

Poland's syndrome: a case report and review of literature

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Abstract

Poland's Syndrome is a rare congenital condition. It is classically characterised by absence of unilateral chest wall muscles and sometimes ipsilateral symbrachydactyly (abnormally short and webbed fingers). The condition typically presents with unilateral absence of the sternal or breast bone portion of the pectoralis major muscle which may or may not be associated with the absence of nearby musculoskeletal structures. Most of the Poland Syndrome is sporadic. We report a 19-year-old patient with variant of Poland Syndrome. To the best of our knowledge, this is one of the few documented cases of a patient with Poland Syndrome reported from Bangladesh.

Keywords: Poland's syndrome.

Introduction

The Poland's Syndrome (also Poland syndrome, Poland's syndactyly, Poland sequence, and Poland's anomaly) (PS) was first described in 1841 by Sir Alfred Poland. In 1962, a New Zealand-born British plastic surgeon, Patrick Clarkson, coined the term Poland syndactyly. Baudinne et al (1967) reported a case of Poland's Syndrome, a term that is more accurate because the group of anomalies include more than just syndactyly.¹

PS cases present with absence or under-development of pectoralis major muscle, associated in some cases with or without a hypoplasia of the breast, an agenesis of 2,3,4 and 5 ipsilateral costal cartilage, an athelia, and an ipsilateral webbing of the fingers (cutaneous syndactyly).^{2,3} Therefore, PS may occur with different gravity. Currently, it is assumed that PS is characterised by a missing sternocostal bundle of the pectoralis major muscle.⁴

Its incidence is difficult to determine, but current estimates range between 1:7,000 and 1:100,000 births, with higher frequency among males (ratio: 2:1-3:1). In 75% of the cases, it is located on the right hemithorax in the unilateral form.^{2,4-7} Affected individuals may have

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variable associated features, such as under-development or absence of one nipple including the areola and/or patchy absence of hair in the axilla.^{2,5} In females, there may be under-development or aplasia of one breast and underlying (subcutaneous) tissues.⁸ In some cases, associated skeletal abnormalities may also be present, such as under-development or absence of upper ribs; elevation of the shoulder blade (Sprengel deformity); and/or shortening of the arm, with under-development of the forearm bones (i.e., ulna and radius).^{2,5} Rare association includes dextrocardia, renal hypoplasia, leukaemia and Mobius syndrome.³

We report a 19-year-old patient with variant of Poland Syndrome. To the best of our knowledge, this is one of the few documented cases of a PS patient reported from Bangladesh.

Case Report

A 19-years-old boy, farmer, presented in June 2013 with a flattening of the right anterior chest wall since birth. He wanted to know whether or not he is fit for going abroad as a worker. He was the second child in a family of four. There was no familial disorder and specifically a coagulation disorder. He had no physical complaints. Examination revealed a young man with normal growth parameters and good mental status. There was no abnormal cranio-facial finding. His chest wall was asymmetric with the hypoplasia of right side (Figure-1). There was no oligosyndactyly (OS). He had flattening of right chest wall with absence of breast. The pectoralis major muscle was under-developed but the pectoralis minor muscle as well as the anterior serratus muscle were present (Figure-2). The movements of the right shoulder were possible with the muscle power at adduction estimated at 4/5, otherwise normal muscle power of arm and hand on the right side was seen. Opposite arm and hand muscle power was 5/5. He had normal heart sounds, and respiration and breath pattern were normal as well. The limbs were normal and symmetric, and all fingers were found to be normal. Radiological examination of the chest showed no abnormalities of the ribs or heart. X-ray of bones of the ipsilateral upper limb did not show any abnormality. Abdominal ultrasound showed no abnormality. Based on physical findings, a diagnosis of PS was made. No surgical treatment was offered, and the



Figure-1: Underdeveloped of right pectoralis major muscle.
(Source: Personal collection from chamber)

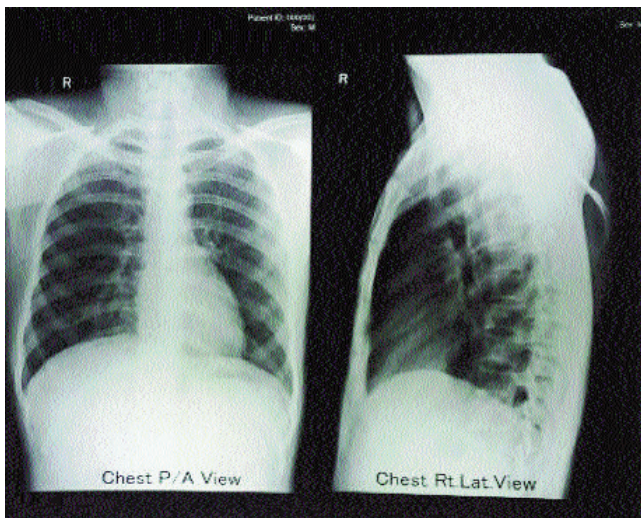


Figure-2: Chest X-Ray of the subject (P/A and Lateral View).
(Source: Personal collection from chamber)

patient and his family were counselled properly and he was given clearance to go abroad as a worker.

Discussion

The PS case presented here is one of the few cases described from Bangladesh and had peculiar presentation as it involved only the unilateral aplasia of the pectoralis major muscle without any other associated defects.^{2,4} It presented in the right hemithorax as the right side is the most frequently involved.^{4,5} The exact aetiology of PS is unknown. It is assumed that the aplasia of the pectoralis muscles and associated chest defects, as the athelia, aplasia of costal cartilages, are consequences of an interruption of early embryonic blood supply of subclavicular artery branches.^{6,7} A combination of the blockage of various branches could lead to different manifestations of PS. It is known that thoracic wall is supplied by medial thoracic branches, intercostals artery, and the thoracic artery from axillary artery, the thoracoacromial artery and the lateral thoracic artery. All these branches come from the subscapular artery or axillary artery. The interruption of the blood supply is caused by thrombus or embolus, which prevent the blood from reaching the developing tissue. Another cause of blood supply interruption is the maldevelopment of vessels.

However, there have been case reports of PS associated with unusual defects, which cannot be explained on the basis of compromised blood supply alone. On the other hand, a study⁴ described an unusual presentation of Poland's anomaly without any vascular alteration, raising the question as to the true pathogenesis of PS.

Most cases of PS are sporadic, which means they are not inherited and occur in people with no history of the disorder in their families. Rarely, this condition is passed through generations in families. In these families the condition appears to be inherited in an autosomal dominant pattern, which means one copy of an altered gene in each cell is sufficient to cause the disorder, although no associated genes have been found. There are rare instances where more than one individual has been identified with PS either in the immediate^{7,10,11} or extended family.¹⁰ Therefore, some authors believe that an inherited abnormal vasculature formation may be the central underlying mechanism for this condition. Several reconstructive procedures are available to correct the functional and structural deformities associated with this syndrome. As for the chest deformity, customised silicone prosthesis is simply and safely used. Transposition of the latissimus dorsi muscle for soft-tissue reconstruction has been used by many authors with satisfactory aesthetic and functional results.⁹

Conclusion

The paper adds to the knowledge of health professionals

in the region about this rare congenital condition.

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