Nigerian Journal of Surgical Research
Vol 8 No 1 – 2 ,2006 : 97– 98

Case Report

Poland's syndrome: an incidental finding on routine medical examination

[1] A. U. Hamidu [2],A Musa and [3]M. C. Tahir

[1]Department of Radiology Ahmadu Bello University Teaching Hospital Zaria. [2]Department of Orth. Surgery University Of Maiduguri Teaching Hospital. [3]Department of Surgery University of Maiduguri Teaching Hospital.
Request for Reprints to Dr Hamidu .A.U Department of Radiology Ahmadu Bello Hospital University Shika-Zaria

Introduction

The presence of syndactyly and partial absence of the pectoralis major muscle comprise Poland's syndrome named after Alfred Poland of Guys Hospital in London. In 1841 Alfred[1] described a man who had syndactyly, absence of the middle phalanges and muscular anomalies of the ipsilateral chest wall. Recognised complications include short fingers, costal cartilage defects and herniation of the lungs[2,3.] . The etiology remains obscure. The clinical features are most consistent with defective embryological development of the upper limb[5.] The aim of this case study is to add to the few documented cases of Poland's Syndrome.

Case Report

SM is a 20 year, old young man who came for routine medical check for recruitment into a local football club. Born to a 45 year old primigravida. He had no previous history of illness suggestive of poliomyelitis and no family history of congenital anomalies. Physical examination revealed webbing of the index fingers of the right hand and decreased prominence of the right hemithorax (when compared to the left) because of lack of the pectoralis major musculature (Fig.1).Radiographic examination of the chest showed relative hyperlucency of the right right hemi thorax compared to the left and absence of the pectoralis major muscle shadow on the right side (Fig.2). Abdominal ultrasound showed normal kidneys, liver and gallbladder.
Diagnosis of Poland's Syndrome was made

.

Table 1. **Summary of frequency and types of abnormalities on record in 55 cases of Poland's syndrome J.W. Mace**

Abnormalities	Incidence	Incidence(%)
Absent Pectoralis Major	55/55	100
Syndactylism (Partial or complete)	49/55	89
Brachydactyly	48/55	87
Hypoplasia of Wrists and hands	49/55	89
Abent Phalanges or digits	25/55	45
Hypoplasia of forearm	20/55	37
Hypoplasia of arm	4/55	7
Thoracic cage defect	14/55	25
Absent Nipple	6/55	11
Herniation of the lung	5/55	9
Axillary Web	4/55	7

Discussion

The major features of Poland's Syndrome as found in the 55 previously reported cases are listed in table 1[5]
As an essential feature of the syndrome absence of at least the sternal head of the pectoralis major muscle must be present. More than 90% of the patients with Poland's Syndrome presented with
syndactyly or brachydactyly or both along with hypoplasia of the involved hand [5.] How often the pectoralis minor muscle is involved is not known

because of the difficulty in examining this[1] . The male to female ratio is 3:1 about the same sex ratio as reported for isolated unilateral syndactyly[6] .75% of the defect occurred on the right side , which is in agreement with our case study. The reason for the apparent predilection on the right side is unknown. Approximately 40% of the the patients have hypoplasia of the ipsilateral forearm or absent digits or both;thoracic cage defect occurred in 20%[7].The hypoplasia of the forearm is is usually mild but may be severe and may approach total aplasia. Herniation of the lungs, ipsilateral axillary webs or bands and absence of the nipples are all documented features (5&7). Poland's Syndrome has never been reported to involve both sides (1&5). The anatomical classicifications of hand anomalies in Poland's Syndrome are well documented in the works of Minguella-so7. The etiology of the Syndrome is unknown but Timothy J.D[8] suggested that drug toxicity could be the cause. It was also reported that the mothers of the patients took an unknown dose of ergonovine maleate while pregnant, this drug has the basic structure of lysergic acid and lysergic acid diethylamide, which might be the cause of limb deformities (4&6).

The role of radiology is to confirm diagnosis and aid in surgical management. Radiographic examination of the chest reveals hyperlucency of the affected side, mimicking a radical mastectomy. A dense ascending line as a result of the absence of the pectoralis major muscles replaces the normal downward curve of the axillary fold. Other causes of unilateral hyperlucency include;poliomyelitis, compensatory emphysema, Macleod's syndrome and Markedly rotated normal chest X-ray. Plain radiograph of the upper limb show various forms of shortening, fusion or aplasia of the digits. Treatment of patients is dependent on the degree of involvement of the upper extremity and is centered around reconstructive surgical procedures on the hand. They may however be some functional deficits in the released fingers[9].

Reference:

1. Marylyn P, Tofeichow and Eugene F. Poland's Syndrome , Paediat. Radiol. 1971,101:619-623
2. Chantara EA, Frire N. Poland's Syndrome. Br Med. J, 1971,4 :812-819.
3. Carakushansky G, Neu RL Gardner. (1969), Lysergide and Cannabis as Possible teratogens in man. Lancet 2:1087-1092.
4. 4. Hecht F, Beals RK, Lees MH,. Lysergic –acid di`ethylamide and Cannabis as Possible teratogens in man, Lancet 1968,2:1087.
5. 5. Mace JW, Kaplan JW., Poland's Syndrome, Clinical Pediatr; 1972, 11:98
6. 6.Timothy JD.. Nature and Etiology of the Poland's anomaly, New Engl. J of Med 1972,10:287.
7. Minguella-sol J, Cabrer-Gonzalez M. Poland's Syndrome, a report of 36 cases,An Esp Pediatr, 1998,. 48:143.
8. Jasonni V, Lelli-Chiesa PL, Repett P, Torre M, Nobili F, Mazzola C, Martucciello G. Congenital deformities of the chest wall, surgical treatment, Minerva Pediat. 1997.49: 407.
9. Gatti JE. Poland's deformity reconstruction's with a customized extra soft silicon Prosthesis; Ann Plast Surg. 1997,39:122.
10. .