July-December 2020 Volume 29 Issue 2
PRINT ISSN: 2277-1867
ONLINE ISSN: 2277-8853

JOURNAL OF FORENSIC MEDICINE SCIENCE AND LAW

Official Publication of Medicolegal Association of Maharashtra

MULTISPECIALITY, MULTIDISCIPLINARY, NATIONAL
PEER REVIEWED, OPEN ACCESS, MLAM (SOCIETY) JOURNAL

Editorial Office Address
Department of Forensic Medicine & Toxicology, Third Floor, Library Building, Seth G S Medical College & KEM Hospital, Parel, Mumbai, Maharashtra, India. Pin-400 012. Email id: mlameditor@gmail.com Phone: 022-24107620 Mobile No. +91-9423016325.

JOURNAL OF FORENSIC MEDICINE SCIENCE AND LAW
(Official Publication of Medicolegal Association of Maharashtra)
Email.id: mlameditor@gmail.com

PRINT ISSN: 2277-1867
ONLINE ISSN: 2277-8853

Case Report

Death due to Pulmonary and Splenic lymphangiectasia: A Rarely Diagnosed Disorder.

Laxman G. Phad[a], Rajesh V Bardale[b], Sandeep V. Haridas[b], Pradeep G. Dixit[c]

[a]Assistant Professor, [b]Associate Professor, [c] Professor and Head,

Department of Forensic Medicine & Toxicology, Government Medical College and Hospital, Miraj, Dist. Sangli, Maharashtra, India.410410

Article Info

Received on: 27.05.2020
Accepted on: 26.10.2020

Key words
Lymphatic dilatation, Developmental disorder, Lymphangioma, Hamartoma.

Abstract

Pulmonary lymphangiectasia is a rare developmental disorder involving the lungs, characterized by pulmonary subpleural, interlobar, perivascular and peribronchial lymphatic dilatation. Splenic lymphangiectasia is characterized by lymphatic dilatation in the spleen with impaired splenic function. The current case is taken for presentation considering the rarity of the disease condition and the mortality arising out of it is still rarest. It also aims to highlight the clinical presentation of disease because most of the times cases are sporadic and aetiology is not completely understood so early suspicion of case, diagnosis and prompt treatment may help to save the precious life.

1. Introduction

Congenital pulmonary lymphangiectasia is a rare developmental disorder involving the lungs, characterized by pulmonary subpleural, interlobar, perivascular and peribronchial lymphatic dilatation. Pulmonary lymphangiectasia presents at birth with severe respiratory distress, tachypnea and cyanosis, with a very high mortality rate at or within a few hours of birth. Most reported cases are sporadic and the etiology is not completely understood.[1] Although there have often debates on the classification and differential diagnosis of the disease, it can usually be divided into primary (congenital) and secondary forms.[2] Normal pulmonary artery wedge pressure and radiographic appearance of increased interstitial markings should lead to suspicion of pulmonary lymphangiectasia. The substernal thickening in three patients is also characteristic of substernal lymphatic collection.[3] Pulmonary lymphangiectasia presents with dilated pulmonary lymphatics as part of a generalized form of lymphangiectasia, i.e., truncal lymphangiectasia, which is usually associated with lymphedema.[1]

Splenic lymphangiectasia is characterized by lymphatic dilatation in the spleen with impaired splenic function. Splenic lymphangioma is a neoplasm or a hamartoma, most researchers support the latter opinion; its formation is proposed to be due to abnormal congenital development of lymphatic vessels. It can also be attributed to bleeding or inflammation in the lymphatic system, which causes obstruction and consequent lymphangiectasia.[4,5] Along with these two forms the other form which is prominently found is Intestinal lymphangiectasia. Which is characterized by dilated lymphatics, protein-losing enteropathy, hypoalbuminaemia and oedema, and patients with this disease lose albumin, immunoglobulins, and lymphocytes into the bowel.

How to cite this article: Phad LG, Bardale RV, Haridas SV, Dixit PG. Death due to Pulmonary and Splenic lymphangiectasia: A rarely diagnosed disorder. J For Med Sci Law 2020;29(2):70-74.

