University Journal of Surgery and Surgical Specialities

ISSN 2455-2860 2019, Vol. 5(9)

Poland syndrome a case report

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Abstract : Poland syndrome is a rare anomaly. It is classically characterized by absence or hypoplasia of chest wall muscles on one side of the body. Ipsilateral hand and digit anomalies and absence or hypoplasia of breast are other associated features. Here we present a case of Poland syndrome for its rarity.

Keyword : Poland syndrome, Pectoralis major, absence of breast nodule.

Introduction:

Poland syndrome also known as Poland anomaly, Poland sequence, Poland Syndactyly, Hand and Ipsilateral Thorax Syndrome. Poland syndrome affects right side more than left side in two thirds of cases. It is 75 percentage more common in males than females. Incidence of Poland syndrome is 1 in 7000 to 1 in 100,000. Poland syndrome consists of unilateral hypoplasia or absence of pectoralis major muscle most frequently sternocostal portion and a variable degree of ipsilateral hand and digit anomalies. Other associated anomalies are malformations of ipsilateral anterior chest wall and breast. Rare malformations include dextrocardia, lung herniation, renal, vertebral and lower limb malformations.[1] The exact etiology of Poland syndrome is unknown. It probably results due to interruption of early embryonic blood supply to subclavian arteries, vertebral arteries and their branches.

Case presentation:

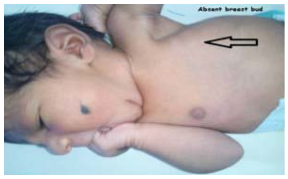
A term female neonate delivered labour naturale in a Primary Health Centre was referred for chest deformity. The baby born at 39 weeks of gestation, with a birth weight of 2800 Gms and cried at birth. She was a second order child born to non consanguinous parents. There was no similar illness in the family and elder sibling ,a male child was normal . Antenatal period was uneventful and fetal sonography was normal.

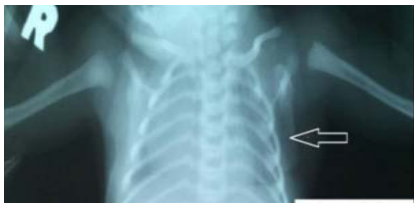
On examination:

Baby was appropriate for gestational age. Baby was alert and active. Chest asymmetry noted with wasting on the left side. There was no breast nodule, nipple, areola on the left side and anterior axillary fold was thinned out. The right side chest examination was normal. No other obvious external anomalies noted and the System examination was normal. Baby was taking breast feeds since birth. Chest x- ray AP view showed hyperlucenct left chest wall. Echocardiography and Ultrasonogram abdomen was normal..

Follow up:

Baby was on regular follow up till one month of age and was doing well. After one month baby had aspiration pneumonia after native medication in home and was admitted in a nearby Government Hospital for two days and was on treatment. Baby died due to sepsis.





X ray showing hyperleucency of left side of the chest

Discussion:

Alfred Poland in 1841 first described this condition by doing autopsy in a man who has difficulty in crossing limb across the chest wall and found the absence of sternocostal head of pectoralis major muscle. There was associated hand anomalies in that patient. Later in 1962 Patrick Clarkson coined the name Poland syndactyly and he reported 3 cases and named it as a syndrome after Poland.[4] The etiology of Poland syndrome is unclear. No genetic predisposition is still identified and Poland syndrome occurs sporadically .Very rarely more than one

individual in the same family is affected. Various hypothesis have been put forward. Most widely accepted hypothesis is that, at the end of sixth week of gestation, when upper limb bud adjacent to chest wall is still in a stage of development, the interruption of embryonic blood supply causes hypoplasia of ipsilateral subclavian artery or one of its branches.[5,8] Hypoplasia of internal thoracic artery could cause absence of sternocostal portion of pectoralis major muscle; whereas hypoplasia of brachial artery may lead to hand abnormalities. Another hypothesis is that disruption of lateral plate mesoderm (from which pectoralis major muscle develops) between 16 to 28 days after fertilization may account for all the defects.

Poland syndrome is associated with absence of sternocostal head of pectoralis major muscle in all cases. [1,2]In some cases pectoralis minor muscle ,latissimus dorsi, external oblique, serratus anterior muscles are also affected. Breast involvement may vary from mild hypoplasia to complete absence. The nipple and areola are usually hypoplastic and lightly pigmented or even absent.. Rib defects are present in 15% of the patient with right sided pectoralis major defects and ribs II to IV or III to V are commonly involved. The chest wall deformities may be evident mostly during growth periods. Other systems involved are gastrointestinal tract, renal (agenesis or hypoplasia of kidneys), ureteric anamolies vertebral segmentation anamolies, abnormal humerus and radius, scapular anomaly and sparsity of axillary hair on affected side. Occasional signs include Encephalocoele /exencephaly, microcephaly, diaphragmatic hernia and dextrocardia. [3,4]Cases of Poland syndrome have been known to be associated with leukemia, Non Hodgkin's lymphoma, cervical carcinoma, and lung carcinoma.[4,5] Diagnosis of Poland syndrome is by the characteristic clinical picture. chest x- ray may show hyper lucent hemi thorax and CT chest may delineate the extent of involvement of muscles. Management usually depends on the extent of the disease. Children with hand abnormalities need plastic surgical correction. Adolescent female with hypoplastic or absent breast needs breast reconstruction for cosmetic purposes. All children with Poland Syndrome should be screened for malignancies frequently. Prognosis of Poland syndrome is excellent. [16] Our neonate was a female child, and she had left side involvement which is a less common presentation. But there are no associated anomalies.

Take home message:

All children with chest wall asymmetry and absent breast bud should be suspected to have Poland syndrome and should be screened for associated anomalies. Poland Syndrome is not familial. No clear genetic predisposition is identified. Prognosis is usually excellent.

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