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Case Report

Poland Syndrome-A Typical Presentation

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1. Abstract

Poland's syndrome is a rare congenital disorder with unilateral aplasia of the pectoralis major muscle (PMM) associated with other anomalies like ipsilateral synbrachydactyly. We present a female baby with Left side congenital absence of pectoralis major muscle with normal hands at birth which can be a milder form of Poland's syndrome in contrast to the commonly reported right sided Poland Syndrome with male predisposition.

2. Keywords

Poland's syndrome, Limb anomalies, Skeletal abnormalities, Subclavian artery, Aplasia pectoralis major, Synbrachydactyly

3. Case Report

Poland Syndrome is a rare congenital anomaly characterized by unilateral absence (aplasia) of chest wall muscles and abnormally short, webbed fingers (symbrachydactyly) of the hand on the same side (ipsilateral). It occurs in 1:30,000 live births and it was named after Sir Alfred Poland, who first published it in 1841 [1]. In this condition, there is typically absence of sternocostal part of pectoralis major, unilateral absence of the pectoralis minor and the sternum. Affected individuals may have variable associated features, such as underdevelopment or absence of one nipple (including the areola) and/or patchy absence of hair in axilla. Poland Syndrome affects males more commonly than females and most frequently involves the right side of the body. In females, there may be underdevelopment or absence of one breast and subcutaneous tissues. Other clinical manifestations include skeletal abnormalities like underdevelopment or absence of upper ribs, elevation of the shoulder blade and/or shortening of the arm, with underdevelopment of the ulna and radius [2].

Here we present a case of a newborn with the classical features of Poland syndrome, and with no limb deformities.

We report a term female neonate appropriate for gestational age born by spontaneous vaginal delivery to a non-consanguineous parent at 37 weeks weighing 2740 gms with Apgar of 8 and 9 at 1st and 5th minute of birth. Her head circumference was 33cms and length was 51cms. She did not develop any respiratory distress; hence feeds were initiated within 30 minutes of birth. During routine head to toe examination left anterior chest wall deformity (**Figure 1**) was noted with displaced and hypoplastic nipple left nipple (**Figure 2**) along with a fibrous strand of tissue running from the lateral end of clavicle when the left arm is raised passively (**Figure3**). There were no hand or feet abnormality of the same side. Hairs and nails were normal. Rest of the examination was completely normal. Neurologic and cardiovascular examinations were unremarkable. Infantogram was also within normal limits. Ophthalmological evaluation was also insignificant. There were no other anomalies seen on further systemic evaluation. Skeletal survey was normal.

PS is usually sporadic and rarely is it inherited. The exact etiology is unknown. At 6th week of gestation, the pectoral mass splits into a clavicular head and a costal head, and this is also the time when tissues between the digits of the hand start to disappear to form web spaces. It is proposed that an insult during the 6th week of gestation may be responsible for anomalies in the-pectoralis muscle and hand anomalies [3]. Poland's syndrome with its unknown causes and lots of proposed theories, one of the other reasonable explanation is interruption to the blood supply to the subclavian arteries in 6th week of gestation, also called as Subclavian artery supply disruption sequence (SASDS) has been postulated elsewhere very commonly. Another hypothesis suggested

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disruption of the lateral embryonic plate mesoderm between 16 and 28 days after fertilization, which may account for all the defects [4]. PS is also associated with other syndromes, including Mobius syndrome (congenital bilateral facial paralysis with an inability to abduct the eyes) and Klippel-Feil syndrome. Lots of associations have been described earlier with leukemia, lymphoma, carcinoma of hypoplastic breast, neuroblastoma, renal hypoplasia, agenesis, duplication of renal collecting system, dextrocardia out of which Ispilateral limb involvement is one of the most common signs of Poland's syndrome[5].Al-Qattan MM in the year 2001 reviewed 20 cases of Poland's syndrome and offered a classification of the hand anomalies. The hand anomalies are divided into seven types according to the severity of the deformity. Three adult female patients with hypoplasia of the sternocostal head of the pectoralis major muscle and ipsilateral breast hypoplasia and the hand in these three patients were normal (type 1). Some authors proposed that isolated pectoral hypoplasia should not be considered as Poland's syndrome. However, is not valid because in familial Poland's syndrome one family member may show an isolated pectoral hypoplasia and another member may show the combined hand and pectoral deformity and this clearly indicates the variable expression of the syndrome[6].According to our report of a female baby with left sided involvement in contrast to the right side involvement in males, we conclude that the absence of limb deformities, in the presence of abnormal pectoralis muscle, does not rule out PS. Also advised for regular follow up for oncological screening and reconstructive surgery if needed at earlier ages[7].

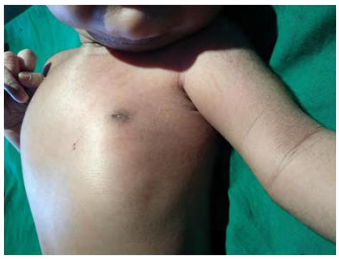

Figure 1

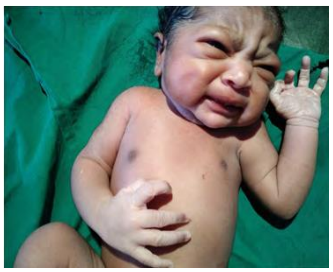

Figure 2

Figure 3

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