***Corresponding author:** Dr Rajesh V Bardale, Associate Professor, Govt. Medical College and hospital Miraj, Sangli, Maharashtra, India. Pin-410410. Email: bardalerv@yahoo.co.in (M): +91- 9420254589

If the development of splenic atrophy results from prolonged lymphocyte depletion, it is likely that patients with intestinal lymphangiectasia will develop abnormalities of splenic function over the period of time.[6]

As far as global burden of disease is concerned, various Autopsy studies suggest that approximately 0.5–1% of infants who are stillborn or die in the neonatal period have Pulmonary lymphangiectasia.[7] This condition carries a poor prognosis with a mortality rate ranging from 50% to 98%, and the incidence of congenital chylothorax is about 1:10,000–15,000 pregnancies, with a male-female ratio of 2:1.[8]

Recent advances in intensive neonatal care have changed the previously nearly fatal outcome of pulmonary lymphangiectasia at birth. Patients affected by pulmonary lymphangiectasia who survive infancy, present medical problems which are characteristic of chronic lung disease.[1]

The current case is taken for presentation considering the rarity of the disease condition and the mortality arising out of it is still rarest. The aim is also to highlight the clinical presentation of disease, as most reported cases are sporadic and the etiology is not completely understood so early suspicion of case, diagnosis and prompt treatment may help to save the precious life.

2. Case report

A fourteen-year old male child brought to casualty of government medical college and hospital with complaints of severe pain in abdomen and one episode of vomiting in since 3 hours. Patient was apparently alright before this episode. The pain in abdomen is sudden in onset, progressive in nature and confined to epigastric region. Patient is irritable and restless. No history of chest pain, cough, fever, palpitation. The Sleep, appetite, bladder and bowel habits were normal before the episode.

The patient was examined thoroughly and given symptomatic treatment for the same and routine blood investigations and other lab investigations were sent.

On examination

Patient is restless with pain in abdomen; tachypnea was present and prominent cyanosis present, pallor present, profuse sweating seen, Pulse rate was 136 /min.

On systemic examination: cardiovascular system shows S1, S2 was normal, no murmur were auscultated. Respiratory system showed bilateral air entry with tachypnea, cyanosis, and pallor. Per abdominal examination shows tenderness in epigastric region. Central nervous reflexes were within normal limits.

Investigation.

Blood and lab investigation:

Total Serum bilirubin, direct serum bilirubin and indirect serum bilirubin were 0.6 mgs/dl, 0.3mgs/dl, and 0.3 mgs/dl respectively. The indirect serum bilirubin was slightly elevated other two parameters within normal limits. The Serum electrolyte values sodium was 145 mmol/lit and potassium was - 4.4 mmol/lit which are within normal limits. The Serum creatinine was 1 mg/dl, which is within normal limit. Blood urea level was- 15 mg/dl, which is within normal limit. Blood sugar level was- 212 mg/dl, which is elevated. The complete blood count including platelet count was within normal limit no any abnormality seen.

But three hours after admission the patient become restless, tachypnic and was not responding to any stimuli and declared dead. The cause of death remains undiagnosed in spite of meticulous examination and all laboratory investigations and the disease picture is was something different so they referred to Forensic Medicine Department for autopsy to know actual cause of death.

Postmortem examination:

The body was brought to autopsy room in the same hospital for examination along with police inquest and requisition letter.

On external examination the body was male aged 14 years, thin built, Cloths were intact Rigor mortis partially present, postmortem lividity present on back and buttocks and not fixed faint in colour. Nail, tip of nose and lips were cyanosed. No any other external injury or abnormality seen over the body.

On internal examination of thoracic cavity the pleura is pale and loosely attached to lungs with no collection is noted in pleural cavity. Lungs were pale, oedematous, enlarged and heavy with granular appearance, patchy consolidation was present at places. On cut section small cystic spaces were present at places with whitish coloured thin fluid in it. Spleen was enlarged, mushy in appearance. On

cut section cystic dilated spaces seen. The cranial cavity was unremarkable brain and its coverings were pale. Liver was pale and unremarkable, kidneys were pale and unremarkable, and heart was pale and unremarkable.

Histopathological examination

The lungs show findings of lymphangiectasia, focal granulomatous inflammation, and few thrombi in medium sized pulmonary vessels. Spleen shows evidence of lymphangiectasia.

Figure1: Showing granular appearance of lungs.

