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Case report:

Poland syndrome with dextrocardia and congenital heart disease: A case report.

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Abstract

The Poland syndrome is a rare anomaly. It consists of unilateral absence or hypoplasia of the pectoralis muscle, most frequently involving the sternocostal portion and a variable degree of ipsilateral hand and digit anomalies. The combination of Poland syndrome and dextrocardia is uncommon. We describe a case of Poland syndrome with dextrocardia and Patent ductus arteriosus.

Key words – dextrocardia, patent ductus arteriosus, rib hypoplasia, pectoralis major

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Introduction

Poland syndrome is a rare anomaly, also known as hand and ipsilateral thorax syndrome, limb/body-wall defect, fissure thoracis lateralis, acropectoral- renal field defect, or subclavian artery supply disruption sequence.Poland syndrome consists of unilateral absence or hypoplasia of the pectoralis muscle, most frequently involving the sternocostal portion and a variable degree of ipsilateral hand and digit anomalies [1]. Other usual anomalies in Poland Syndrome are malformations of the anterior chest wall and breast. Dextrocardia, lung herniation, renal, vertebral and lower limb malformations have been described in rare cases. The exact etiology of Poland's syndrome is unknown. It probably results due to interruption of early embryonic blood supply to subclavian arteries, the vertebral arteries and or their branches [2]. Poland syndrome affects the right side in more than two third of cases [3].It is more common in males than females. The combination of Poland syndrome and dextrocardia is uncommon. Patent ductus arteriosus (PDA) in Poland syndrome is not reported.

Case presentation

A term male baby weighing 2750 gms, born by spontaneous vaginal delivery at 39 weeks of gestation after normal antennal period. Child was noticed to have left sided depressed hemithorax with loss of normal chest contour, areolar hypoplasia and widened 3rd intercostal space due to hypoplasia of left pectoral muscle

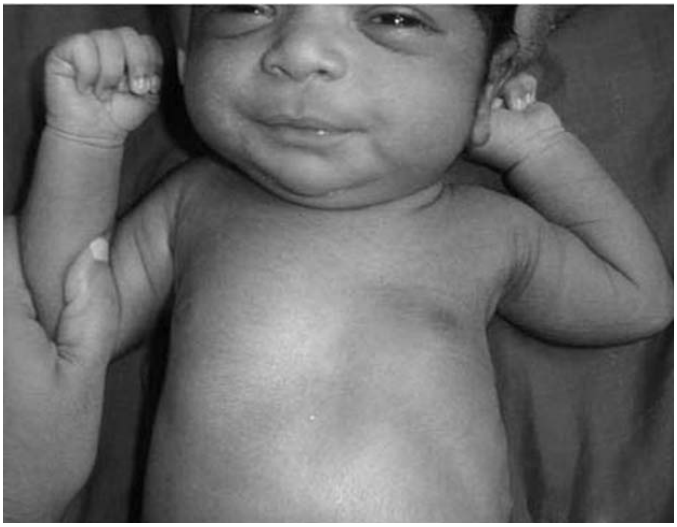

Figure 1. *Shows depressed left chest wall and hypoplasia of left pectoral muscle*

(Fig. 1). On examination heart sounds were well heard on the right side, liver 2 cm below the right costal margin. A chest X-ray showed dextrocardia with rib hypoplasia *(*Fig. 2). Echocardiography showed dextrocardia, Patent ductus arteriosus with left to right shunt. Abdominal ultrasound confirmed right sided liver and normal kidneys on both sides. There were no digital anomalies or features suggestive of Möbius syndrome. The karyotype was normal. The baby was otherwise healthy and being followed up.

