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Rare Case of Pulmonary Sequestration

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Abstract

Pulmonary sequestration is a rare congenital lung malformation resulting in a mass of dysplastic lung tissue supplied by aberrant systemic vessel and separated from the normal bronchopulmonary tree, commonly misdiagnosed, and not treated in a timely manner, patient often exposed to unnecessary investigations and invasive procedures before diagnosis been made. We present one of our patients who was misdiagnosed and had two surgeries before diagnosis was made and successfully treated.

Case report

30-year-old Sudanese patient who immigrated to Australia before10 years presented to ED with cough, chest pain and Haemoptysis, on arrival to hospital, he was febrile at 37.8, normal heart rate, blood pressure and oxygen saturation, cardiac examination showed dual heart sounds and no murmurs, examination of the chest did not show any clinical signs of consolidation, patient used to smoke for 15 years but stopped smoking for the last 5 years, he did not have any comorbidity and not on regular medications, CXR was normal, CT chest showed a cystic lesion in the left lower zone.

Full blood count, biochemistry and blood culture was unremarkable apart from CRP was 30 mg/Dl(I mg/Dl), patient diagnosed as pneumonia and discharged on one week of antibiotic, one month later patient admitted with symptoms of pneumonia and haemoptysis, CXR and CT did not show any new changes, bronchoscopy and bronchoalveolar lavage were normal, patient was reassured and refereed to tuberculosis Clinic because of his African descent, patient received BCG in his past, his Mantoux test was mildly positive consistent with past vaccination, induced sputum was examined for Acid fast bacilli, culture for TB and deaminase, patient scheduled to be reviewed again in 6 weeks time, four weeks later, patient was admitted because of dry cough, fever and haemoptysis, D dimer was positive, CTPA ruled out pulmonary embolism, Coagulation screen was normal, induced sputum did not confirm tuberculous infection, chest phycision, decided to treat him empirically as pulmonary TB because of his country of origin, patient was started on Rifamycin, INH, ethambutol, pyranezemide and pyridoxine for two months followed by rifamycin and INH for four months, two month later patient was readmitted with haemoptysis, repeated CT chest did not change, patient was discussed by a multidisciplinary team, they agreed that small cystic mass in the left lower lobe should be resected, patient informed with decision of the multidisciplinary team and agreed, mass was resected, pathological examination was nonspecific, mass did not show any features of malignancy,

biopsy was examined for culture cytology, histochemical staining immunohistochemical staining, electron microscopy, flow cytometry, genetics, cytogenetics, fluorescent in situ hybridization, molecular genetics, PCR, gene expression microarrays, DNA sequencing and microbiology testing.

Patient was discharged home, to be seen in the clinic in 6 weeks for discussion about result of lung biopsy and other tests, patient was feeling well for two months, all biopsy testing were negative for cancer or any specific pathology, patient was told that he is cured, and asked to stop taking any treatment for TB.

Three months later patient was readmitted with fever and haemoptysis, repeated blood testing was unremarkable, CTPA and CTA ruled out pulmonary embolism but showed a very small cystic mass less than 1cm in the left lower lobe with a small feeding vessel arising from the descending aorta into the small cystic mass, diagnosis had been made as a intralobular pulmonary sequestration.

Patient was treated with endovascular therapy for the aberrant artery and resection of the diseased mass, patient felt better with no recurrence of the disease, reviewed in 6 months' time at the outpatient clinic where he was asymptomatic with normal clinical examination and normal CTA.

Patient considered cured and discharged from the clinic.

Discussion

Lung sequestration is a very rare disease which manifested by very common manifestations like pneumonia, haemoptysis and occasionally heart failure, patients can present to different subspeciality including GP, ED, chest physician, cardiologist and occasionally gastroenterologist and haematologist for chronic anaemia, and occasionally oncologist for a lung mass and rarely multidisciplinary team for investigating hypoxia, most patients will not be diagnosed from the first presentation due to the rarity of the disease, almost all patients will undergo repeated invasive investigations like bronchoscopy, lung biopsy and some patients will have unnecessary surgeries like lung resection, few patients would undergo Video- assisted thoracoscopic surgery.

Pulmonary sequestration represents up to 6% of congenital pulmonary malformations, characterised by dysplastic lung tissue supplied by aberrant systemic artery, the abnormal segment of the lung is disconnected from the tracheobronchial tree, this anomaly is explained in the most literatures by.

Formation of accessary lung bud inferior to the normal lung bud during embryological formation and development and getting the blood supply from the foregut vessels, the abnormal segment of the lung and the rest of the lung will have the same pleura if the accessary lung bud develops before the formation of pleura, this sequestration is called interlobar sequestration, it is the most common type of lung sequestration and commonly give rise to recurrent illness and needs treatment.

Most of the interlobar pulmonary sequestration occur in the medial and posterior basel segment of the left lung, overall 96% occur in the lower lobes, bilateral involvement is rare, associated congenital abnormalities included diaphragmatic hernia, congenital valvular disease, pectus excavatum, bronchogenic cyst, congenital cystic adenoid malformation are very rare in interlobar sequestration than extra lobar sequestration, systemic vascular blood supply in interlobar sequestrations is from descending abdominal aorta in 75% of cases, other 25% included splenic artery, celiac vessel, subclavian artery, internal thoracic, intercostal artery, internal thoracic and pericardiophrenic arteries pulmonary artery, left gastric artery, coronary and renal artery had been reported, almost all venous drainage is via pulmonary veins.

