2 Cases of Poland Syndrome

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ABSTRACT

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Poland's syndrome, (Poland's anomaly) is a rare birth defect characterized by underdevelopment or absence of the chest muscles (pectoralis) on one side of the body and webbing of the fingers (cutaneous syndactyly) of the ipsilateral hand mostly common on the right side of body and found more in males than females. It is usually considered a unilateral condition.

Because of the rarity of the disease and chance of missing it in routine clinical examination we are presenting two patients with Poland's syndrome seen in our clinic in the same month who were completely un aware of their anomalies

Keywords: Poland's syndrome, Poland's anomaly, Cutaneous syndactyly

*See End Note for complete author details

INTRODUCTION

In 1841, Sir Alfred Poland who was a medical student then described a characteristic chest wall anomaly based on findings of one cadaver dissection. He specifically noted absence of the sternocostal portion of the pectoralis major with an intact clavicular origin, absence of the pectoralis minor, hypoplastic serratus anterior and external oblique muscles.

Named after him, Poland syndrome also includes features of ipsilateral breast and nipple hypoplasia and aplasia, deficiency of sub cutaneous fat and axillary hair and also absence of the sternal head of the pectoralis major muscle, hypoplasia of the rib cage, and hypoplasia of the upper extremity.

Many of the patients are asymptomatic and unless a thorough physical examination is done these features are likely to be missed. We describe here 2 cases of Poland's syndrome seen in our clinic in the same month.

CLINICAL DETAILS

Case 1

44 year old male patient presented to our OPD with complaints of recurrent pain right side of chest, low back ache, neck pain for 3 years duration.

Patient also gives previous history of rib fracture (right side) 1 ½ years back after a fall.

On examination

General examination

Salient features noted in this patient were syndactyly of left hand, pectus excavatum.

There was also asymmetry of left side of chest with wasting of pectoralis major and minor muscles. Diminished chest expansion on the left side was also noted.

Vitals- Pulse rate-84/min BP –150/70 mmHg Resp rate -16/min

Systemic examination

CVS, RS-NAD Rest of systems examination-WNL

Investigations

CXR showed left hemi thorax translucent, due to absence of left pectoralis major and Scoliosis to left, old healed fracture 7,8,9 ribs anteriorly on right side.

Rest of parameters in other investigations was normal including USG Abdomen, which ruled out any renal anomalies (see Figure1)

Case 2

27 year old male patient presented to our OPD with complaints of lower abdominal pain and post micturition syncope at night.

He gave history of similar episode of loss of consciousness after micturition 5 years back.

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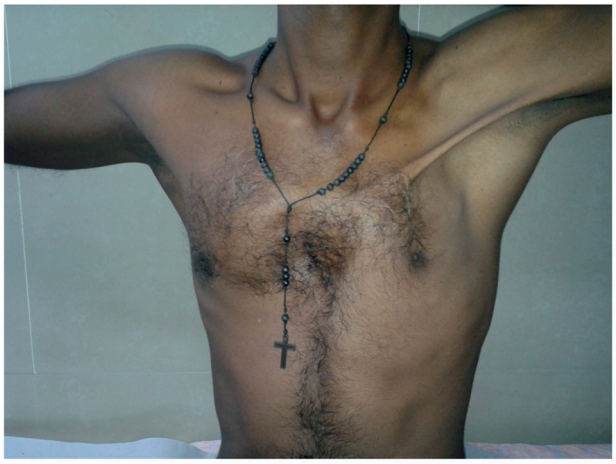

Figure 1. **Showing asymmetry of chestwall, wasting of pectoralis major, prominent skin fold on left side – Case 1**

On examination

General examination-

Salient features noted in this patient were syndactyly of left hand with grade 2 clubbing. There was also asymmetry of left side of body with wasting of pectoralis muscles, kyphoscoliosis of left side with prominence of clavicles on left side. The chest expansion was also decreased on left side.

Vitals- Pulse rate-78/min BP –140/70 mmHg Resp rate -18/min

Systemic examination

CVS, RS-NAD Rest of systems examination-WN (See Figure 2)

Investigations

CXR showed wasting of pectoralis major muscle and upward displacement of scapula

Rest of parameters in other investigations was normal including USG Abdomen ,which ruled out any renal anomalies.

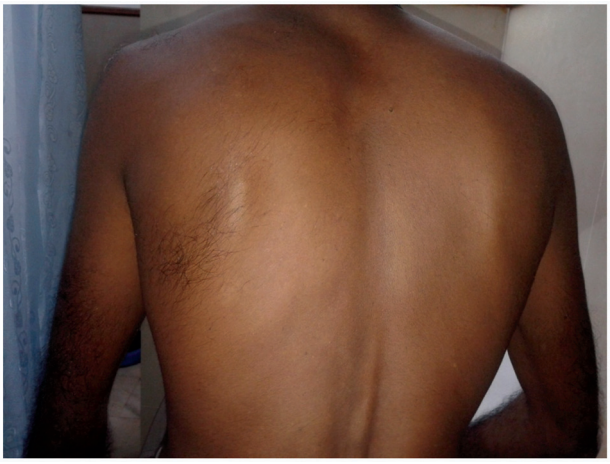

Figure 2. **Showing asymmetry of back on left side –Case 2**

DISCUSSION

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 alone may be seen. Involvement of adjacent muscles, including the pectoralis minor, serratus, latissimus dorsi, and external oblique, also has been described. Poland syndrome can also present with ipsilateral renal agenesis. Renal structural anomaly may be an integral part of this syndrome .Therefore ultrasound abdomen & renal imaging studies must be performed on all patients with Poland syndrome.

The skin of the affected area is thin and the axillary hair may be absent. The ipsilateral nipple is often smaller and higher in both males and female patients, and the breast is generally hypoplastic in female patients.

Skeletal deformities may involve absence of portions of the ribs or costal cartilages anteriorly. In severe cases, anterior lung herniation may be present. The scapula may be smaller with winging; this is termed Sprengel deformity. The upper extremity also may be hypoplastic. The upper arm, forearm, and fingers may be shortened; brachysymphalangism, Complete, or incomplete syndactyly can also be found in patients with Poland syndrome.

Because the functional disability in Poland syndrome is mild, patients usually present later for evaluation and discussion on aesthetic options. Chest wall abnormalities and the presence of the latissimus muscle may require evaluation with CT scan.

Poland syndrome is uncommon but not rare. While plastic surgeons encounter more female patients than male patients with this deformity (because the female patients seek out treatment of breast asymmetry), no gender predilection is exhibited. Many men remain undiagnosed. Since Poland syndrome is under reported and infrequently diagnosed, the exact incidence is difficult to determine. In one review, the incidence of Poland syndrome was reported as 1 in 30,000.[1] The right side is affected twice as often as the left.[2]

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 syndrome has been associated with other syndromes, including Möbius syndrome (congenital bilateral facial paralysis with inability to abduct the eyes) and Klippel-Feil syndrome.[3] Hematopoietic malignancies, including leukemia and

non-Hodgkin lymphoma, have been described in patients with Poland syndrome.

Although several theories have been advanced regarding the etiology of Poland syndrome, most evidence indicates that it results from a vascular developmental anomaly during the sixth week of gestation, with hypoplasia of the subclavian artery causing musculoskeletal malformations.

In male patients, 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. In females breast anomalies can be corrected by reconstruction surgery or with implant expander.

END NOTE

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