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Pulmonary lymphangiectasia: A case report

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Abstract
Congenital pulmonary lymphangiectasis [CPL] is a rare, poorly documented disease, characterized by abnormal dilatation of pulmonary lymphatics without lymphatic proliferation. This disease is seen almost exclusively in infancy and early childhood. It can usually be divided into primary [congenital] and secondary forms. The primary form presents in neonates, and the patients mostly die due to the respiratory distress, shortly after birth. Here we present a case of pulmonary lymphangiectasia in an one month old child as incidental finding on autopsy.

Keywords: pulmonary, lymphangiectasia, autopsy, congenital

Introduction
CPL is a rare developmental disorder involving the lung and characterized by pulmonary subpleural, interlobar, perivascular and peri bronchial lymphatic dilatation. CPL presents at birth with severe respiratory distress, tachypnoea and cyanosis with a very high mortality rate at or within a few hours of birth. It has been suggested that in CPL, pulmonary lymphatic channels of the foetal lung do not undergo the normal regression process at 20 weeks of gestation. Secondary developmental pulmonary lymphangiectasia [PL] may be caused by a cardiac lesions. Autopsy studies suggest that approximately 0.5- 1% of infants who are stillborn or die in the neonatal period have PL [1].
CPL is a uniformly developmental defect that is related to an apparent failure of the communication between the pulmonary lymphatics and the systemic lymph channels. Most cases of CPL are diagnosed clinically or radiologically. However, there are rare fatal cases that were pathologically confirmed on autopsy [2].
In 1959, Laurence published article reporting on 10 cases of stillbirth or neonatal death that showed evidence of dilated pulmonary lymphatic vessels on post-mortem examination. This was the first time the term ‘congenital pulmonary lymphangiectasis’ had been used in a publication. This condition is now generally known as congenital pulmonary lymphangiectasia’, but may also still be referred to as congenital pulmonary lymphangiectasis [3].
PL is an abnormal dilatation of the lymphatics draining the interstitial and subpleural (SP) space of the lungs. It is a rare condition, first described by Virchow in 1856. In PPL, the lymphatic abnormality can be localised to the lung or be part of a more widespread abnormality of lymphatic drainage. Early descriptions of pulmonary lymphangiectasia from the pathology literature reported an incidence of 1% in consecutive necropsies on stillbirths and neonates. The true incidence of PPL after the new born period is not known and reports of PPL in infants and children are restricted to isolated case reports [4]. Although first described by Virchow over a century ago, only 45 cases have been reported to date [5].

Case Report
A female child with an unknown gestational and delivery history, died at the age of one month. She has a history of cyanosis and not passing of urine and stool for 4-5 days. Autopsy revealed pleural effusion with cardiopulmonary oedema. Both the lungs were pale externally with numerous tinny cystic spaces within the lung parenchyma on cut sections. All other organs were congested. Under light microscopy of both the lungs [Fig.1, 2], there was widespread dilatation of lymphatics under the visceral pleura and in the interlobular septa and adjacent to pulmonary veins.

The interlobular septa are widened and prominent. These lymphatics were much larger than the neighbouring veins, which was converse of the normal ratio. Dilated lymphatics were also found in the peri-broncho-vascular bundle area. Each lung had a normal lobation. Other organs were unremarkable on microscopy. We arrived at a diagnosis of primary/ congenital pulmonary lymphangiectasia after ruling out interstitial oedematous changes, emphysematous changes and lymphangiomatosis of both the lungs.

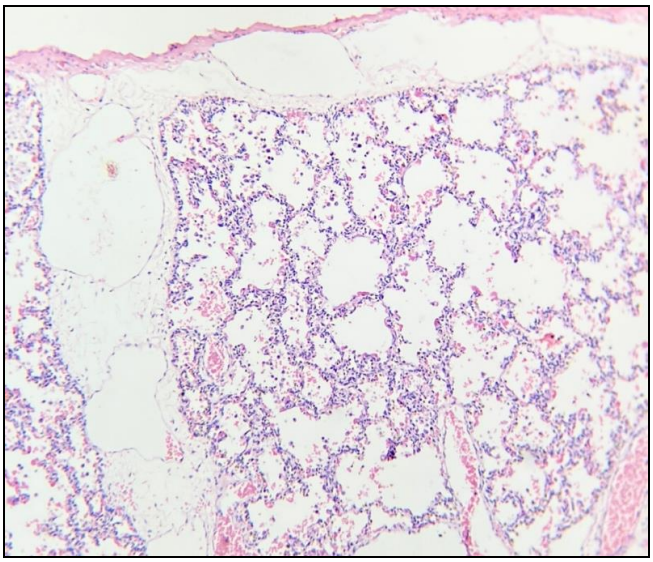

Fig 1: Shows dilatation of lymphatic channels lined by flat endothelial cells in the subpleural area and interlobar septa. [H & E: 10X]

