A Rare Chest Wall Deformity; Poland Syndrome: Case Report

Nadir Görülen Göğüs Duvarı Deformitesi: Poland Sendromu: Olgu Sunumu

Ali Çelik, Kerim Tülüce, Muhammed Sayan, İsmail Cüneyt Kurul

Department of Thoracic Surgery, Faculty of Medicine, Gazi University, Ankara, Turkey

ABSTRACT

Poland Syndrome is a rare chest wall anomaly which is often noticed by the patient's family in childhood. Physiologic and psychologic problems may develop in the following years but rarely need surgery for this pathology. A 13 year-old boy was admitted to our clinic for chest wall shape defect. After the clinical workup, Poland Syndrome was diagnosed. The patient, who had no indication for operation, is being followed up by informing his family. *(Gazi Med J 2011; 22: 49-51)*

Key Words: Costa agenesia, syndactyly, pectoral muscle absence, winging scapula

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ÖZET

Poland Sendromu nadir görülen ve sıklıkla çocukluk döneminde hastanın ailesi tarafından fark edilen göğüs duvarı anomalisidir. Nadiren cerrahi gerektiren bu patolojiler ilerleyen yıllarda fizyolojik ve psikolojik sorunara neden olabilir. Onüç yasında erkek çocuk göğüs duvarı sekil bozukluğu nedeniyle kliniğimize basvurdu. Tetkikler sonrası hastaya Poland Sendromu tanısı konuldu. Operasyon endikasyonu olmayan hasta ailesinin de bilgilendirilmesiyle takibe alındı. *(Gazi Med J 2011; 22: 49-51)*

Anahtar Sözcükler: Kosta agenezi, sindaktili, pektoral kas yokluğu, kanat skapula

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INTRODUCTION

Poland Syndrome occurs in 1/7.000-100.000 births and is characterized by the absence of pectoral muscles, besides costa agenesia, abnormal posture, chest wall collapse, abnormalities of the scapula, patchy absence of hair under the arm on the affected side, underdevelopment of subcutaneous fatty tissue, syndactyly, brachydactyly, underdevelopment or absence of breast or nipple. Surgical repair is generally performed for paradoxic respiration, advanced depression of the chest wall or sternum and breast anomalies in girls.

CASE REPORT

Thirteen year-old boy was admitted to our clinic for chest wall deformity. History revealed a surgery carried out 3 years earlier for a congenital adhesion between the second and third fingers of his right hand. No one else in his family had such a disorder. Physical examination revealed absence of pectoral muscles on the right side, decrease and abnormal localization of hair in the same side axilla (Fig. 1), wing scapula on the opposite side (Fig. 2) and scar tissue secondary to surgery on the right hand (Fig. 3). Chest x-ray was normal. Thorax tomography showed no

Address for Correspondence/Yazışma Adresi: Ali Çelik, Department of Thoracic Surgery, Faculty of Medicine, Gazi University, Beşevler 06500 Ankara, Turkey Phone: +90 530 404 54 67 - +90 506 406 70 67 E-mail: alicelik78@gmail.com
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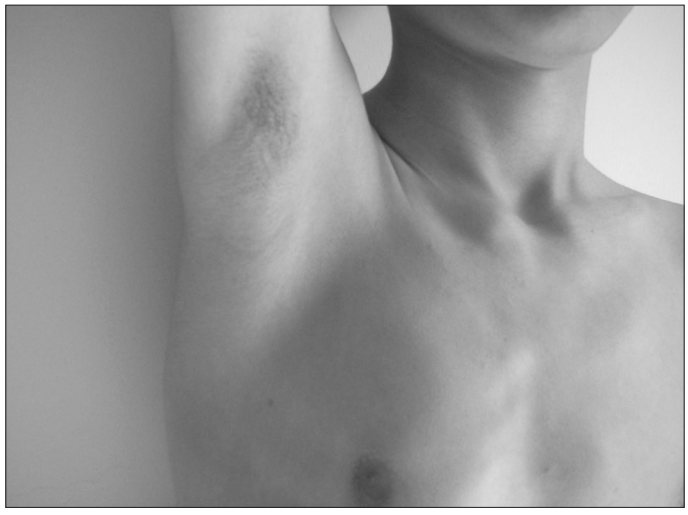

Figure 1. Malposition of axillar hair with absence of pectoral muscles and mild lower sternal depression (2816x2112 Pixel image)

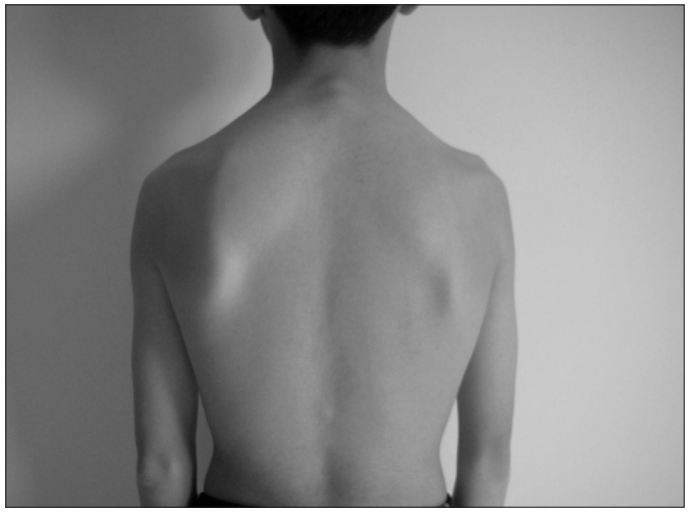

Figure 2. Protrusive appearance of scapula on posterior view (Winging scapula) (2816x2112 Pixel image)

