

Poland syndrome (anomaly) with congenital hemangioma: A new association

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ABSTRACT

Unilateral defect of pectoral muscle and ipsilateral syndactyly constitute Poland syndrome. Absence or hypoplasia of the breast and nipple, axillary hair loss and dermatoglyphic abnormalities have also been reported in this syndrome. The primary defect could be in the development of the proximal subclavian artery with early deficit of blood flow to the distal limb and the pectoral region, resulting in partial loss of tissue in those regions. However, the association of congenital hemangioma with Poland sequence has not been observed so far. Such an association is being reported here in a 1-year-old infant, second-born of nonconsanguineous parents, who also had polydactyly instead of the documented syndactyly.

Key Words: Congenital hemangioma, Poland syndrome, Polydactyly

INTRODUCTION

In 1841 Poland reported unilateral absence of pectoralis minor and the sternal portion of pectoralis major muscle in an individual who also had syndactyly of the hand on the same side.^[1] Simple, complete or incomplete syndactyly can occur. This unique pattern has been noted in several cases and the incidence is about 1/30,000. Other associations observed are hypoplasia of the upper extremity (brachysymphalangism); absence or hypoplasia of the breast, nipple; loss of axillary hair and dermatoglyphic abnormalities. It has been estimated that 10% of the patients with syndactyly of the hand have Poland sequence. We report a case of Poland sequence in a girl with congenital hemangioma and polydactyly of the affected side.

CASE REPORT

A 1-year-old baby girl, second-born of nonconsanguineous parents, was evaluated for lower respiratory infection. She had a depression on the right side of chest at birth itself and after 5 days, she developed a small compressible red spot, which later extended up to the midline and right upper limb. There was no family history of similar complaints. There was no history of convulsions and her development was normal. Her only sibling, a 5-year-old boy, was normal.

Examination revealed an active child whose weight was 8 kg, length 76 cm and head circumference 45 cm. She had a high arched palate but no facial dysmorphism. The sternocostal part of pectoralis

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major was absent on the right side. The right nipple was placed at a lower level. There was an extensive well-defined erythematous compressible lesion with surface smooth in most of the areas and lobulated in other areas involving the right side of chest, axilla, flexor surface of the right upper limb and thenar eminence (C₄₋₇ and T₁ dermatomes) [Figure 1], suggestive of strawberry angioma. The baby also had preaxial polydactyly on the right side but no syndactyly, brachydactyly or oligodactyly. There was no wasting of muscles, nail changes or Sprengel's deformity (i.e., elevation of scapula on the affected side). Other systems were clinically normal.

Her hemogram and urine analysis were normal. Histopathology of the skin lesion was consistent with strawberry hemangioma. Computerized tomography and MRI scan of the chest confirmed the absence of sternocostal part of pectoralis major muscle on the right side. Radiograph of thoracic spine and ultrasound scan of abdomen did not show any



Figure 1: Extensive hemangioma affecting right upper limb and chest with polydactyly

abnormality.

DISCUSSION

The exact etiology of Poland's sequence is unknown. The male-female ratio is 3:1. In 75% of cases, right side is affected. Bouvet^[2] *et al.* and Bavinck^[3] *et al.* have suggested that the primary defect may be in the development of the proximal subclavian artery with diminished blood flow to the affected side, leading to partial loss of tissue in that region. Vascularization of the fetal skin begins during the third month of intrauterine life, but these vessels do not anastomose with the deeper vasculature until late in gestation.^[4] It has been suggested that infantile hemangioma arises where islands of embryonic cutaneous angioblastic tissue fail to establish normal contact with the rest of the developing vascular system.^[5]

Since Poland's sequence and congenital hemangiomas are of vascular etiology, their association may be due to an intrauterine vascular anomaly rather than an accidental coincidence. Parent-to-child transmission is possible with marked variability in expression. In some patients, absence of the pectoralis muscles alone or in others syndactyly alone can occur.

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