CASE REPORT

An Unusual Presentation of Poland's Syndrome

Rajesh Kumar Gupta, Chander Rashmi Gupta, Deedar Singh

Abstract

We are presenting a rare case of Poland Syndrome which in addition to its usual features has facial nerve palsy.

Key Words

Poland syndrome, Facial nerve palsy

Introduction

Poland's syndrome is a rare congenital anomaly. Only 400 cases have been reported in world literature, whereas in Indian literature only 4 cases have been reported so far (1). In 1841 Alfred Poland first described the association of congenital thoracic anomalies with ipsilateral syndactyly. The term Poland's syndactyly was first used to describe this group of congenital anomalies by Clarkson in 1962 (2), although the observation had been made by many authors prior to that date. More recently many authors have reported small series of cases of Poland's syndactyly. Since Baudinne and co-workers reported a case of "Poland's syndrome" in 1967, the condition has been referred to as syndrome (3).

Case Report

A four year old boy weighing 18 kgs was brought to us by his parents with chief complaint of webbing of fingers of right hand since birth. The boy was delivered normally by a multigravida mother. The boy was born second of the twins. Prenatal history was unremarkable. There was no history of drug ingestion, radiation exposure or infection during pregnancy. There was no family history of congenital anomalies. Findings on general physical examination were within normal limits except for facial nerve palsy on the left side (Fig. 1). The right hemithorax was less prominent than the left. The pectoralis major muscle was absent. The anterior axillary fold was less marked on right as compared to left. The right nipple was present but hypoplastic. The right shoulder movement was normal. The right upper extremity was atrophic with hypoplastic musculature. There was shortening of forearm segment of about three cms (Fig 2). The right hand was hypoplastic. There was syndactyly of right hand involving the index, middle, ring and little fingers. The syndactyly was simple incomplete type (Fig. 3) X-ray chest revealed no rib anomalies. X-ray examination of the hand revealed hypoplasia of the middle and distal phalanges of index, middle, ring and little fingers (Fig. 4). Surgical correction of the syndactyly was done by release of syndactyly between ring and middle fingers with reconstruction of web by Sgook Technique (4).

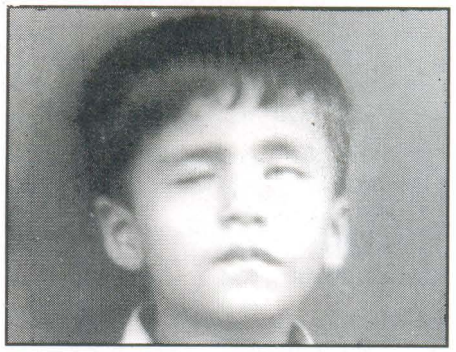

Fig. 1. Photograph showing facial nerve palsy on left side.

From the Department of Orthopaedics, Govt. Medical College, Jammu (J&K) India.
Correspondence to : Dr. R. K. Gupta, Deshmesh Nagar, Digiana, Jammu-180010 (J&K) India.

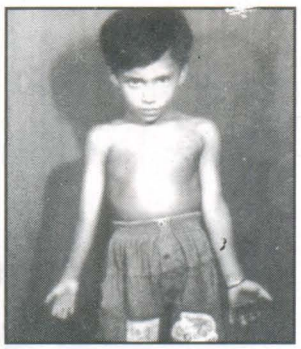

Fig. 2. Photograph showing atrophic, hypoplastic (R) upper extremity with short forearm segment.

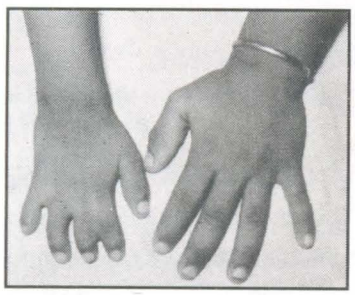

Fig. 3. Photograph showing hypoplastic (R) hand with syndactyly.

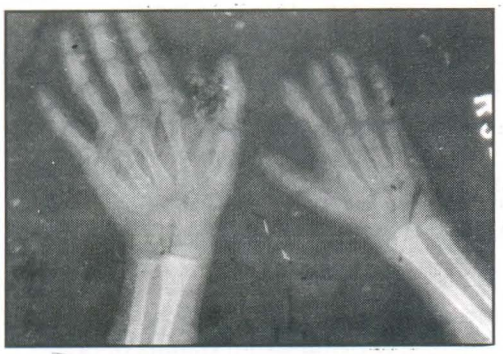

Fig. 4. X-ray of hands showing hypoplasia of middle & distal phalanges of index, ring, middle and little fingers of right hand.

Discussion

The etiology of Poland's syndrome is unknown. Although hereditary traits have been demonstrated for some anomalies of the hand such as polydactyly, only one report of Poland's syndrome described familial incidence (5). Poland's syndrome is more common in boys and more common on right than left. The reported incidence of Poland's syndrome with syndactyly of the hand ranges from 2.5 to 13.5% (6,7). It is thought to be due to vascular insult early during gestation when the limb bud and chest wall development are dependent on end artery circulation. The clinical features are variable but always include congenital aplasia and syndactyly. The middle phalanges are hypoplastic or absent so that effectively there is only one interphalangeal joint. The syndactyly is usually incomplete and simple. It may involve all fingers and frequently includes the thumb, which then lies in the same plane as the fingers. Poland's syndrome may also include hypoplasia of the nipple and breast, hypoplasia of the upper ribs, herniation of the lung, contracture of the anterior axillary web and elevated scapula. The arm and more frequently the forearm are hypoplastic. In 1984 Gausewitz *et. al.* reported mild facial weakness in one case out of the ten cases presented by them (8).

Summary

A case of Poland's syndrome with facial nerve palsy has been presented because of its rarity as only one reference of facial nerve involvement with this syndrome has been reported in English literature so far.

References

1. Lohokare SK, Krishna M, Panse MP. Poland's syndrome: a report of four cases. *IJO* 1991 ; 25 (1) : 43-44.
2. Clarkson, Patrick. Poland's syndactyly. *Guy's Hosp Rep* 1992 ; 111 : 335 - 46.
3. Baudinne P, Bovy GL, Wasterlain A. Us cas de syndrome de. *Poland Acta Paediat Belgica* 1967 ; 32 : 407-10.
4. Skoog, Tord. Syndactyly. A clinical report on repair. *Acta Chir Scand* 1965 ; 130 : 537 - 49.
5. Trosev K, Cervenka Cervenka J & Gerberova E. Congenital dysplasia of the pectoralis muscles associated with congenital malformations of the hand and fingers of the ipsilateral upper limb. *Acta Chir Orthop Traum Cechoslovaca* 1966 ; 33 : 320 - 27.
6. Epstein LI, Bennett JE. Syndactyly with ipsilateral chest deformity. *Plastic and Reconstr Surg* 1970 ; 46 : 236 - 40.
7. Velasco JG, Broadbent TR, Woolf RM. Syndactylism. *British J Plast Surg* 1967 ; 20 : 364 - 68.
8. Gausewitz HS, Meals RA, Setoguchi Y. Severe limb deficiencies in Poland's syndrome. *Clin Orthop* 1984 ; 185 : 91.