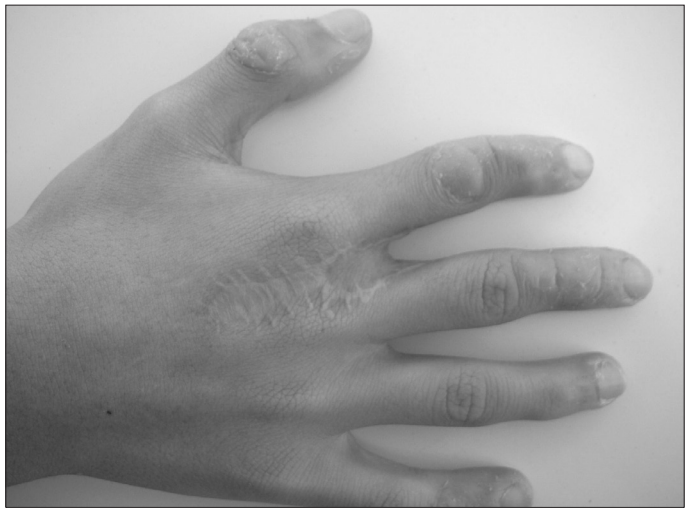

Figure 3. On the right hand, operated patient for adhesion between second and third phalanx three years previously, scatris tissue (2816x2112 Pixel image)

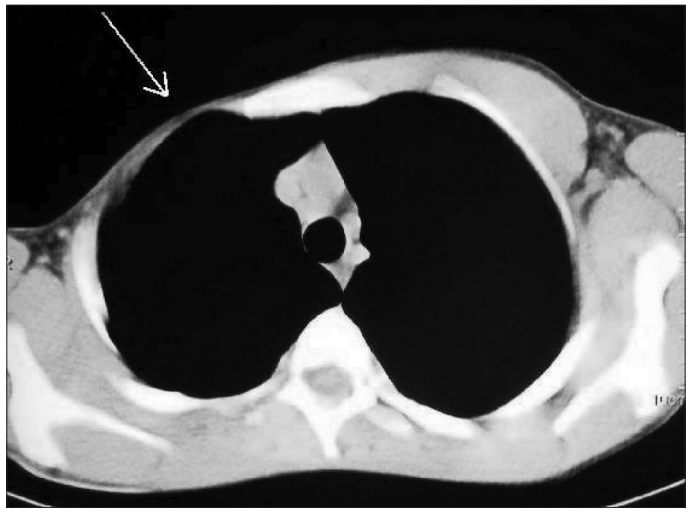

Figure 4. Thorax CT: Absence of pectoral muscles and poor subcutaneous fat tissue (600x450 Pixel image)

other pathologic finding except for absence of pectoral muscles on the right side (Fig. 4). After informing the patient and his family, he registered for annual controls.

DISCUSSION

In 1841 Alfred Poland, when he was just a medical student, reported the results of a patient's anatomic dissection (1). In this patient, syndactyly existed together with absence of pectoralis major and pectoralis minor. The exact spectrum of concomitant anomalies for this syndrome was described by Thomson. Incidence is reported as 1/ 7.000-100.000 births (2). Subclavian hypoplasia was thought to be a reason although etiology was unknown. But David (3) and colleagues showed that subclavian hypoplasia is not a reason but a result. Chest wall involvement has an extended spectrum, from absence of pectoral muscles to especially the 2nd to 4th rib cartilage depression, hypoplasia or aplasia; breast involvement varies from total absence of breast to the absence of the nipple (4). Decrease of subcutaneous fat and absence of axillar hair on the affected side can be seen. Hand deformities described by Poland; hypoplasia (brachydactyly), adhesion of fingers (syndactyly) and claw hand may seen rarely. There is no correlation between thorax anomalies and hand anomalies. In our patient, the scar of the syndactyly surgery was made three years previously, and right sided absence of pectoralis minor and pectoralis major muscles was concomitant.

For surgical correction, evaluation of concomitant muscle and skelatal anomalies are very important. Computerized thorax tomography (thorax CT), is very important for evaluating the thoracal cavity and presence of concomitant pathologies (5). If only pectoral muscels are involved and deformity is not severe, surgical correction is not required. In our patient, surgical correction was not required because the deformity was not severe and it was not causing any physiologic disorder. However, especially in female patients, absence of the breast besides pectoral muscles is a very important cosmetic problem. So, some surgical techniques are used such as breast reconstruction and silicone implantation to subcutaneous tissue (6). Fokin and Robiscek (4) accepted advanced chest wall collapse, unguarded mediastinum, paradoxal movement of the chest wall, absence or decrease of breast tissue and cosmetic defects especially

in females, as surgical indications. If costal cartilages are absent or depressed, surgical correction of the chest wall must be considered. Thus the concave structure of the thorax will come to a neutral position and paradoxal movement will be blocked.

In 1966 Ravitch described the correction technique as wedge osteotomy, which allows rotation of the sternum by resection of unilateral costal cartilages by posterior cartilage deplacement and fixation with a Rehbein bar and Steinmann needle (7). In the absence of medial costal parts, reconstruction of the defective region is made possible by taking grafts from the other side (8, 9). If required, meshes can be used and rarely covered by latissimus flaps in selected cases, but it is not preferred.

In conclusion, Poland Syndrome is a rare chest wall anomaly. In this syndrome, in addition to chest wall anomalies, the other skeletal anomalies, absence of pectoral muscles, breast anomalies, axillary hair underdevelopment can be seen. Both of the genders paradoxal respiration is the result of costal agenesia, advanced malposition of sternum and in females cosmetic problems as a result of breast anomalies are surgical indications. In cases which do not require operation must be monitered and notified about sternal deformity.

Conflict of Interest

No conflict of interest was declared by the authors.

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