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Case Study

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CASE REPORT OF AN EXTRALOBAR SEQUESTRATION HYBRID LUNG LESION ASSOCIATED WITH TWO CYSTIC PULMONARY AIRWAY MALFORMATIONS IN THE IPSILATERAL LEFT HEMITHORAX

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ABSTRACT

Extralobar sequestration is a rare congenital anomaly characterized by a mass of pulmonary parenchyma with it is own pleura, lacking communication with the tracheobronchial tree, with a blood supply from descending aorta and varied venous drainage. In this case we report the association of a congenital cystic adenomatoid malformation, a lesion that communicates with the tracheobronchial tree and has a direct arterial supply to the pulmonary circulation. The risk factors and etiology are not completely elucidated, and the scarcity of reports about the occurrence of both of these pathologies makes the rarity of the case even greater and demonstrates the relevance of this study. We describe a case of ipsilateral left hemithorax lesions in a newborn patient who was diagnosed prenatally by a 32-week ultrasound. Early surgical intervention on the ninth day after birth instituted an effective operative excision with curative intention to treat.

KEYWORDS: Extralobar sequestration, case report, congenital cystic adenomatoid malformation, left hemithorax.

INTRODUCTION

Huber described in 1777 a pulmonary segment that presented its own blood supply disconnected from the tracheobronchial tree, and posteriorly, in 1946, the term "sequestration" was coined by Pryce for this rare congenital anomaly.[1] Concomitantly, the first report about a congenital cystic adenomatoid malformation was originally described in 1949 by Ch'In e Tang.[2]

Pulmonary sequestration is a rare congenital malformation characterized by a non-functioning mass of pulmonary parenchyma with it is own pleura, lacking communication with the tracheobronchial tree, with a blood supply from the descending aorta and varied venous drainage. It is classified as intralobar when it shares the visceral pulmonary pleura and extralobar when it has its own pleural investment.[3,4,5]

Usually receive arterial blood supply from the descending aorta.[3,6] and venous drainage can be from the pulmonary or systemic vein, such as the subclavian, hemiazygos, and portal veins.[7,8] Already, congenital cystic malformations are usually communicated with the tracheobronchial tree, with the blood supply to the pulmonary circulation.[4]

Congenital pulmonary malformations account for up to 6.6% of all lung diseases.[9,10] intralobar being more prevalent than extralobar ones.[11] In contrast, congenital cystic adenomatoid malformation has an incidence of 25%, being one of the most common pulmonary pathologies.[4] Both of those have no predilection for gender or a specific side of the chest.[13,14] but extralobar sequestration has a higher incidence in men.[15] The etiopathogenesis of these pathologies is not fully understood, but the concomitant incidence of both suggests that they have the same embryological origin.[4]

Clinically, patients may be asymptomatic with a random diagnosis or develop symptoms such as repeated infections, cough, or hemoptysis.[16] With large cysts, symptoms appear earlier, such as progressive respiratory failure due to lesion growth and compression of adjacent lung tissue, which may progress to pulmonary hyplopasia.[4] Diagnosis is confirmed by prenatal magnetic resonance imaging.[17] and postnatal computed tomography.[5,18] Attendant, for congenital cystic

adenomatoid malformations, prenatal diagnosis can be made from the 17th week, assisting better management.[19,20,21] The treatment for both types of lesions is surgical resection, lobectomy or sequestrectomy being performed for pulmonary sequestration[4]. When asymptomatic, expectant management is recommended.[16,22,23] For congenital pulmonary malformations, early intervention with lung lesion resection is recommended, even when there are no symptoms, due to the risk of cancer evolution.[4]

The morbidity and mortality are similar to that of other conditions such as diaphragmatic hernia or pulmonary hypoplasia,[24,25,26] mostly with good prognosis.[27,28] Considering the evolution and rarity of these concomitant lesions, early diagnosis and surgical treatment are of valued importance to the best patient outcome and prognosis.

CASE REPORT

Clinical presentation

This is a case of three pulmonary lesions, ipsilateral on the left hemithorax. During a routine antenatal ultrasound scan (US) at 24 weeks, increased echogenicity on left lung base was observed. Fetal magnetic resonance imaging was performed (Figure 1-A), describing a morphologic alteration with volumetric increased of the left lung, but no anomalous vascularization was characterized.

At 32 weeks, another US was performed (Figure 1-B), identifying an extralobar sequestration with an anomalous supply from the aorta. At 37 weeks, a baby boy was born by cesarean delivery without complications, with Apgar scores of 8 (first minute) and 9 (fifth minute). No respiratory distress was observed, but he was sent to the NICU for observation and further evaluation. A CT scan was performed (Figure 1-C and D) and revealed an extensive lesion from the upper third to the base of the left lung, without clear communication with the main tracheobronchial tree and causing a mass effect on the left superior and inferior lobes with contralateral deviation of the mediastinal structures. A chest X-ray was also performed (Figure 1-E).

