ORIGINAL RESEARCH PAPER
Volume-8 | Issue-10 | October - 2019 | PRINT ISSN No. 2277 - 8179 | DOI : 10.36106/ijsr
INTERNATIONAL JOURNAL OF SCIENTIFIC RESEARCH

POLAND SYNDROME,CONGENITAL DEFECT OF THE CHEST WALL:A CASE REPORT

Medicine

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ABSTRACT

POLAND SYNDROME is rare disorder characterised by underdeveloped/ absent chest muscle on one side of body. Incidence, l in 10,000 to 1 in 1,00,000.The authors describe a case of 65years old man, with history of breathlessness, on clinical examination of head to toe revealed defect of chest wall on left side. The authors presents a variant of poland's syndrome to emphasize the risk of trauma to underlying cardia, may impair significant movement on one side of body, as condition is asymptomatic and often not diagnosed & psychological toll if diagnosed in teenagers .This is few of documented cases of poland's syndrome from india.

KEYWORDS

Poland Syndrome,chest Wall,sporadic.

INTRODUCTION-

" Poland's syndrome is a rare congenital anomaly characterized by hypoplasia of the breast and nipple, scarcity of subcutaneous tissue, absence of the costosternal portion of the pectoralis major muscle, lack of the pectoralis minor muscle, aplasia or deformity of the costal cartilages or ribs II to IV or III to V, alopecia of the axillary and mammary region, and unilateral brachysyndactyly" [1].

"The extent and involvement of these components are variable and it is rare for all features to be present in the same individual" [2, 3]. "There is also a lack of correlation between the extent of hand, breast, and thoracic deformities" [4, 5]. "Poland's syndrome is mostly, if not always, unilateral, although a single bilateral case has been reported" [6].

"Clinical manifestations of Poland's syndrome are extremely variable" [3]. Unknown cause. Always sporadic ,tends to occur on right side. Common in boys than girls. Treatment surgical correction of chest deformities.

PRESENTATION OF CASE:

Patient admitted in critical care ward IPNO-6453.A 65 year old male patient presented with breathlessness.He had no family or personal history of pathological relevance.He was chronic bidi cigarettes smoker for 10years.He was not suffering from koch's,asthma,IHD.

On physical examination the patient was alert and oriented but defect of chest wall on left side (FIG1 & 2).Defect extended from 2nd to 5th ribs associated with absence of axillary hairs & folds on left side.Complain of breathlessness so the patient underwent for ECG(FIG 3) shows 'p' pulmonale in leads 2,3,avf.

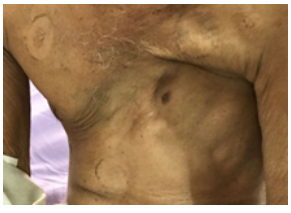

FIG 1:ANTERIOR VIEW OF CHEST WALL.

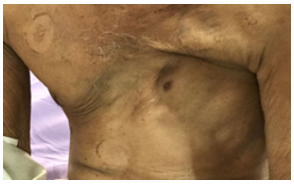

FIG 2:LATERAL VIEW OF CHEST WALL WITH ABSENCE OF LEFT AXILLARY HAIRS

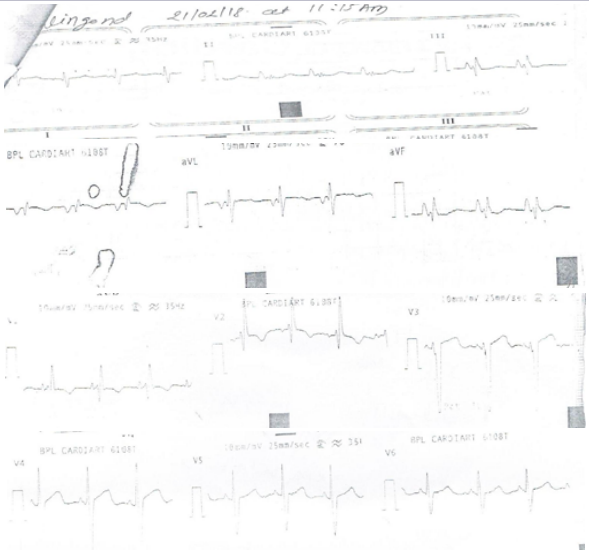

FIG 3:ECG changes of the patient.

CHEST XRAY-

A chest x-ray was performed in which hyperlucency on left side and dextrocardia(FIG 4).

