Case Report

A rare case report of Poland syndrome in neonates

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Abstract

The Poland's anomaly was first described in 1841 by Sir Alfred Poland as a syndrome presenting with absence or underdevelopment of pectoralis major muscle, associated in some cases with a hypoplasia of the breast, an agenesis of 2, 3, 4 and 5 ipsilateral costal cartilages, and an ipsilateral webbing of the fingers (cutaneous syndactyly). Other associated abnormalities may include dextrocardia, diaphragmatic hernia and renal anomalies etc. Poland syndrome most often affects the right side of the body, and occurs more often in males than in females. It is usually considered a unilateral condition but rarely, bilateral.

Introduction

It was first named in 1962 by Patrick Clarkson, a New Zealand-born British plastic surgeon. The incidence of this condition ranges from 1: 20,000 to 1:50,000 live births as reported by different authors [1, 2].

The cause of Poland syndrome is unknown. However, an interruption of the embryonic blood supply to the arteries that lie under the collarbone (subclavian arteries) at about the 46th day of embryonic development is the prevailing theory. Although several theories of etiogenesis have been proposed, the vascular theory appears to be the most favoured by many [3].

Case Report

A newborn, preterm (34 wks POG) male presented at Subharti medical college at 6 hours of life born via LSCS (cause leaking PV since 2 days and oligohydramnios) for ongoing care of prematurity. Baby cried immediately after birth and no signs of distress present. The baby was first issue of non consanguineous marriage. No significant Natal history present except PV leaking.

On examination pulse was 136 per minute, BP 44/26(34) mm of hg, Length 48 cms, weight 1.810 kgs and Head circumference 35cms were noted. There was some abnormality of right side of chest with asymmetrical hypoplasia and absence of anterior axillary fold [Figure 1]. Ipsilateral fingers were short and webbed (symbrachydactyly) [Figure 2]. No systemic abnormality was noted.

Xray chest showed no abnormalities of the ribs or heart but increased translucency over the ride side due to the absence of pectoralis major muscle [Figure 3] and X-ray of the affected limb showed aplasia of the 1st to 4 th fingers. Axial computed tomography scan showed the presence of pectoralis major on the left side but not on the Right [Figure 4].

Discussion

The case of Poland Syndrome we present is the first described in Rwanda and is of the pure presentation as it consists only on the unilateral aplasia of the pectoralis major muscle without any other associated defects [1,4]. The cause of Poland syndrome is uncertain and it often occurs sporadically. The disorder is currently considered “a non-specific developmental field defect” occurring at about the 6th week of fetal development.

Geneticists currently hold the view that Poland syndrome is rarely inherited and generally is a sporadic event. There are rare instances where more than one

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individual has been identified with Poland syndrome either in the immediate [5, 6, 7] or extended family. [8, 9, 10] Therefore, some authors believe that an inherited abnormal vasculature formation may be the central underlying mechanism for this condition.

Fig 1: Showing some abnormality of right side of chest with asymmetrical hypoplasia and absence of anterior axillary fold

Fig 2: Xray showing ipsilateral fingers were short and webbed

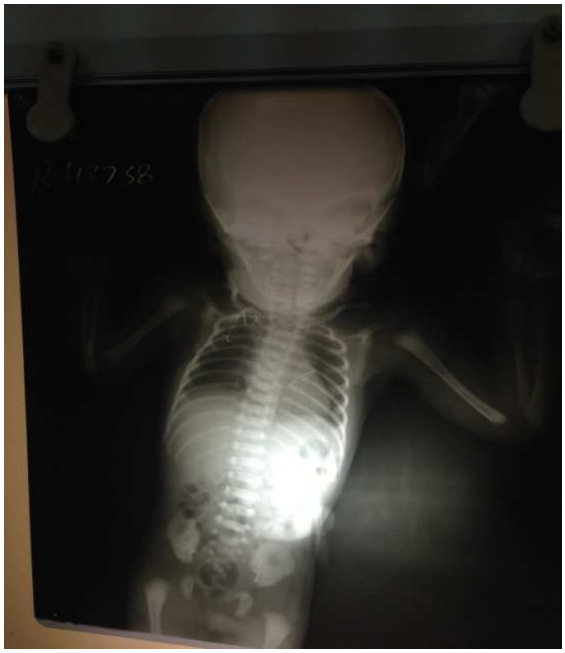

Fig 3: Xray showing increased translucency over the ride side

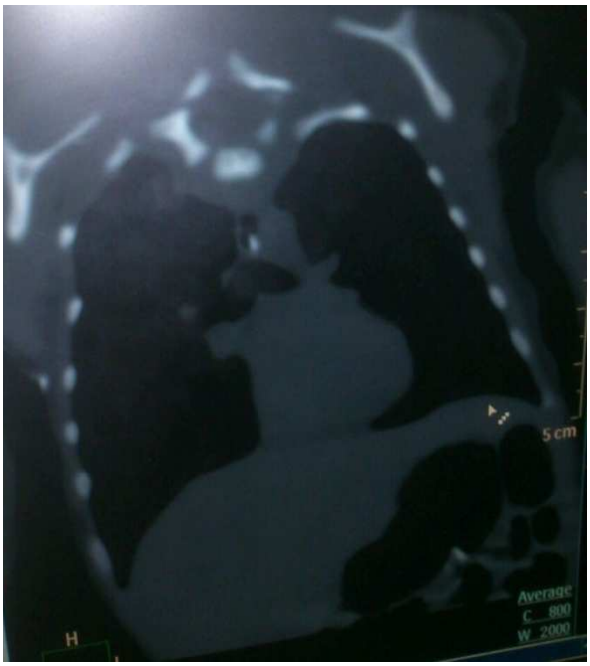

Fig 4: Showing presence of pectoralis major on the left side and absent on right side.

Conclusion

It is a rare inherited disorder with aplasia of pectoralis major muscle on either side with or without associated disorders. A team approach is required for management of patient with Poland syndrome. Several reconstructive procedures are available to correct the functional and

structural deformities associated with this syndrome. As for the chest deformity, customized 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 esthetic and functional results [11].

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