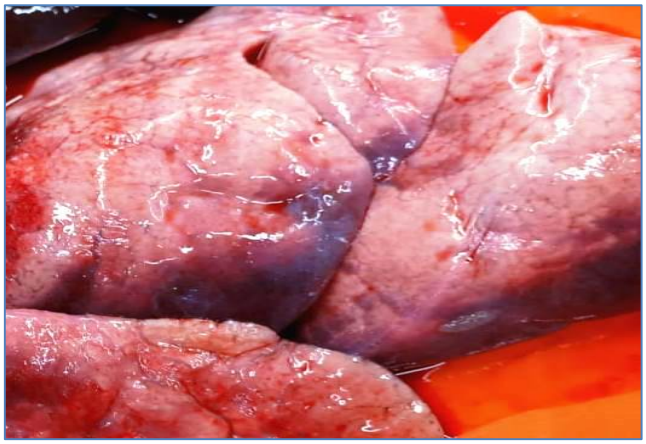
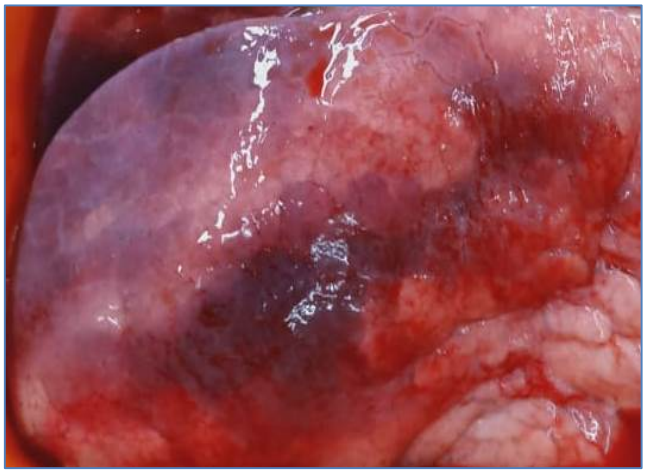


Figure 2: Showing areas of patchy consolidation over the lungs.



3. Discussion

Noonan et al. classified the pulmonary lymphangiectasis into three groups. Group 1 is a generalized form of lymphagiectasis (lymphedema with intestinal lymphangiectasis), group 2 is due to pulmonary venous hypertension or obstruction associated with cardiovascular anomalies, and group 3 includes patients compromised by a primary developmental defect of the pulmonary lymphatics.[9] New classification divides the lymphangiectasis into the primary (congenital) and secondary forms to differentiate it from the lymphangiomatosis, and they noted that the primary form presents in neonates and is usually fatal. The secondary form of lymphangiectasis results from a variety of processes that impair lymphatic drainage and increase lymph production. They proposed that primary and secondary lymphangiectasis can be distinguished by the age of the patients and their clinical courses.[10]

Pulmonary lymphangiectasia may present at birth as a stillbirth or with severe respiratory distress, tachypnea, and cyanosis, with a very high mortality rate at or within a few hours of birth.[11] The etiology of PL is not known. Although no consensus has yet been reached on whether splenic lymphangioma is a neoplasm or a hamartoma, most researchers support the latter opinion; its formation is proposed to be due to abnormal congenital development of lymphatic vessels.[5] It can also be attributed to bleeding or inflammation in the lymphatic system, which causes obstruction and consequent lymphangiectasia.[12,13]

It has been suggested that Pulmonary lymphangiectasia, lymphatic channels of the fetal lung do not undergo the normal regression process at 20 weeks of gestation, and thus there is a persistence of the large lymphatic vessels that are normal form of the maturation developmental process at 9–16 weeks of gestation.[1] Obstruction of pulmonary lymphatics or veins, or the actions of infectious agents have also been taken into consideration.[11]

The lymphatics of the lungs have valves that direct the flow of lymph toward the hilum.[14] From the hila, the lymph is carried by the broncho mediastinal trunks to the subclavian veins. There are right and left broncho mediastinal trunks with many anastomotic channels between them. Most of the lymph of the lungs goes to the right broncho mediastinal trunk, except the left upper lobe which is drained mainly by the left broncho mediastinal trunk.[15] Pulmonary lymphangiectasis may be the result of dilatation and ectasia of the pulmonary lymphatics due to incompetent valves or agenesis, or interruption of the thoracic duct with establishment of collateral pathways through the diaphragm and parietal pleura to the internal mammary chain.[3]

Hennekam Lymphangiectasia syndrome is a rare disorder characterized by presence of intestinal and renal lymphangiectasia, dysmorphic facial appearance and mental retardation. The syndrome is familial and was first reported in two male and two female children of consanguineous parents. The pattern of transmission was autosomal recessive.[16] Eom M. et al reported two cases of primary CPL in a 13-day-old male neonate and a one-day-old male neonate, showing prominent lymphatic dilatation in the septal, subpleural, and peri-bronchial tissue throughout both lungs. The latter case was associated with congenital cardiac anomaly including single ventricle. These are unique cases of CPL in Korea of which the diagnosis was established through post-mortem examination.[10]

