Case Report for Discussion (No 7).

Poland's syndrome in association with clinodactyly of the thumb

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Abstract

This report describes the case of a 7-year-old Caucasian male infant with clinical and radiological findings indicative of Poland's syndrome and clinodactyly of the thumb. Although Poland's syndrome associated with upper limb abnormalities has been described, we believe that this is the first documentation of Poland's syndrome associated with clinodactyly of the thumb.

Introduction

In 1841 Alfred Poland described the association of ipsilateral absence of pectoralis major muscle and syndactyly[1]. Since then several authors have reported this rare congenital sporadic disorder, which became known as "Poland's syndrome"[2]. Although, the presence of coexisting anomalies with Poland's syndrome is well known[3], the patient described herein does not appear to have any of these. This report describes a case of Poland's syndrome associated with clinodactyly of the thumb. We believe that this is the first documentation of Poland's syndrome described in association with clinodactyly of the thumb.

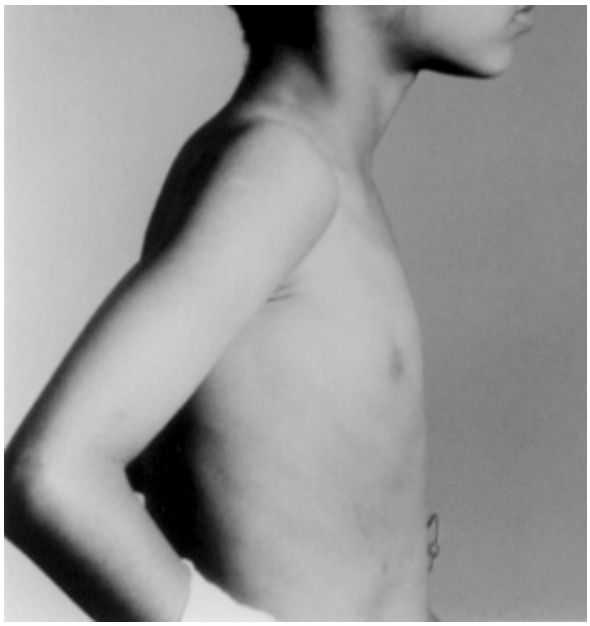

Fig 1

Case Report

A 7-year-old Caucasian male infant was referred to the Plastic and Reconstructive Surgery Unit at Charing Cross Hospital with a congenital deformity of his right upper limb. He was born to a 36-year-old female at full term by normal delivery and it was the sixth child in family. There was no prior family history of congenital anomalies. The patient was first seen at Hospital at the age of 2-years-old and no operation was recommended at that time. Due to developmental delay, he attended a school for children with special educational needs. His family had become increasingly concerned about his right hand because of the psychological problems resulting from this handicap.

At the physical examination there was brachysyndactyly of the right hand with a simple complete syndactyly of the index and middle fingers. The little finger was essentially normal but all the other digits were hypo-plastic with only two phalanxes. In addition he had clinodactyly of the thumb with an abnormal proximal phalanx. There was a 30 degree deviation in ulnar direction. Examination of the shoulder girdle revealed absence of the sternal head of the pectoralis major. A clinical diagnosis of Poland's syndrome in association with clinodactyly of the thumb was made. Apart from the deformity of the upper limb on the right side, he had no other coexisting abnormalities.

Fig 2

Radiographic examination of the right upper limb showed abnormalities only in the right hand. There was fusion of the index and middle fingers with hypoplasia of the proximal phalanx of the middle finger and an absent proximal epiphysis. The middle phalanxes of the index, middle, and ring fingers were absent while the distal phalanxes of the same fingers show prominent epiphyses and coned shaped metaphysis. The middle phalanx of the little finger was hypo plastic. The tips of the distal phalanx of the index and little fingers were abnormally blunted. The appearance of the carpal bones and wrist was also indicative for a delay in the bone age.

Under general anaesthesia, surgery for syndactyly was performed through a zig zag incision. A dorsal quadrelateral flap was elevated and the two digits were separated. Full thickness skin grafts from the right groin were grafted in the areas where skin was missing. For the clinodactyly of the thumb an osteotomy of the proximal phalanx was performed. A wedge shaped bone graft from the right iliac crest was introduced and fixed with a K-wire.

Discussion

Poland's syndrome is a rare congenital abnormality, where the most common clinical appearance is unilateral brachysyndactyly and ipsilateral deficiency of the sternal head of the pectoralis major. The brachydactyly is due to hypoplasia of the middle phalanxes. The syndactyly involves most often the index and the middle finger and the thumb is usually normal[1].

The presence of coexisting anomalies with "Poland's syndrome" is well known. Wilson et al. reported missing fingers, hypo plastic metacarpals or phalanxes and flexor tendon absence as coexisting hand anomalies[3]. They also associated other abnormalities such as : scoliosis, seizure disorder, heart murmur and dextrocardia, Mobius syndrome, Bonnevie-Ullrich syndrome, thrombocytopenia, foot anomalies and hypoplastic ribs, clavicle or scapula, with "Poland's syndrome". However their study or any other study in the literature has never included clinodactyly in the associated deformities coexisting with "Poland's syndrome".

This patient appeared to have Poland's syndrome without any of the coexisting abnormalities already described by some authors[3]. However he was presented with clinodactyly of the thumb. To our knowledge, such a coexistence, has never been documented.

The incidence of clinodactyly in normal children varies between 1% and 19.5 %[4]. It may be sporadic and often associated with a big number of syndromes, but it has not been reported in conjunction with Poland's syndrome.

References

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