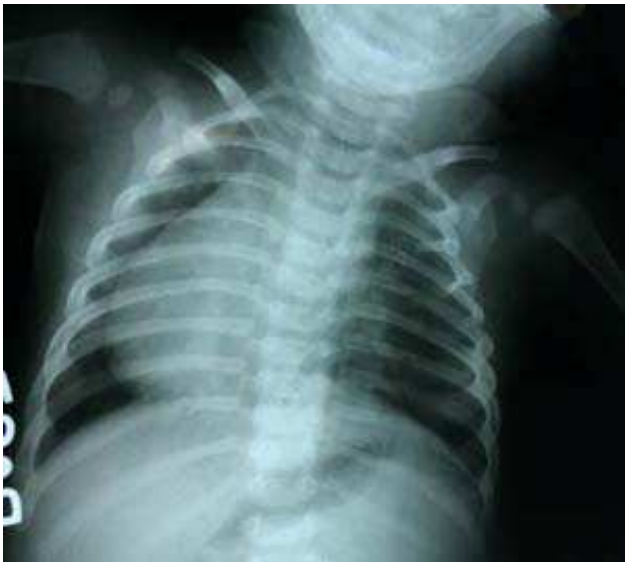

Figure 2. *Chest X-ray showing dextrocardia and left sided rib defect.*

Discussion

Alfred Poland in 1841 described a man who had syndactyly, absence of the middle phalanges and muscular anomalies of the ipsilateral chest wall. In 1962, Clarkson coined the name Poland's syndactyly [4]. Although the disease pathology underlying Poland syndrome is not well understood, various hypotheses have been put forward. Most widely accepted hypothesis is that, at the end of the sixth week of gestation, when the upper limb bud adjacent to the chest wall is still in a stage of development, the interruption of the embryonic blood supply causes hypoplasia of the ipsilateral subclavian artery or one of its branches[5].

Hypoplasia of the internal thoracic artery could cause the absence of the sternocostal portion of the pectoralis major muscle, whereas hypoplasia of the brachial artery may lead to hand abnormalities[6]. Another hypothesis is that disruption of the lateral plate mesoderm (from which the pectoralis muscle develops) between 16 and 28 days after fertilization may account for all the defects[7]. Geneticists currently hold the view that Poland syndrome is rarely inherited and generally a sporadic event. There are rare instances where more than one individual has been identified with Poland syndrome in the same family[8].

Poland's syndrome is associated with the absence of the sternocostal head of the pectoralis major muscle. In most of the cases, the pectoralis minor muscle is also absent. In some cases of Poland's syndrome, the latissimus dorsi, external oblique, and serratus anterior muscles are also affected. Breast involvement may vary from mild hypoplasia to complete absence [9]. The nipple and areola are usually hypoplastic and elevated, lightly pigmented, or even absent. Ribs II to IV, or III to V are most commonly involved, but the second rib is less frequently affected. Rib defects are present in 15% of patients with right-sided pectoralis major defects [10]. The progression of the chest deformity occurs mostly during growth periods. A rare association between Poland syndrome associated with microcephaly, cerebral atrophy, disorders in myelination, situs inversus or dextrocardia, hemivertebra, gastroschisis, paralysis of the cranial nerve, psychosocial retardation, hypospadias, and urinary system anomalies have also been reported[11]. Cases of Poland's syndrome have been known to be associated with leukemia, non-Hodgkin's lymphoma, cervical cancer, leiosarcoma, and lung cancer[12].

Isolated dextrocardia (situssolitus) represents an anomaly with normal situs but a right-sided heart. The frequency of isolated dextrocardia is about 1 in 30,000 live births[13]. There are only about twenty reported cases of Poland syndrome with Dextrocardia. In all these patients, Poland's syndrome was left-sided and associated with rib defects [14]. Congenital cardiovascular anomalies have not been reported in Poland syndrome with dextrocardia. Our case was associated with patent ductus arteriosus. All the cases of Poland syndrome with dextrocardia had rib defects.

Patients with significant deformities of the chest wall and overlying soft tissue may need surgical reconstruction, generally recommended after the completion of growth. Several reconstructive procedures are available to correct the functional and structural deformities associated with this syndrome. As for the chest deformity, customized silicone prosthesis can be used. Transposition of the latissimus dorsi muscle for soft-tissue reconstruction has been used with satisfactory results [15].

Conclusion

Poland syndrome with dextrocardia can be associated with other congenital heart diseases. During clinical examination and echocardiography one should evaluate carefully for the other anomalies.

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