Foster et al described a patient with intestinal lymphangiectasia who developed hyposplenism and speculate that it resulted from chronic loss of lymphocytes into the gut. This patient was saved with meticulous investigations and prompt, vigorous treatment.[6] Lahiri et al described the duodenal and splenic lymphangiectasia can exist in a scenario of chylous ascites without any obvious obstruction of lymphatic channels and in the absence of yellow nail syndrome. They describe the case of a 54-year-old man presenting with chylous ascites, lymphangiectasia and nephrotic syndrome with focal segmental glomerulosclerotic lesion in his kidney.[17]

4. Conclusion: Pulmonary lymphangiectasia is a rare developmental disorder involving the lungs, characterized by pulmonary subpleural, interlobar, perivascular and peribronchial lymphatic dilatation. Splenic lymphangiectasia is characterized by lymphatic dilatation in the spleen with impaired splenic function. Recent advances in intensive neonatal care have changed the previously nearly fatal outcome of pulmonary lymphangiectasia at birth. Patients affected by pulmonary lymphangiectasia who survive infancy, present medical problems which are characteristic of chronic lung disease. As far as forensic aspect of the disease is concerned the autopsy surgeon must look for the findings of this particular condition to reach the appropriate cause of death and also the associated organ involvement, which will help the clinicians in early diagnosis and prompt treatment may help to save the precious life.

Conflict of interest: None declared.

Funding: None

References:

1. Bellini C, Boccardo F, Campisi C, Bonioli E. congenital pulmonary lymphangiectasia, orphanet J Rare Dis. 2006;1:43.
2. Hilliard R, Mckendry J, Philips M. congenital abnormalities of the lymphatic system: a new clinical classification. Pediatrics 1990; 86: 988-94.
3. Hernandez R, Aron M, Amnon Rosenthal, Pulmonary Lymphangiectasis in Noonan Syndrome, Am Journal of Roentgenology 1980;134:75-80.
4. Chung S, Park Y, Jo Y, et al. asymptomatic lymphangioma involving the spleen and retroperitoneum in adults. World J Gastroenterol. 2009;15(44):5620–5623.
5. Ioannis I, and Kahn A, splenic Lymphangioma, Archives of Pathology & Laboratory Medicine: February 2015; 139(2): 278-282
6. Foster P, Bullen A, Robertson D, Chalmers D, Losow sky M, Development of impaired splenic function in intestinal lymphangiectasia. Gut 1985; 26:861–864.
7. Esther C, Barker P, Pulmonary lymphangiectasia: Diagnosis and clinical course. Pediatr Pulmonol 2004;38:308-313
8. Bukowski R, Saade G, Hydrops fetalis, Clinical Perinatol, 2000;27:1007-1031.
9. Noonan J, Walters L, Reeves J. Congenital pulmonary lymphangiectasis. Am J Dis Child 1970; 120: 314-9
10. Eom M, Choi Y, Kim Y, Cho M, Jung S, Lee H. Clinico-pathological characteristics of congenital pulmonary lymphangiectasis: report of two cases. J Korean Med Sci. 2007;22(4):740-745.
11. Bellini C, Mazzella M, Campisi C, Taddei G, Mosca F, Toma P, Villa G, Boccardo F, Sementa AR, Hennekam RC, Serra G: Multimodal imaging in the congenital pulmonary lymphangiectasia-congenital chylothorax-hydrops fetalis continuum. Lymphology 2004; 37:22-30.
12. Abbott R, Levy A, Aguilera N, Gorospe L, Thompson W, From the archives of the AFIP: primary vascular neoplasms of the spleen: radiologic pathologic correlation. Radiographics. 2004;24(4):1137–1163.
13. Patti R, Iannitto E, DiVita G. Splenic lymphangiomatosis showing rapid growth during lactation: a case report. World J Gastroenterol. 2010;16(9):1155–1157.
14. Trapnell D, The peripheral lymphatics of the lung. Br J Radio 1963;36 :660-672.

15. Roujeau J. La circulation lymphatique du poumon et sa pathologie. Rev Tuberc Pneumol 1971 35:267-280.
16. Hennekam R, Geerdink R, Hamel B, Hennekam F, Kraus P, Rammeloo J, Autosomal recessive intestinal lymphangiectasia and lymphedema, with facial anomalies and mental retardation. Am J Med Genet 1989;34:593-600.
17. Lahiri, D., Agarwal, R., Roy, M, Chylous ascites and lymphangiectasia in focal segmental glomerulosclerosis – a rare coexistence: a case report. J Med Case Reports 2015; 9: 34.