Bilateral Poland's Syndrome: A Case Report

Bilateral Poland Sendromu Olgusu

Bilateral Poland's Syndrome

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Özet

Poland sendromu nadir gözlenen konjenital bir sendromdur. Genellikle major ve minör pektoral kasın hipoplazisi veya yokluğu, meme ve/veya meme başının anomalisi, cilt altı yağ dokusunun hipoplazisi, göğüs deformitesi, pektoral ve aksiller kıllanma yokluğu ve el anomalileri ile karakterizedir. Poland sendromu genellikle tek taraflıdır. Litaretürdeki ikinci bilateral Poland sendromu olgusunu sunmaktayız. Bilateral pektoral kas yokluğunun yanında birçok bilateral kas yokluğu bulunan 20 yaşında bir erkek hasta sunmaktayız. Hastamızın aynı zamanda bilateral pektoral ciltaltı yağ dokusu hipoplazisi ve bilateral pektoral kıllanma yokluğu ve bilateral sprengel deformitesi mevcuttu. Unilateral bir sendrom olarak bilinen Poland sendromunun bilateral de olabileceğini göstermesi nedeniyle olgumuzun önemli olduğunu düşünmekteyiz.

Anahtar Kelimeler

Bilateral Poland Sendromu; Sprengel Deformitesi; Pectoral Kas Agenezisi

Abstract

Poland's syndrome is a rare congenital anomaly with absence or hypoplasia of major and minor pectoral muscles, anomaly of breast or nipple, hypoplasia of subcutaneous tissue, chest wall deformity, pectoral alopecia and axilla, and hand anomalies. Poland's syndrome is mostly unilateral. Here, we present the second case of bilateral Poland's syndrome in the literature. The case of a 20 years old male with absence of bilateral pectoralis major muscles and bilateral many other muscles, is hereby presented. He also had bilateral hypoplasic pectoral subcutaneous tissue, pectoral alopecia and Sprengel's deformity. We think that our case is important since it shows that Poland's syndrome which was defined as unilateral may be bilateral.

Keywords

Bilateral Poland's Syndrome; Sprengel's Deformity; Agenesis of Pectoral Muscle

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Introduction

Poland syndrome is a rare congenital anomaly with absence or hypoplasia of major and minor pectoral muscles, anomaly of breast or nipple, hypoplasia of subcutaneous tissue, chest wall deformity pectoral alopecia, and axillary and hand anomalies (1,2). While absence of many upper extremity muscles may accompany to syndrome; Sprengel deformity, Klippel-feil and Möbius syndrome may be observed (3). One of accepted features of the syndrome is being unilateral. However, Karnak and friends reported a bilateral case in 1998 (4). We want to re-review the definition of Poland syndrome with our case which is an example of bilateral Poland syndrome.

Case Report

A 20 years old male patient was admitted due to chest deformity and restriction in upper extremity mobilization. Bilateral pectoralis major muscles and many other muscles could not be palpated in physical examination (Figure 1). It was seen that subcutaneous tissue was hypoplasic and there was alopecia on pectoral area. Left nipple was smaller and asymmetric with the other. Abductions of both extremities were noticed as ½ limited. Bilateral Sprengel's deformity was detected (Figure 2). Thoracic CT displayed the bilateral absence of pectoralis major, latissimus dorsi, serratus anterior and trapezius muscles, and bilateral hypoplasia of pectoralis minor muscles (Figure 3). The patient was diagnosed as bilateral Poland syndrome. Any surgical intervention was not considered and the follow-up was decided for the patient.