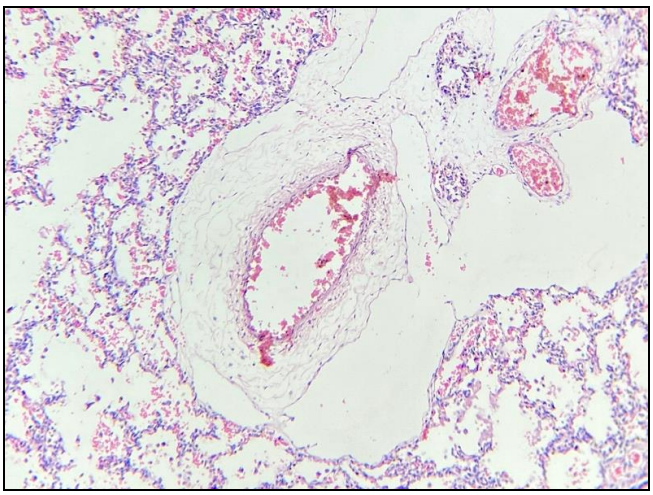

Fig 2: Shows dilatation of lymphatic channels lined by flat endothelial cells around the pulmonary vein. [H & E: 10X]

Discussion

First described by Virchow in 1856, CPL is a rare lymphatic vessel malformation characterized by abnormally dilated and thin-walled pulmonary subpleural, interlobar, perivascular, and peri bronchial lymphatic channels. In secondary pulmonary lymphangiectasia, dilated lymphatic vessels most commonly develop in conjunction with a variety of congenital heart defects, such as total anomalous pulmonary venous return, stenosis of the pulmonary and mitral valves, hypoplastic left heart, cortriatriatum, atresia of the pulmonary veins and atrioventricular canal defects. More widespread abnormalities of lymphatic drainage have also been reported in cases of pulmonary lymphangiectasia involving dilation of lymphatic vessels in bones, viscera, and soft tissues. The incidence of CPL is not clearly defined since only a few isolated cases or small case series have been reported. Autopsy studies suggest, however, that approximately 0.5-1% of perinatal deaths may be attributed to CPL. Most cases of CPL are sporadic and although familial occurrences have been described, no underlying genetic aetiology has yet been identified. Two familial conditions in which CPL may occur are Hennekam syndrome, which presents as congenital lymphedema with facial anomalies, intestinal lymphangiectasia, as well as varying degrees of mental retardation, and Njolstad syndrome, which is characterized by CPL along with facial and lower limb lymphedema. CPL has also been associated with 46,XY/46,XX mosaicism, ichthyosis congenita, Noonan's syndrome, Turner's syndrome, Fryns syndrome, and Down's syndrome. The mortality rate for CPL is variable, ranging from 50% to 98%. CPL associated with systemic lymphangiectasia seems to have a slightly better prognosis than isolated CPL. The combination of CPL with hydrops fetalis, bilateral chylothorax or the immediate onset of severe respiratory distress at birth has the worst prognosis [6].

Recently, Faul *et al*. also divided 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. Macroscopically, the lungs of pulmonary lymphangiectasis appear heavy and noncompliant. A small amount of collagen and smooth muscle may be found in the walls of vessels, particularly in the secondary form of pulmonary lymphangiectasis. Flat cells lining the lymphatic spaces are immunohistochemically stained for CD31, and this process indicates their endothelial nature, and it excludes the diagnosis of interstitial emphysema. In pulmonary emphysema, the spaces may be devoid of lining cells but may be connected in some areas with alveoli. The immunohistochemical staining for cytokeratin also showed the pneumocytes, lining intact alveolar walls. Interstitial emphysema can be distinguished from congenital cystic adenomatoid malformation because in interstitial emphysema there are areas of normal lung between the cysts. By contrast, adenomatoid malformation is composed of abnormally arranged immature lung, in which the cysts are usually lined with prominent epithelium including goblet cells. It is also occasionally difficult to differentiate pulmonary lymphangiectasis from lymphangiomatosis by histological examination because both conditions have similar clinical manifestations and histological features. Tazelaar *et al*. proposed that the term ''lymphangiectasis'' be reserved to describe those extremely unusual congenital or secondary lesions in which the primary alteration is a dilatation of existing lymphatic channels, without an increase in their number or complexity, while the term ''lymphangiomatosis'' be used for those diffuse lesions characterized primarily by an increased number of complex anastomosing lymphatic channels in which dilatation is a secondary phenomenon. Lymphangiomatosis and CPL share a similar immunohistochemical profile for vimentin, factor VIII-related protein, CD31, CD34, and smooth muscle actin. Histological findings of the lungs in our case reveals diffuse dilatation of existing lymphatic channels rather than an

abnormal increase in their number or complexity. Therefore, the possibility of lymphangiomatosis can be ruled out. Recently, it was reported that another monoclonal antibody D2- 40 is a highly sensitive and specific marker of lymphatic endothelium in normal tissue and vascular tumour [2].

Conclusion

CPL is a rare but serious condition marked by dilatation of the pulmonary lymphatic vessels. It can manifest in utero as hydrops fetalis and/or polyhydramnios; immediately or shortly after birth as cyanosis, tachypnoea and respiratory distress; or weeks to months into the neonatal period.[3]

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