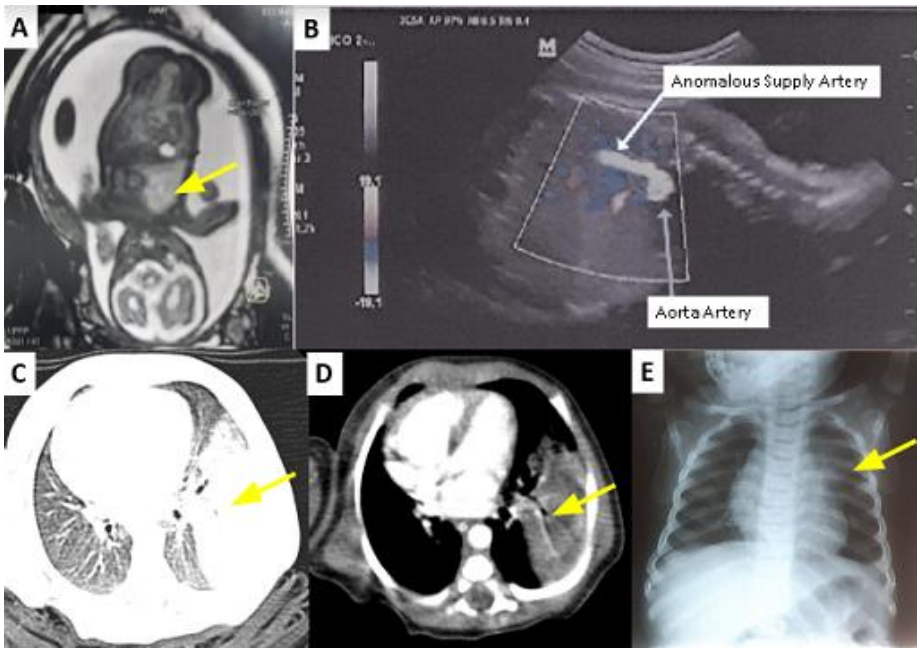

Figure 1- A: Fetal Magnetic Resonance Imaging. B: USG demonstrating anomalous vessel. C: Chest computed tomography showing a pulmonary base lesion; D: Computed tomography showing a lesion in the middle third of the lung, in segment IV. E: Chest X- Ray.

On the ninth day after birth, an effective operative excision was undertaken through a left thoracotomy (Figure 2). The extralobar sequestration was classical, located on the base of the left hemithorax, with an anomalous arterial supply from the aorta and drainage to the left pulmonary vein. Two other lesions were found in segments IV and I of the left superior lobe, compatible with a congenital cystic adenomatoid malformation. The three lesions were resected and sent for anatomopathological analysis (Figure 2).