If the accessary lung bud develops after the formation of the pleura, this will result in the sequestrated lung tissue develops its own pleura covering resulting in extra lobar sequestration. Extra lobar sequestration can occur in the chest cavity, diaphragm and occasionally in the abdomen, they are associated with other anomalies like pectus excavitum, congental valvular disease, pericardial cyst, hind gut duplications, diaphragmatic hernia, or pericardial defects.

Extra lobar sequestrations are usually Asymptomatic but occasionally they can cause feeding problems, hypoxia, cardiac or chest symptoms, although it is benign, patients still at risk to develop infection, haemorrhage, or malignant transformation, histologically the extralobar sequestrations is Dilated airways lined by bronchial epithelium and remenants of cartilaginous bronchi and bronchiole, the sequestrated

segment receives blood supply from aberrant systemic circulation.

Although most masses in the posterior mediastinum are neurogenic, extralobar sequestration should be in the differential diagnosis even in the absence of aberrant artery.

Although interalobar sequestration is believed to be congenital, some authors think it could be acquired due to chronic infections causing lung tissue to be disconnected from the normal bronchial tree causing hypertrophy of the nearby systemic artery which become the aberrant feeding vessel, in a large study for patients with pulmonary sequestration, CT showed that 50% of mass lesion were cystic, 30% were cavitatory and the rest were consolidation, in another study 78% of aberrant vessels originated from descending thoracic aorta, and the rest stemmed from one aberrant vessel, Venous return of interalobar sequestration is via the pulmonary vessels in more than 96%, usually symptoms are prominent in the first 10 years of life, sequestrations are not supplied by bronchial artery.

In another study which included 133 extralobar sequestrations, 78% were located between the diaphragm and lower lobe, aberrant supply was from descending thoracic aorta, few cases were supplied from venous branched of pulmonary vessel.

In another study antenatal diagnosis of pulmonary sequestrations was possible by antenatal ultra-sound after week twenty in 10 patients, systemic aberrant feeding vessels was identified in four of them, spontaneous resolution occurred in eight of them, two of them manifested with hemithorax and polyhydramonis which was treated with paracentesis and the amountic fluid allowed the pregnancy to continue until full term.

Bilateral pulmonary sequestration had been reported in the literatures with a bridging Isthmus which was treated surgically after failing to treat endoscopically.

In Conclusion

Pulmonary sequestration is a rare condition manifested by common clinical signs and symptoms in the form of recurrent pneumonia and haemtysis, the accessary lung mass could be cystic or solid.

The diagnosis is usually missed because of the rarity of the condition, red flag is the common involvement of the left posterior basel segment of the lung, incomplete response to medication and repeated presentation, associated congenital malformation are common in extralobar sequestration, guidelines advise that every patient should have CT/MR angiography.

Symptomatic patients should be treated with resection and endovascular therapy, there is no agreement that A symptomatic sequestration should be treated, except if they become symptomatic, every clinician should have a low threshold to consider the diagnosis of pulmonary sequestration in patients with repeated pneumonia and hemoptosis if they did not respond to standard of care management, this will avoid morbidity and mortality from avoidable invasive investigations, procedures and surgery.

References

1. Lin CH, Lin CT, Chen CY, Phen H, Phen HC, et al. (1994) Pulmonary Sequestration. Chinese Medical journal. 53: 168-172.
2. Webb WR. (2006) Fundamentals of body CT. (Third Edition).
3. Sade MR, Clouse M, Ellis H. (1974) the spectrum of pulmonary sequestration. Ann Thorac Surg. 18: 644-658.
4. Halkic N, Cuenoud PF, Corthesy ME, Ksontini R. (1998) Puklmonary sequestration: a review of 26 cases. EUR J Cardiothorac Surg. 14(2): 127-133.
5. Smith RA. (1956) A Theory of the origin on intralobular Sequestration of Lung. Thorax. 11(1): 10.
6. Gupta S, Kim S. (2013) Pulmonary Sequestration. N Engl J Med. 368: e6.
7. Albanese CT and Rothenberg SS. (2007) Experience with 144 consecutive pediateric. Thoracoscopic lobectomies. Journal of laparoendoscopic advanced surgical Techniques. 17(3): 339-341.
8. Yucel O, Gurkok S, Gozubuyuk A, Caylak H, Sapmaz E, et al. (2008) Diagnosis and surgical Treatment of pulmonary Sequestration. 56(3):154-157.
9. Montjoy C, Hadique S, Graeber G, Ghamande S. (2021) Intralobar Bronchopulmonary Sequestra in adults over the age of 50: case series and review. WV Med J. 108(5):

8-13.

10. Szmygin M, Pyra K, Sojka M, Jariello T. (2021) Successful Endovascular treatment of Intralobar pulmonary Sequestration- an effective alternative to Surgery. Pol J Radiol. 86: e 114.