

Poland's Syndrome Associated with Opposite Side Retractable Testicle

Karşı Taraf Retraktıl Testis İlişkili Poland Sendromu

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Abstract

Poland's syndrome is a rare congenital anomaly of unknown cause characterized by unilateral, partial or complete chest wall hypoplasia and ipsilateral hand abnormalities as especially on the right side. Many abnormalities have been described in association with this syndrome as dextrocardia, dextroposition, renal malformation and tumours. The case of a 37 years old man with absence of left pectoralis major muscle, hypoplasia of the ribs II and III so depending on this lung herniation on the anterior wall of the chest and reduced apex lung volume but with right retractile testicle; was reported. Especially; genitourinary system investigations must be performed on patients with Poland syndrome.

Key words: Lung herniation, Poland's Syndrome, retractile testicle

Özet

Poland sendromu, özellikle sağ tarafta olduğu gibi, tek taraflı, kısmi veya tam göğüs duvarı hipoplazisi ve ipsilateral el anormallikleri ile karakterize, nedeni bilinmeyen nadir bir konjenital anomalidir. Bu sendromla ilişkili olarak dekstrocardi, dekstropozisyon, böbrek malformasyonu ve tümörler olarak bir çok anormallik tanımlanmıştır. Sol pektoralis major kas yokluğu, kaburgaların II ve III nolu hipoplazinin göğüs ön duvarındaki akciğer fitiğine ve apeksin akciğer hacminde azalmaya yol açtığı, ve sağ retraktıl testis olan 37 yaşında bir erkek olgu sunulmuştur. Özellikle genitoüriner sistem araştırmalarının, Poland Sendrom'lu hastalarda yapılması önerilmektedir.

Anahtar kelimeler: Akciğer fitiği, Poland Sendromu, retraktıl testis

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Introduction

Poland's syndrome, also known as Poland's Anomaly or Poland's Sequence; is a severe congenital anomaly classically characterized by unilateral absence of at least the sternal head of the pectoralis major muscle is pathognomonic and unilateral brachysyndactyly (1,2). In addition; it consists of many thoracic deformity features with lack of the pectoralis minor muscle, hypoplasia of the breast and nipple, aplasia or deformity of the costal cartilages or ribs II to IV or III to V, alopecia of the axillary and mammary region with anterior axillary fold contracture (3). The incidence of Poland's Syndrome ranges from 1 per 7.000 to 1 per 100.000. The male and female ratio is 2:1 to 3:1 (2,3). In males the right side is affected more than 2:1; however, in females there is no favored side (3).

Case

A 37-year-old man presented with coughing and difficulty in breathing and was admitted to a hospital. Physical examination revealed absence of the left pectoralis major muscle, unilateral syndactyly especially between second and third fingers and right retractile testicle (figure 1 and figure 2). Chest was on the right side with lateral displacement of the left nipple due to the absence of the left pectoralis major muscle (figure 1). In pulmonary function test; all values were at nearly limit. Cardiovascular examination revealed normal without gallop. Laboratory findings were within the normal ranges. The family history was unremarkable.

In echography, renal and abdominal sonography was normal. Scrotal sonography confirmed the

physical examination as right retractile testicle with normal testicles structure. Radiograph of left hand showed hypoplasia of the middle phalanx of fingers and distal phalanx of fifth finger. Chest radiograph revealed absence of the anterior portions of the second and third ribs (figure 3).

Figure 1. Photograph showing the hypoplasia of the left anterior chest wall



Figure 2. Photograph showing left hand with syndactyly



Discussion

In 1841, Alfred Poland, a student–demonstrator in anatomy defined a case as unilateral absence of sternocostal part of pectoral major and total absence of pectoralis minor and deficiency of serratus anterior and external abdominal oblique muscles with ipsilateral syndactyly (4). But earlier from his reports, in 1826 (by Lallemand) and in 1839 (by Froriep) similar reports were

The chest computed tomography revealed absence of left pectoralis major and minor muscle, hypoplasia of the ribs II and III, herniation on the anterior wall of chest associated with left hemithorax and reduced apex lung volume of left lung (figure 4).

Figure 3. Frontal chest radiography

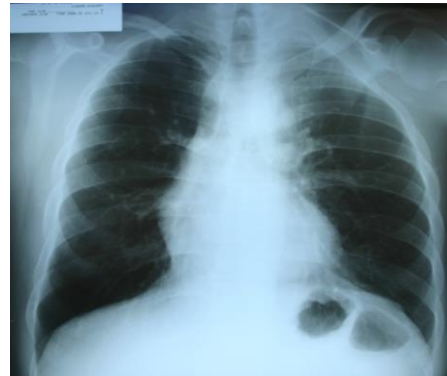


Figure 4. Chest computed tomography showing the absence of the left pectoralis muscles and arrow shows the lung herniation.



described. Patrick Clarkson (1962) called syndactyly with absence of pectoralis major muscle as Poland's syndactyly. In 1967 Poland's Syndrome received its name by Baudinne and colleagues (2,3,5).

The etiology of Poland's syndrome is still unknown (3,6). At the moment vascular defect hypothesis is valid and according to this hypothesis an interruption or reduction of the

embryonic blood supply of the subclavian artery or vertebral arteries and/or their branches during the sixth week of gestation is the pathogenesis of Poland's, Moebius and Klippel-Feil Syndromes (7,8). Bavinck and Weaver hypothesized with suggesting a term "subclavian artery supply disruption sequence" According to this hypothesis, an interruption of the subclavian artery the point where is proximal to the origin of the internal thoracic artery and distal to the origin of the vertebral artery (8). Although most reported cases of Poland's syndrome is generally sporadic as our case and syndrome defined as congenital disorder with low (<1%) risk of reoccurrence in the same family, there are several reports of familial occurrence (1,5,6,7). Soltan and Holmes suggested that familial occurrence of the vascular anomalies results the some malformations as Poland's syndrome (9).

In some cases Poland's syndrome was associated with different malignancies as leukaemia, non-Hodgkin's lymphoma, cervical cancer, leiomyosarcoma, lung cancer, gastric cancer and breast cancers (3). Lung herniation reported in 8% of patients with Poland's syndrome as in this case (3).

Renal anomalies in Poland's syndrome reported as unilateral renal agenesis or duplication of the urinary collecting system (3,10); however, in the literature, we could not find any case of Poland's syndrome associated with retractile testicle especially the opposite side.

As Poland's syndrome is associated with unexpected disorders, we recommend genitourinary system investigations also be performed on patients with Poland's syndrome.

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