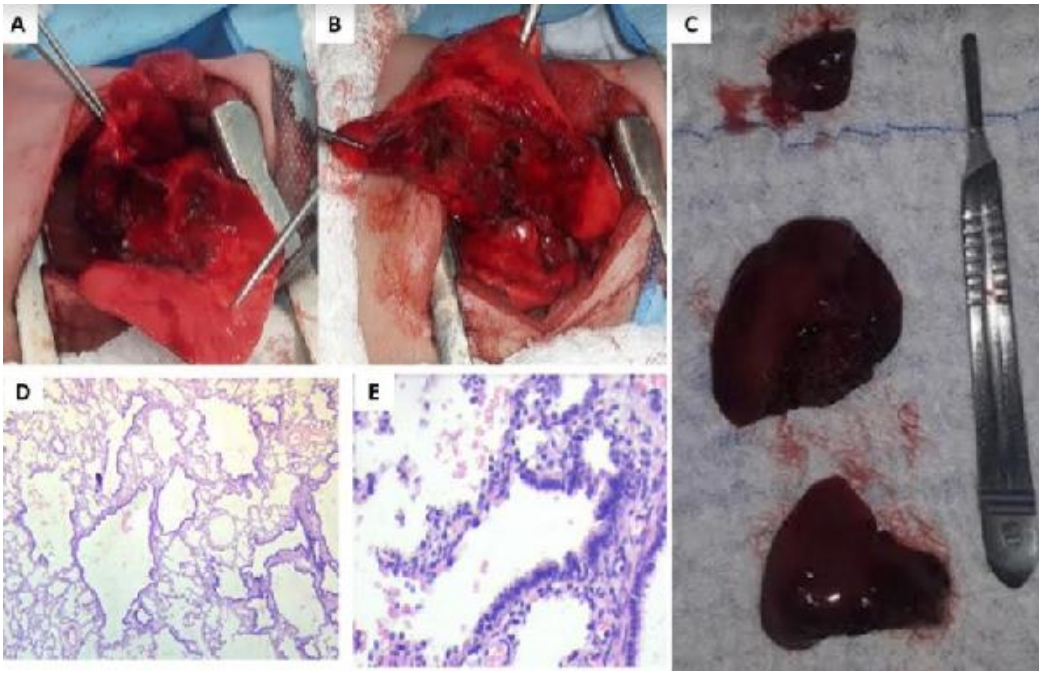

Figure 2: A: Lower lobe after pulmonary sequestration resection with anomalous artery ligation. B: Upper lobe after resection of cystic lesions areas C: From the top to the bottom, resected cystic malformation segment I, segment IV, Extralobar sequestration resected. D: Anatomopathological analysis at low magnification, mature lung tissue with dilated bronchiolar structures. E: High magnification, clustered cystic areas, lined with pseudostratified ciliated columnar epithelium, surrounding elastic tissue, and adjacent alveolar structures.

The child presented an excellent recovery, with hospital discharge 14 days after surgery. At the time of this report, he had been faring well with no postsurgical complications for 14 months.

MATERIALS AND METHODS

This study has ethics committee aproval of Hospital Pró-Cardíaco – ESHO Empresa de Serviços Hospitalares*:* CAAE: 08330819.1.0000.5533 – 3.197.288 (Plataforma Brasil)

Pathologic Evaluation

Histologic sections from tissue samples of the pulmonary sequestration resection and of the cystic malformation segment, had been processed routinely in 10% buffered formalin and embedded in paraffin, than were stained with hematoxylin and eosin (H&E) for confirmation of the diagnosis. (Figure 2 D,E)

RESULTS

The anatomopathological results were equal on the three lesions, congenital cystic adenomatoid malformation type 1. At low magnification, mature lung tissue with dilated bronchiolar structures, at high magnification, clustered cystic areas, lined with pseudostratified ciliated columnar epithelium, surrounding elastic tissue, and adjacent alveolar structures. (Figure 2 D,E).

DISCUSSION

Pulmonary sequestration is the second most common congenital lung anomaly in pediatric respiratory disease.[13] On prenatal US, extralobar sequestration appears as a well-defined echodense, homogeneous mass, and color-flow Doppler of a systemic artery from the aorta to the fetal lung lesion is pathognomonic. Magnetic resonance imaging is a very useful tool to diagnose pulmonary malformations during pregnancy; it may help to differentiate congenital pulmonary malformations from extralobar sequestration but will not always show vascular development of these lesions.[29] The main indications for resection are respiratory symptoms after birth due to a large mass or hydrothorax/hydrops, increased risk of infection, and occult malignant transformation.[4] When surgery is indicated, postoperative length of stay usually is short and survival around 100%.[14]

CONCLUSION

Extralobar pulmonary sequestration with hybrid congenital cystic adenomatoid malformation are unusual; some reports describe a range from about 34% to approximately 50 % for Stocker type II, and even more rare is the association with ipsilateral congenital cystic adenomatoid malformation lesions on the upper left lobe. To the best of our knowledge, this is the first case of extralobar sequestration in association with two more independent CPAM lesions in the left hemithorax.