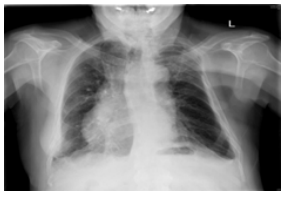

FIG 4:CHEST XRAY

OTHER INVESTIGATIONS:

TC-5940celIs/cmm,RBC 5.09millions/cmm,hb-14.4gm/dl,pIatelet count 1.491akhs/cmm,RBS-152mg/dI,UREA- 56mg/dl,serum creatinine-O.7mg/dI,serum sodium-134mg/dI,serum potassium3.6mg/dl.

CLINICAL DIAGNOSIS OF POLAND SYNDROME ESTABLISHED.

DISCUSSION-

POLAND'S SYNDROME first described by Alferd POLAND in 1841.Sporadic disorder due to vascular abnormal event occur during sixth week of fetal development called subclavian artery supply disruption sequence.

Incidence,1 in 10,000 to 1 in 1,00,000.75percent affects right side.symptoms occur on one side of body absence of chest muscles pectoralis major (most common) and minor,axillary hair absent on same side[7].

In sporadic cases of Poland's syndrome, there is a male predominance; also the right side is affected more than twice as often in males, whereas in females, the number of left- and right-side lesions is almost equal. incidence of Poland's syndrome in males and females is approximately the same, and there is no right-side predominance.

"Poland's syndrome, absence of the upper portion of the serratus anterior, which has been attributed to the decrease of blood flow in the suprascapular arteries, leads to elevation and winging of the scapula (Sprengel's deformity or scapula alata)" [8,9].

"Poland's syndrome may also coexist with Klippel-Feil syndrome, which is characterized by shortness of the neck. The latter is attributed to fusion of the cervical vertebrae and to abnormalities of the brainstem and cerebellum, due to a delay in the development of the vertebral arteries" [8].

" The association of aplasia of the pectoralis major muscle with renal anomalies, such as unilateral renal agenesis or duplication of the urinary collecting system, is defined as an acro-pectoral-renal field defect, and may cause renal hypertension [10, 11]". For this reason, renal studies are recommended for all patients with aplasia of the pectoralis major muscle.

DIFFERENTIAL DIAGNOSIS-

Amazia and Amastia.

TREATMENT:

RECONSTRUCTIVE SURGERY.

"Surgical intervention may be indicated for the following reasons:

(1) unilateral depression of the chest wall and the possibility of its progression,
(2) lack of adequate protection of the heart and lung,
(3) paradoxical movement of the chest wall,
(4) hypoplasia or aplasia of the female breast, and
(5) cosmetic defect due to lack of the pectoralis major muscle and axillary fold in male patients[12]".

CONCLUSION -

Condition is benign and asymptomatic ,severity varies so often not diagnosed or reported .Reconstructive surgery viable option.

REFERENCES:

[1]. Ravitch MM. Poland's syndrome. In: Ravitch MM, ed. Congenital deformities of the chest wall and their operative correction. Philadelphia, London, Toronto: WB Saunders, 1977:233–71.
[2]. Urschel HC Jr, Byrd HS, Sethi SM, Razzuk MA. Poland's syndrome: improved surgical management. Ann Thorac Surg 1984;37:204–11.
[3]. Urschel HC. Poland's syndrome. Chest Surg Clin N Am 2000;10:393–403.
[4]. David TJ. The Poland anomaly and allied disorders. Pediatr Res 1981;15:1184.
[5]. Shamberger RC, Welch KJ, Upton J III. Surgical treatment of thoracic deformity in Poland's syndrome. J Pediatr Surg 1989;24:760–5.
[6]. Karnak I, Tanyel FC, Tuncbilek E, U¨ nsal M, Bu¨ yu¨ kpamukcu N. Brief clinical report. Bilateral Poland anomaly. Am J Med Genet 1998;75:505–7.
[7]. rarediseases.info.nih.gov/diseases/7412/poland-syndrome.
[8]. Bavinck JNB, Weaver DD. Subclavian artery supply disruption sequence: hypothesis of a vascular etiology for Poland, Klippel-Feil, and Mo¨bius anomalies. Am J Med Genet 1986; 23:903–18.
[9]. Hegde HR, Shokeir MH. Posterior shoulder girdle abnormalities with absence of pectoralis major muscle. Am J Med Genet 1982;13:285–93
[10]. Hedge HR, Leung AKC. Aplasia of pectoralis major muscle and renal anomalies. Am J Med Genet 1989;32:109–11.
[11]. Pranava VM, Rao PSSS, Neelachalam A, Sailendra VH. Poland's syndrome with renal hypertension. JAPI 2000;48: 452–3.
[12]. Alexander A. Fokin, MD, PhD, and Francis Robicsek, MD, PhD. Poland's Syndrome Revisited. Ann Thorac Surg 2002;74:2218–25).