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LEFT HEMI THORAX INVOLVEMENT IN POLAND SYNDROME – A RARE CASE REPORT WITH REVIEW

Paediatric Surgery

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ABSTRACT

Background-- Poland syndrome is a rare anomaly. It was first described by Poland a medical student on anatomic dissection in 1841. [1] The incidence is 1 in 30000-35000 live births. [2]. **Clinical description--** It is characterized by absence of sternocostal portion of pectoral muscles on one side with other associated anomalies like hand deformities, aplasia or hypoplasia of second to fifth ribs, underdeveloped breast tissue etc. Sternocostal portion of Pectoralis major muscle is always absent on one side in this syndrome. Syndactyly and variable degree of brachydactyly and absence of latissimus dorsi muscle is present in some cases of Poland syndrome. The etiology is of this syndrome is uncertain. It commonly involves right hemi thorax but in our case left side was involved. **Management--** Very few children need surgery with severe chest deformity. **Conclusion –** Although most of the cases are asymptomatic , but It can be diagnosed by proper clinical examination of chest showing bulging of localized part of chest wall due to muscular defect especially in newborn.

KEYWORDS

Pectoralis major, Poland's syndrome, Rib defects

INTRODUCTION—
Poland syndrome presents as chest deformity usually without any complaint. Ipsilateral chest wall is depressed due to lack of sternocostal portion pectoralis muscle with or without rib aplasia. Thompson summarizes the full spectrum of anomalies like absence of ribs, absence of axillary hair, and chest wall depression, athelia or amastia, Syndactyly and limited subcutaneous fat in 1895. [3]. The incidence of Poland syndrome is more common in male as compared to female with ratio of 3:1. The involvement of right hemi thorax is about three fourth of total cases. Here we are presenting a very rare case with left hemi thorax involvement. Sometimes it is associated with severe chest deformity. Surgical intervention is required to corrected chest deformity of severe degree.

Case report—
Five month old child came to us with complaint of tachypnoea and localized abnormal movement in left anterior chest wall. Child was diagnosed outside as eventration of left diaphragm. The child was examined thoroughly. He was full term normally delivered child. Developmental milestones were within normal limit. On clinical examination, there was localized depression of left anterior chest wall lateral to sternum. There was localized bulging with each respiration. No other external deformities were present. On palpation there was feeling of gap on that part of chest showing absent ribs and muscles through which lung was herniated during respiration. On X ray chest left hemi diaphragm was slightly elevated. Then contrast enhanced CT scan was done to confirm the diagnosis of Poland syndrome. CECT chest showed rib defect with absence of sternocostal part of pectoral muscles and latissimus dorsi. There was no eventration of diaphragm. As our patient had associated upper respiratory tract infection so antibiotics along with nebulization was done. After recovery from upper respiratory tract infection patient was discharged. Surgical intervention was not planned as chest deformity was not severe.

DISCUSSION –
The first description of Poland syndrome was described in 1841. Poland, a medical student first described it as absence of pectoralis major and minor muscles and Syndactyly in anatomic dissection. The term Poland syndrome was given by Clarkson in 1962. [4]. Thompson summarizes the full spectrum of anomalies like absence of ribs, absence of axillary hair, chest wall depression, athelia or amastia and limited subcutaneous fat. [5].

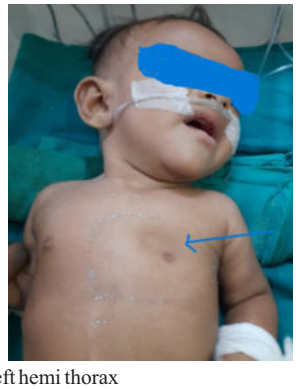
Poland syndrome is charactized by aplasia or hypoplasia of the sternocostal part of the pectoralis major muscle and at least on other associated lesion. Hand anomalies are widely varied. Ireland et. All [6] reported Syndactyly and variable degree of brachydactyly. Thoracic deformity ranges from normal ribs to complete aplasia of second to fifth ribs but second rib is least common. The incidence of Poland syndrome is 30000 to 32000 live births. This is more common in male as compared to female and commonly involve right hemithorax. But in our patient, it was on the left side of chest which is very rare. Etiology of this syndrome is uncertain. This is rarely familial. [7] There are various theories regarding its Etiology. 1) Abnormal migration of the embryonic tissue forming the pectoral muscles, 2) Hypoplasia of subclavian artery, [8]. 3) In utero injuries due to attempted abortion. None of the theory is universally accepted. [9]

CECT Chest is diagnostic tool for Poland syndrome. [10] This helps for assessment of thoracic deformities like aplasia or hypoplasia of pectoral muscles, absent ribs and absence of latissimus dorsi muscle.

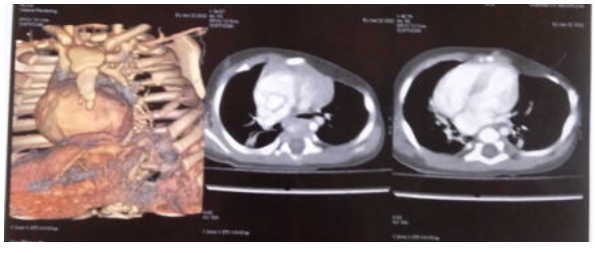
Management— Surgical intervention is required only in case with severe chest deformity. Ravitch described the reconstruction of aplastic ribs with autologous rib grafts and use of latissimus dorsi flap to cover the rib for good cosmesis as well as functional improvement. [11] Haller et. all published report regarding simultaneous latissimus dorsi muscle flap with rib graft. [12] The key step in surgery is correction of abnormal position and rotation of sternum.

CONCLUSION –
Poland syndrome is very rare congenital anomaly presenting with hypoplasia of one side of hemi thorax without any complaint. Contrast enhanced CT scan is investigation of choice. Surgical management is required only in patients with severe chest deformity.

Picture legend



1. Deformity in left hemi thorax



2. CECT chest

3. X – Ray chest

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