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REFERENCES

1. Van Raemdonck D, De Boeck K, Devlieger H, Demedts M, Moerman P, Coosemans W, et al. Pulmonary sequestration: a comparision between pediatric and adult patients. Eur J Cardiothorac Surg, 2001; 19: 388-95.
2. Ch´in KY, Tang MY. Congenital adenomatoid malformation of one lobe of a lung with general anasarca. Arch Pathol, 1949; 48: 221-9.
3. Kestenholz PB, Schneiter D, Hillinger S, Lardinois D, Weder W. Thoracoscopic treatment of pulmonary sequestration. Eur J Cardiothorac Surg, 2006; 29(5): 815-8.
4. Claire Langston. New concepts in the Pathology of Congenital Lung Malformations. Seminars in Pediatric Surgery, 2003; 12(1): 17-37.
5. Gezer S, Taştepe I, Sirmali M, Findik G, Türüt H, Kaya S, et al. Pulmonary sequestration: a single-institutional series composed of 27 cases. J Thorac Cardiovasc Surg, 2007; 133(4): 955-9.
6. Stocker JT, Kagan-Hallet K: Extralobar pulmonary sequestration: Analysis of 15 cases. Am J Clin Pathol, 1979; 72: 917-925.
7. Gamillscheg A, Beitzke A, Smolle-Juttner FM, et al: Extralobar sequestration with unusual arterial supply and venons drainage. Pediatr Cardioi, 1996; 17: 57-59.
8. Kamata S, Sawai T, Nose K, et al: Extralobar pulmonar sequestration with venous drainage to the portal vein: A case report. Pediatr Radiol, 2000; 30: 492-494. 21- Stocker JT, Kagan-Hallet K. Extralobar pulmonary sequestration: analysis of 15 cases. Am J Clin Pathol 1979; 72: 917–925.
9. Stocker JT, Kagan-Hallet K. Extralobar pulmonary sequestration: analysis of 15 cases. Am J Clin Pathol, 1979; 72: 917–925.
10. Vogt-Moykopf I, Rau B, Branscheid D. Surgery for congenital malformations of the lung. Ann Chir, 1999; 46: 141–156.
11. Kalenahalli KV, Garg N, Goolahally LN, Reddy SP, Iyengar J: Infradiaphragmatic extralobar pulmonary sequestration: masquerading as suprarenal mass. J Clin Neonatol, 2013, 2(3): 146–48.
12. Rossel KC, Salinas RG, Kakarieka EW, Espinosa AG. Malformación adenomatoide quística pulmonar Del recién nacido. Rev Chil Pediatr, 1996; 67(4): 167-71.
13. Crombleholme TM, Coleman B, Hedrick H, Liechty K, Howell L, Flake AW, et al. Cystic adenomatoid malformation volume ratio predicts outcome in prenatally diagnosed cystic adenomatoid malformation of the lung. J Pediatr Surg, 2002; 37(3): 331-8.
14. Larsen GL, Accurso FJ, Deterding RR, Halbower AC, White CW. Respiratory tract and mediastinum. In: Hay Jr WW, editors. Current pediatric diagnosis & treatment. Apleton & Lange 15ª. ed., 1999; 418-64.
15. Corbett HJ, Humphrey GM. Pulmonary sequestration. Paediatr Respir Rev., 2004; 5(1): 59-68.
16. Cristiano Feijó Andrade; Hylas Paiva da Costa Ferreira; Gilberto Bueno Fischer. Malformações pulmonares congênitas. J. bras. Pneumol, 2011; 37(2): 259-71.
17. Liu YP, Chen CP, Shih SL, Chen YF, Yang FS, Chen SC. Fetal cystic lung lesions: evaluation with magnetic resonance imaging. Pediatr Pulmonol, 2010; 45(6): 592-600.
18. Jesch NK, Leonhardt J, Sumpelmann R, Gluer S, Nustede R, Ure BM. Thoracoscopic resection of intra- and extralobar pulmonary sequestration in the first 3 months of life. J Pediatr Surg., 2005; 40(9): 1404-6.
19. Kunisaki SM, Barnewolt CE, Estroff JA, Ward VL, Nemes LP, Fauza DO, Jennings RW. Large fetal congenital cystic adenomatoid malformations: growth trends and patient survival. J Pediatr Surg, 2007; 42(2): 404-410.
20. Volpe MV, Pham L, Lessin M, Ralston SJ, Bhan I, Cutz E, Nielsen HC. Expression of Hoxb-5 during human lung development and in congenital lung malformations. Birth Defects Res A Clin Mol Teratol, 2003; 67(8): 550-556.
21. Jancelewicz T, Nobuhara K, Hawgood S. Laser microdissection allows detection of abnormal gene expression in cystic adenomatoid malformation of the lung. J Pediatr Surg., 2008; 43(6): 1044-1051.
22. Laberge JM, Puligandla P, Flageole H: Asymptomatic congenital lung malformations. Semin Pediatr Surg, 2005; 14(1): 16–33.
23. Adzick NS, Harrison MR, Crombleholme TM, Flake AW, Howell LJ: Fetal lung lesions: management and outcome. Am J Obstet Gynecol, 1998; 179(4): 884–889.
24. Rosado-de-Christenson ML, Stocker JT: Congenital cystic adenomatoid malformation. Radiographics, 1991; 11: 865-886.
25. Atkinson JB, Ford EG, Kitagawa H, et al: Persistent pulmonary hypertension complicating cystic adenomatoid malformation in neonates. J Pediatr Surg, 1992; 27: 54-56.
26. Njinimbam CG, Hebra A, Kicklighter SD, et al: Persistent pulmonar hypertension in a neonate with cystic adenomatoid malformation of the lung following lobectomy: Survival with prolonged extracorporeal membrane oxygenation therapy. J Perinatol, 1999; 19: 64-67.
27. Adzick NS: Fetal thoracic lesions. Semin Pediatr Surg, 1993; 2: 103- 108.
28. Souza J.et al. Malformação adenomatóide cística pulmonar em crianças: análise de 10 casos. Arquivos Catarinenses de Medicina, 2003; 32(4): 47-55.
29. Adzick NS: Fetal thoracic lesions. Semin Pediatr Surg, 1993; 2: 103- 108.