Poland Syndrome, a rare entity

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Citation

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Abstract

Poland syndrome is rare entity with average incidence of one in 30,000[4]. Named after Sir Alfred Poland, Poland syndrome includes the features of under development of the chest muscle on one side of the body, cutaneous syndactaly on the same side of the body, and some time the lack of development of the breast bone. We report a case of 12 years old male boy with this rare syndrome.

INTRODUCTION

Poland syndrome can present with ipsilateral involvement of the chest muscles, skin and subcutaneous tissues, bones, and upper extremity. The absence of the sternal head of the pectoralis major muscle is considered the minimal expression of this syndrome. Involvement of adjacent muscles, including the pectoralis minor, serratus, latissimus dorsi, and the external oblique, also has been described. Most Poland syndrome cases arise sporadically. However, several reports exist of family members and twins with the same diagnosis, suggesting some degree of genetic transmission. Poland's Syndrome is three times more common in boys than girls, and affects the right side of the body twice as often as the left. The reasons for these differences are unknown, as is the cause. We are presenting a case report of this rare entity in 12 year old male boy.

CASE REPORT

A 12 years old male child presented to us in the out- patient department with problem of hypoplasia and syndactaly of fingers of right hand. On further evaluation of the patient we found out that there was hypoplasia of the pectoralis major, latissmus dorsi and also the serratus anterior muscle on right side. There was also hypoplasia of nipple on right side as compared to left side. No lymph node was palpable. Blood biochemistry and counts were with-in normal limits. So the diagnosis of ‘Poland Syndrome’ was established. Since the major complaint of the patient was syndactly, so only that part was treated. The patient is with us on regular follow up.

