

Case Report

A Rare Case of Poland's Syndrome

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ABSTRACT

Poland's syndrome is a rare congenital anomaly consisting of a unilateral absence of the pectoralis major, hand anomaly and occasionally associated other malformation. Many structural and functional abnormalities have been described in association with this syndrome. We report an incidentally diagnosed case in a 4 year male child presented to us with brachydactyly.

Key words: Poland's syndrome, congenital anomaly, pectoralis major

INTRODUCTION

Poland described a case of unilateral absence of the pectoralis major muscle in 1841. [1] Thompson first described the full anatomical spectrum of the disease. [2] Baudinne et al described the term Poland's syndrome in 1967. [3] The clinical presentation are variable. The typical components of the syndrome include ipsilateral hypoplasia or absence of the sternocostal portion of the pectoralis major muscle with associated hand defects. [4] The reported incidence of Poland's syndrome ranges from 1:7000 to 1:100000. Most cases are sporadic and tend to occur in male (2-3:1) and is more commonly seen in the right side (2:1). Familial cases are rare, equal in both sexes with no side predominance. [5] 10% of these patient also express other features of Poland's syndrome.

CASE HISTORY

A 4 year old male child presented with complain of small right hand that was present since birth. On physical examination, the pectoralis major muscle was absent (**figure-1**) and brachydactyly was present in right side whereas the left side chest and upper arm was found to be normal (**figure-2**). Routine haematological and biochemical investigation were within normal limit. On hands radiograph, phalanges of right hand were found to be small as compare to left hand (**figure-3**). Chest radiograph revealed hyperlucency of right hemithorax with normal appearing heart shadow (**figure- 4**). Ultrasonography of the abdomen and electrocardiogram did not reveal any abnormalities. None of the family members had reported similar anomalies.



Figure -1 Absent anterior axillary fold and hypoplastic chest on right side



Figure-2 Small finger on right side

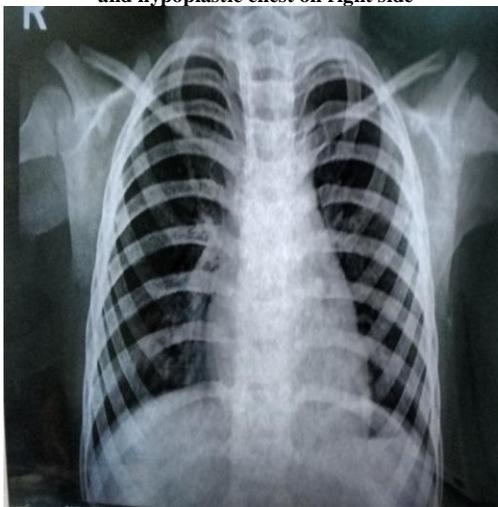


Figure-3 Hyperlucency on right side of lung



Figure-4 Small phalanges on right side

DISCUSSION

Poland syndrome is a rare congenital anomaly typically characterised by unilateral chest wall hypoplasia and ipsilateral hand anomalies.^[6] The absence of pectoralis major muscle on one side constitutes the hallmark. The other abnormalities may include hemivertebrae, renal anomalies, dextrocardia, Sprengel deformity, club foot and submucous cleft palate.^[7] It has been postulated that various factors like congenital vascular maldevelopment, intrauterine insults and drugs could ultimately result in hypoxia to one side of the fetus, as limb bud develops adjacent to the chest wall leading to the development of the syndrome. Various associated anomalies including diaphragmatic hernia and those of the liver and biliary tract, kidney, testes and heart

have been reported and neoplasm such as leukaemia, non-Hodgkin's lymphoma.

This diagnosis is important as this condition may be associated with several visceral anomalies and neoplasm. Chest X-ray and ultrasonography of abdomen should be obtained. This syndrome can also be diagnosed prenatally using ultrasonography. The treatment of patient with Poland's syndrome is individualised and depends on patient's age, gender and pathology and severity. Functional disability is minimal and patient may seek a surgical opinion for cosmetic reasons. Several reconstructive procedures like flaps, lipofilling and breast prosthesis. Treatment for syndactyly should be conducted at preschool age.

CONCLUSION

Poland's syndrome is a rare condition but easy to diagnose. Poland's syndrome may be an incidental finding because of mild functional disability. Patient may present for aesthetic reason. Presence of brachydactyly/ syndactyly should prompt a search for other features of Poland's syndrome. It is important to rule out other developmental anomalies and associated tumours for early diagnosis and treatment.

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