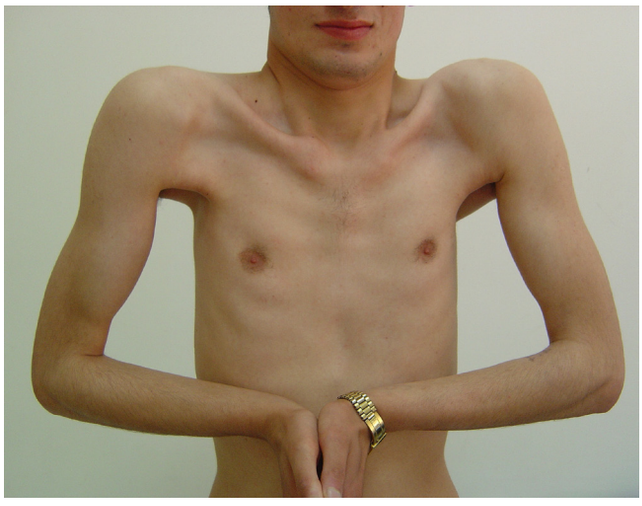


Figure 1. Bilateral Poland Syndrom

Discussion

Poland's syndrome is a rare congenital anomaly characterized by hypoplasia of the breast and nipple, paucity of subcutaneous tissue, partial or complete absence of the pectoralis major muscle, lack of a pectoralis minor muscle, aplasia or deformity of costal cartilages or ribs, alopecia of the axillary and mammary region, and unilateral brachysyndactyly (1).

The main peculiarity of all patients with Poland's syndrome is unilateral hypoplasia or absence of pectoralis major muscle. Patients with at least one of the other associated lesions are considered to have the syndrome (4).

Although the Poland's sydrome is mostly unilateral, a single bilateral case has been reported (4). Our patient has bilateral



Figure 2. Bilateral Sprengel's Deformity

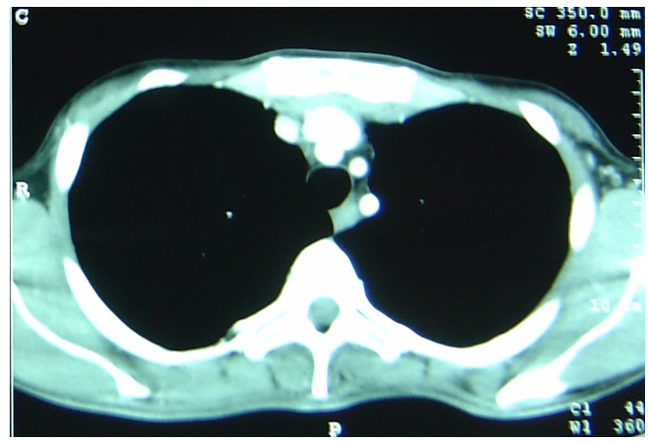


Figure 3. Absence of multiple muscles (CT)

absence of pectoralis major muscles, bilateral hypoplasia of subcutaneous tissue, bilateral hypoplasia of pectoralis minor muscles, alopecia of the mammary region. Bilaterally pectoralis major muscles are absent and there are other associated lesions which are enough to consider to have Poland's syndrome for each side. Therefore our patient was considered to have bilateral Poland's syndrome.

In some cases of Poland's syndrome, the latissimus dorsi, external oblique, and serratus anterior muscle are also affected. In our case, bilateral latissimus dorsi, serratus anterior muscles were absent. Differently bilateral trapezius muscles were absent. Isolated bilateral absence of trapezius muscles has been reported. There are some issues noticed that it may occur as a component of Poland's syndrome (5).

In Sprengel deformity there is elevation and winging of scapula which is hypoplastic. In some patients with Poland's syndrome like ours, partial or complete absence of the serratus anterior leads to varying degrees of elevation and winging of scapula. The hypoplasia of scapula and muscle defects are the result of an interruption of early of embryogenic blood supply in the subclavian arteries, the vertebral arteries and/or their branches as Poland's syndrome(3).

In conclusion, Poland's syndrome was routinely described as unilateral since the syndrome was defined firstly by Alfred Poland in 1841. However after approximately 150 years, a bilateral Poland's syndrome was reported by Karnak and friends. The definition of Poland's syndrome should be reviewed. We think that our case is important example to show that Poland's syndrome can be bilateral.

Competing interests

The authors declare that they have no competing interests.

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