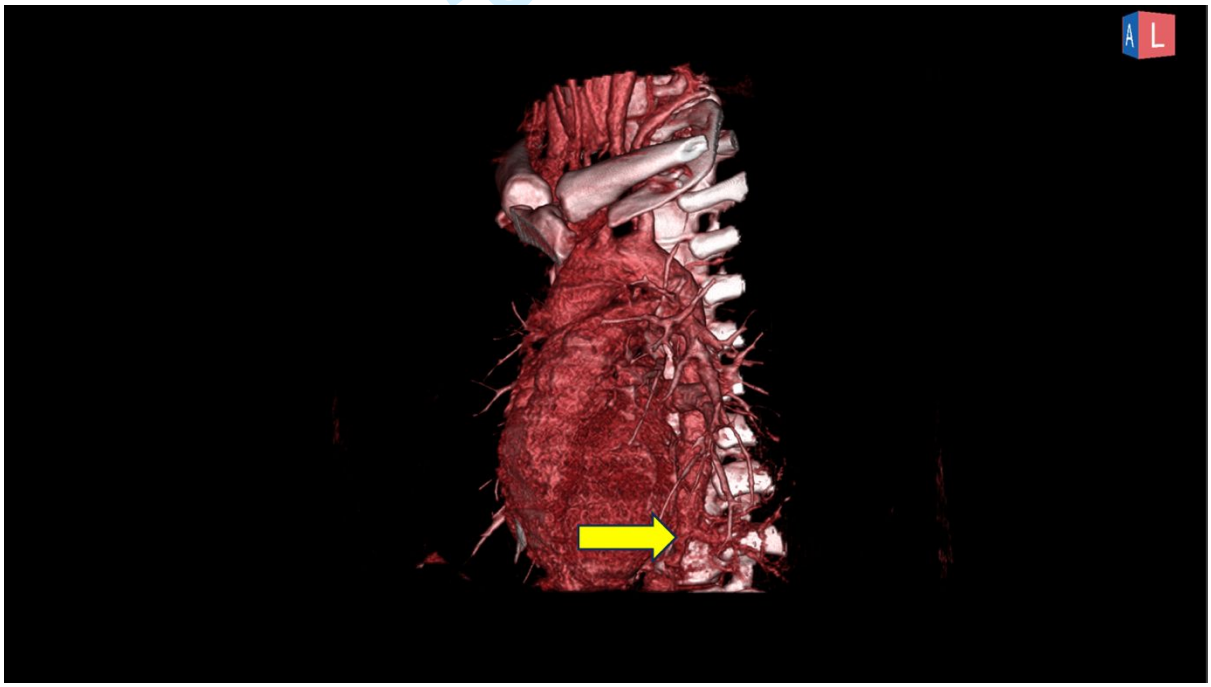


Case 6; two small feeder from DA at level of T9



Case 7; single arterial supply from DA at level of T9