

POLAND SYNDROME: A RARE CASE

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ABSTRACT

Poland syndrome is a rare congenital anomaly characterised by unilateral absence of sternocostal head of pectoralis major muscle and pectoralis minor muscle along with ipsilateral rib anomaly and frequent association with ipsilateral upper limb brachysyndactyly. We present a case of a 10 month old male child having depressed left side of anterior chest wall without respiratory distress. The imaging findings were characteristic and diagnostic of Poland syndrome.

Keywords: Poland syndrome, pectoralis major, brachysyndactyly

Introduction

Poland syndrome is a rare entity, usually sporadic with incidence of 1 in 36000 or 50000¹ with male predilection about 3:1.² It was named after Sir Alfred Poland who first published it in 1841.¹ The syndrome is characterised by unilateral deficiency of pectoralis muscles (absence or hypoplasia of sternocostal portion of pectoralis major and lack of pectoralis minor), absence or hypoplasia of ipsilateral breast or nipple, deformity of ipsilateral ribs, mainly second to fifth and in some cases upper limb abnormalities such as brachysyndactyly.³ Dextrocardia, renal anomalies and lung herniation has been described in some rare cases.⁴ It has rare association with Mobius syndrome, Morning glory syndrome, Pierre Robin syndrome etc.⁵ We report a case of an infant with typical features of Poland syndrome without any limb anomaly.

Case Report

A ten month old baby was presented to us with depressed anterior chest wall on left side since birth without any history of breathlessness. The child was

born by normal delivery at full term and achieved milestones appropriate for age. His weight was 7.5 kg and height 64 cm. Physical examination revealed a depression in left anterior chest wall (Fig. 1). The left nipple was smaller compared to right nipple. There was no other anomaly on further examination. Both

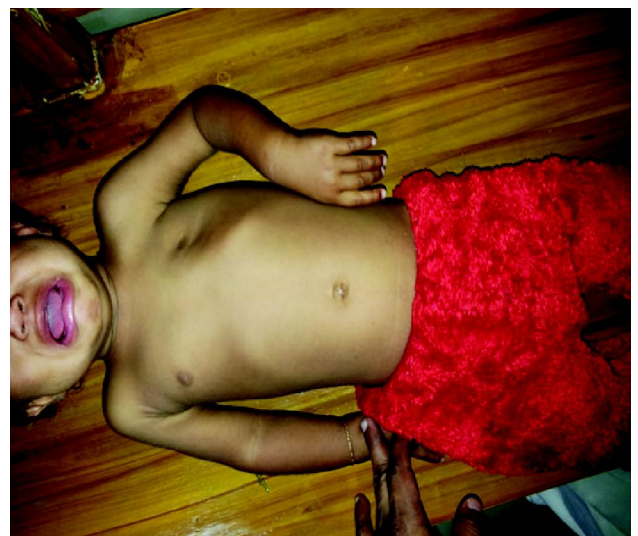


Figure 1: Child with depression in left anterior chest wall and smaller left nipple compared to that of right side.

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upper and lower limbs were normal. The supine frontal chest radiograph showed hypertransradiant left hemithorax. Helical CT scan of thorax done in a multi slice scanner revealed absence of pectoralis major and minor muscles of left side (as shown in Fig. 2) along with deformity and hypoplasia affecting the anterior part of left third rib (Fig. 3). Lungs, pleural spaces and mediastinum were unremarkable.



Figure 2: Axial CECT image of thorax reveals absence of pectoralis major and minor muscles of left side.

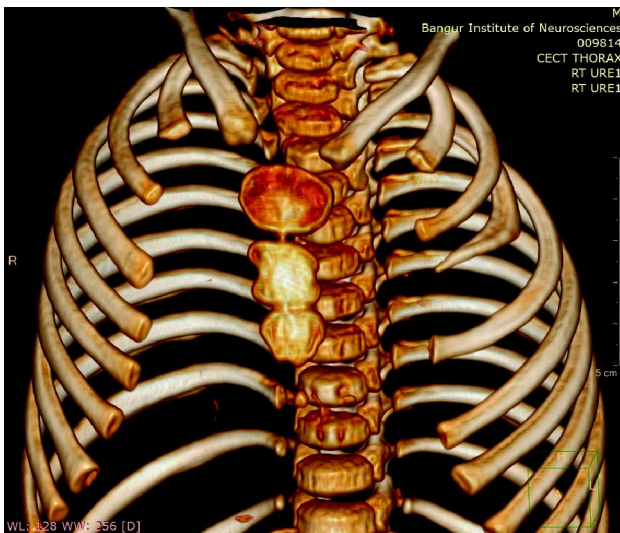


Figure 3: Volume rendered 3D reconstruction shows deformity and hypoplasia affecting the anterior part of left third rib.

Discussion

Poland syndrome is a rare, mostly sporadic in occurrence, though a few authors suggest autosomal inheritance with incomplete penetrance.⁶ In our case, Poland syndrome has occurred as sporadic event.

Embryologically pectoral muscle split into sternal and clavicular head at 6th week. Around this time tissue within the digits of hand also start to disappear to form web spaces. Vascular insult due to interruption in subclavian arterial supply at specific sites produce corresponding patterns of defect.⁷

Absence of pectoralis muscle is usually unilateral and mostly in right side (75%).⁸ But in our case it is found on left side which is even rare. Ipsilateral rib anomaly and hypoplasia of nipple as seen in our case are recognized features of Poland syndrome. Ipsilateral syndactyly and brachydactyly are common associations with Poland syndrome which were not present in our case.

The surgical option is generally recommended after completion of growth. Treatment options are autologous fat injection, pedicled latissimus dorsi muscle transfer, transverse rectus abdominis musculocutaneous flap, custom made chest wall prosthesis and sternal / rib reconstruction, or combination of techniques.⁹

Conclusion

We have reported a case of Poland syndrome, a rare entity, in a ten months old child involving left anterior chest wall without any limb abnormality.

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