Figure 1

Figure 1

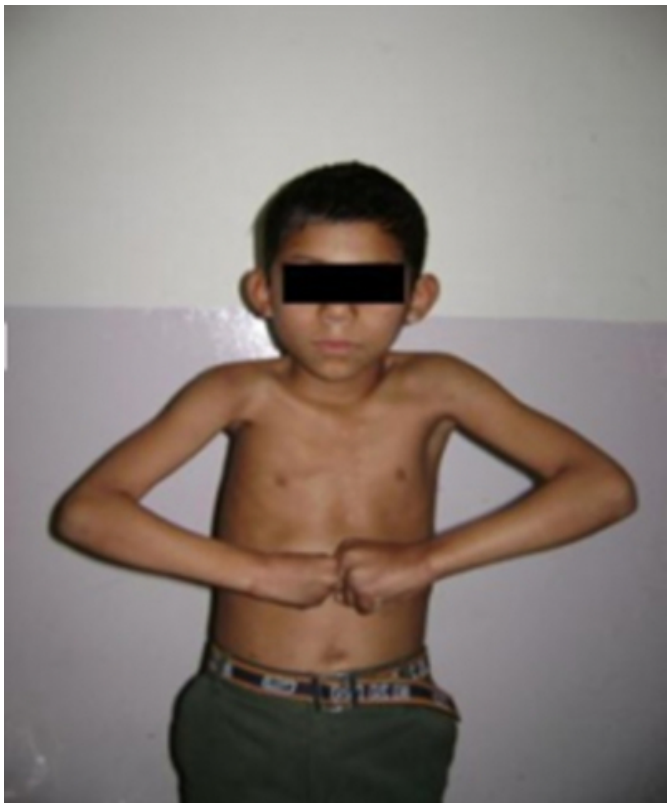

Figure 2

Figure 2

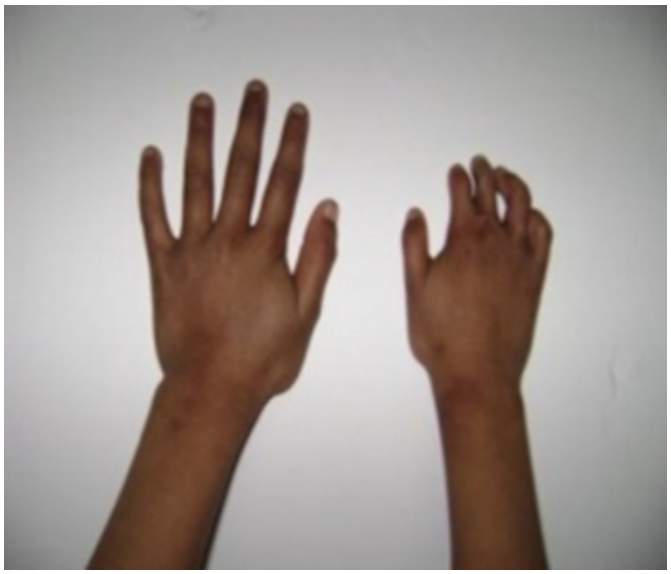

Figure 3

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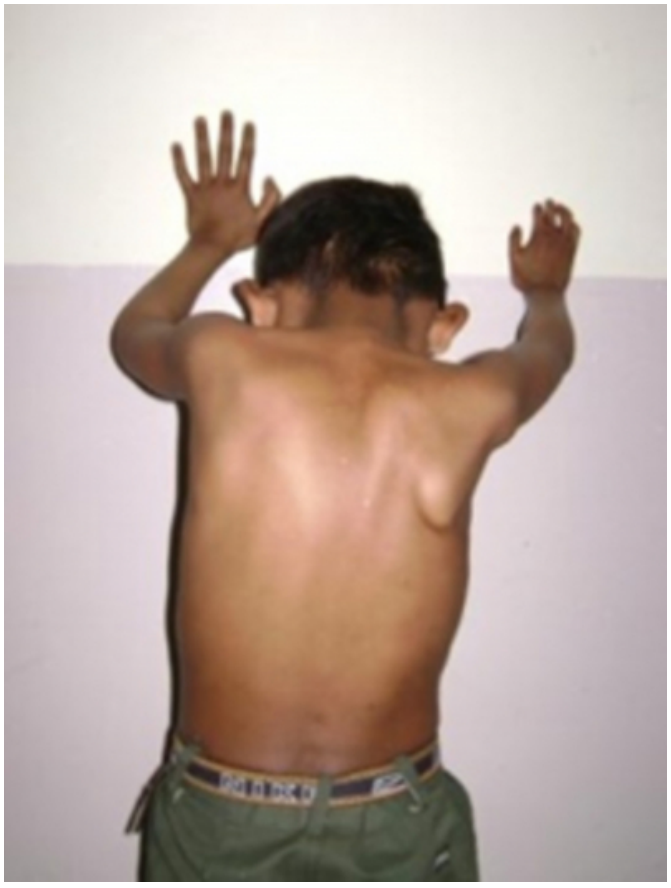
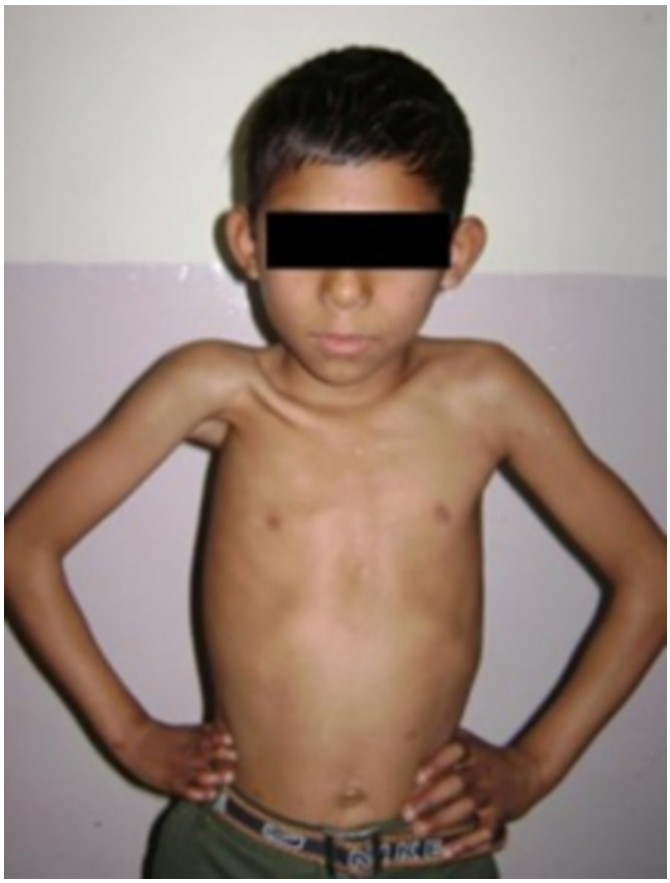


Figure 4

Figure 4



DISCUSSION

Very frequent signs of Poland syndrome are absent pectoral muscles, Brachydactyly (Short fingers), Diaphragmatic hernia, Humerus absent/abnormal, Oligodactyly/missing fingers, Radius absent/abnormal, Rhizomelic micromelia (relatively shorter proximal segment of the limbs compared to the middle and the distal segments),Syndactyly of fingers (webbing),Ulna absent/abnormal, Upper limb asymmetry ,Abnormal rib.

Frequent Signs are Hypoplastic/absent nipples, Scapula anomaly and Occasional Signs Agenesis/hypoplasia of kidneys, Encephalocele, Abnormal morphology and function of hypothalamic-hypophyseal axis, Microcephaly, Preaxial polydactyly, Ureteric anomalies (reflux/duplex system), Vertebral segmentation anomaly.

Although several theories have been advanced to explain the etiology of Poland syndrome, most evidence indicates that it results from a vascular event during the critical sixth week of gestation with hypoplasia of the subclavian artery causing musculoskeletal malformations. The critical vascular event, known as subclavian artery supply disruption sequence

(SASDS), occurs when the medial and forward growth of the ribs forces the subclavian vessel into a U-shaped configuration.

Poland syndrome has been associated with other syndromes including Möbius syndrome (congenital bilateral facial paralysis with inability to abduct the eyes) and Klippel-Feil syndrome. Hematopoietic malignancies, including leukemia and non-Hodgkin lymphoma, have been described in patients with Poland syndrome

Patients with Poland syndrome present for treatment of the chest deformity and breast asymmetry. Three main determinants influence the timing and options for reconstruction: breast development, existence of a latissimus dorsi muscle, and degree of chest wall deformity. If the breasts are not fully developed, use of autologous tissues for reconstruction is delayed until such time. During breast development, females may benefit from provisional breast reconstruction with tissue expansion. The hypoplastic breast can be expanded incrementally to match breast development on the unaffected side. Once the breasts are developed, the latissimus muscle can be used.

In males, the chest deformity can be reconstructed with the latissimus dorsi muscle as early as age 13 years. When rib abnormalities are mild, reconstruction with the latissimus dorsi muscle affords satisfactory chest wall symmetry. However, if severe, associated rib abnormalities should be treated to optimize the eventual outcome.

If the latissimus is absent as part of the Poland syndrome complex, other options for reconstruction include free latissimus muscle from the unaffected side, the transverse rectus abdominus muscle (TRAM) flap, gluteus maximus flap, thigh flap, or free perforator TRAM or gluteus flaps. TRAM should be contraindicated in the females who are intended to be pregnant.

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