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Poland syndrome in young adult: A rare case report

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Abstract

We report rare case of Poland syndrome in young adult. The authors tend to familiarize health care professionals in the region of this rare congenital condition.

Keywords

Poland Syndome; young; adult; absent; pectoralis major; rare.

Introduction

Poland syndrome is a rare congenital anomaly usually presenting with underdeveloped chest wall musculature; mostly unilateral. In addition to this there may be subcutaneous tissue scarcity, deformities of costal cartilages and multiple ribs and unilateral brachysyndactyly of the same side with variable levels of extent and involvement in different individuals [1,2].

Case presentation

We hereby report a case of adult presentation of Poland syndrome presenting with flattened anterior chest wall musculature of the left side since birth with normal growth parameters and good mental status. A 22-year-old boy from Bulandshahr, Uttar Pradesh presented to our hospital, with the flattening of the left anterior chest wall since birth (Figure 1a). There was no significant familial history. All the growth parameters were normal and the patient had good mental status. Computed tomography scan of the chest showed absence of the left pectoralis major and minor muscle and of some of the anterior ends of left ribs (Figure 1b). With the typical features the diagnosis of Poland syndrome was rendered.

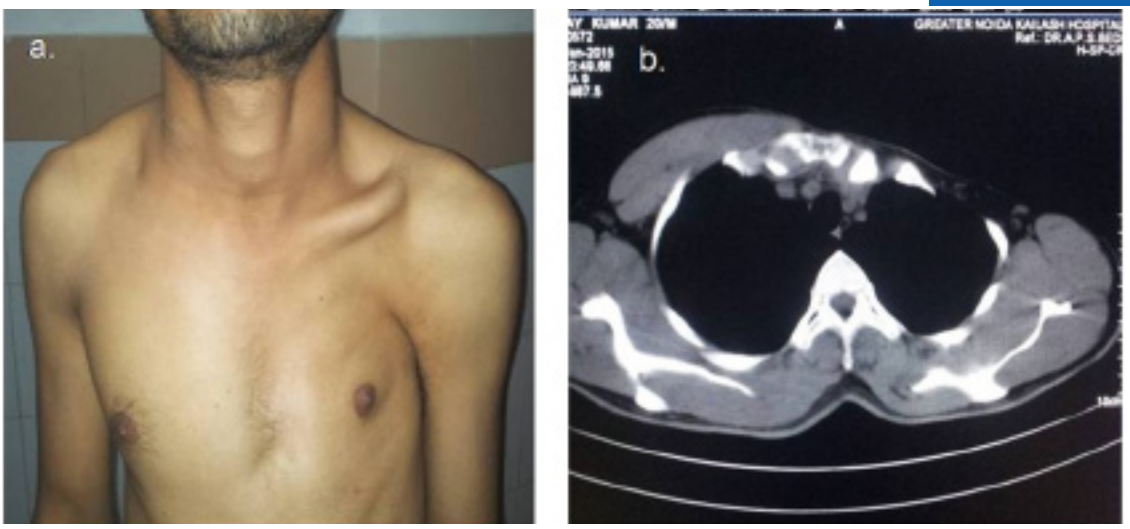

Figure 1: (a) Photograph showing 22 years male with Poland Syndrome. There is chest asymmetry with anterior chest wall depression and flattening of pectoralis region of same side. **(b)** Computed Tomography Scan showing absence of left pectoralis major, pectoralis minor muscle and anterior ends of few left ribs.

Discussion

Poland syndrome (also Poland sequence or Poland's anomaly) was first described in 1841 by Sir Alfred Poland and is characterized by underdevelopment or absence of the chest muscle (pectoralis) on one side of the body, and also usually webbing of the fingers (cutaneous syndactyly) [3]. The reported incidence by different authors ranges from 1:10,000 to 1:100,000 and is observed more frequently in males than in females with the right side of the body affected more often than the left [4]. The condition typically presents with unilateral absence of the sternal or breastbone portion of the pectoralis major muscle which may or may not be associated with the absence of nearby musculoskeletal structures [5,6].

It can be diagnosed at a primary instant by the clinical features as mentioned before. An overwhelming majority of cases occur due to unknown cause. However, researchers suggest that it arises from interruption of early embryonic blood flow during sixth week of fetal development, which predispose to this anomaly. Cases of Poland syndrome which are associated with leukemia and breast carcinoma suggest a relation of this developmental defect and tumors and hence require oncological awareness. This syndrome is treated using mainly reconstructive surgery [7]. Fused fingers are separated as early as possible and breast or chest implants may be used after full physical development has been reached. Today, bioengineered cartilage can be implanted to help and give the chest a more normal appearance [8]. Adequate knowledge of this condition helps in making early diagnosis and timely therapeutic approaches with follow-up in early growth period.

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