

Diagnostic value of chest computed tomography images in adult Poland syndrome: a report of two cases

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Abstract

Poland syndrome is a rare congenital developmental deformity characterized by unilateral agenesis or hypoplasia of thoracic wall soft tissue. We report two adult cases of Poland syndrome detected by computed tomography (CT) images. CT images of the two cases depicted an asymmetric chest wall with the absence of a breast and agenesis of the pectoralis muscles. A physical examination of case 1 showed a thin right chest wall with depression of the right nipple region. Hand deformities were also observed, including brachydactyly and syndactyly. In case 2, hand deformities were not found in a physical examination. Using multi-planar reconstruction, the size, position, origin, and termination of bilateral pectoral muscles could be compared symmetrically. For patients with Poland syndrome, a timely diagnosis and treatment are important. The use of chest CT in clinical practice could play an important role in the early diagnosis and treatment of Poland syndrome.

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Keywords

Poland syndrome, syndactyly, thoracic wall, multidetector computed tomography, pectoral muscle, breast, hand deformity

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Introduction

Poland syndrome (PS) is a rare, sporadic, congenital developmental deformity characterized by unilateral agenesis or hypoplasia of thoracic wall soft tissue. PS may be accompanied by hand deformities, such as ipsilateral brachydactyly and syndactyly.¹ PS in adult patients is occasionally detected by chest computed tomography (CT), in a physical examination, or for other reasons, and it is difficult to be distinguished from muscle deficiency of the chest wall after surgical resection. We report two cases of PS and discuss the importance of chest CT in

detecting adult PS. The reporting of this study conforms to the CARE guidelines.²

Case I

A 46-year-old man visited the hospital for a regular health examination. Chest CT showed agenesis of the right pectoral muscles and the absence of the right breast (Figure 1). His right pectoralis major muscle was small, and its maximal cross-sectional area was approximately one third to half of the left pectoralis major muscle. The right pectoralis minor

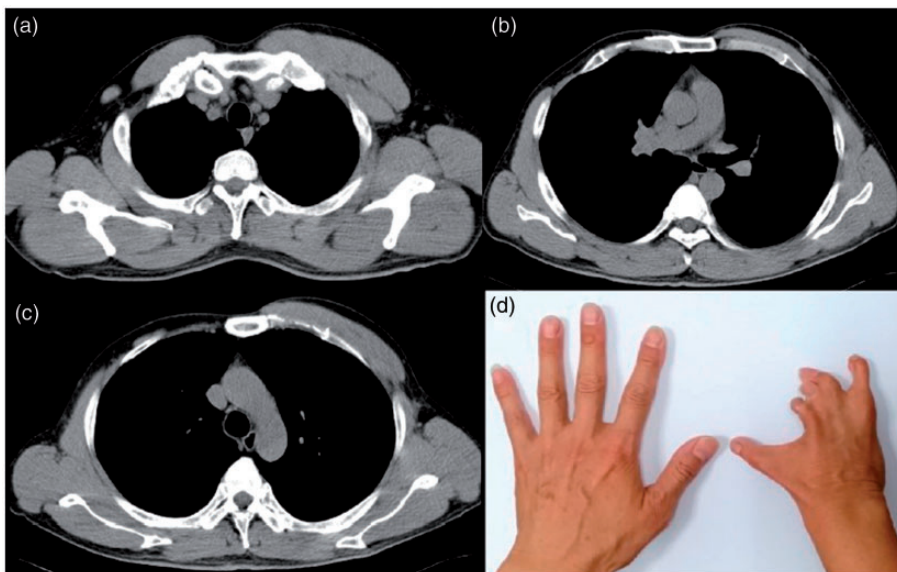


Figure 1. (a–c) Axial chest computed tomography images show an asymmetric chest wall with the absence of the right nipple and agenesis of the right pectoral muscles. (d) A physical examination shows agenesis of all fingers, except for the thumb, and syndactyly of the third and fourth fingers.

was shaped like a bar, and it was located on the lateral side of the right pectoralis major muscle. Additionally, the size of the right anterior serratus was smaller than that of the left. The left nipple and breast were normal, while the right nipple and breast could not be detected in axial images.

The patient reported that his right chest wall had been thinner than the left since birth. There was an areola in the region of the right nipple with depression in the center. Ipsilateral hand deformities were also found at birth, including agenesis of all fingers, except for the thumb, and syndactyly of the third and fourth fingers. The patient had sought consultation for his hand anomalies as a teenager. However, because basic function of his fingers could meet most of his daily needs, his parents decided not to have orthopedic surgery performed.

Case 2

A 69-year-old man was admitted to the Department of Infectious Diseases because

of a fever. Chest CT showed hypoplasia of the right pectoral muscles with the absence of the ipsilateral breast (Figure 2). Scoliosis of the upper thoracic segment and slight depression of the upper right chest wall were detected through multi-planar reconstruction. The right pectoralis major muscle had a smaller size, of which the vertical and transverse diameters accounted for two thirds of the left pectoralis major muscle. Although bilateral pectoralis minor muscles started from the coracoid process, the left pectoralis minor muscle was located on the left third and fourth anterior ribs, while the right was on the right first and second anterior ribs. The width of the left erector spinae was smaller than that of the right. The left breast showed mild hyperplasia with a normal nipple, while the right breast and nipple were absent. Additionally, the right acromion was lower than the left. The patient reported that the right chest wall was thinner than the left, with the absence of the right breast and nipple since birth. A hand deformity

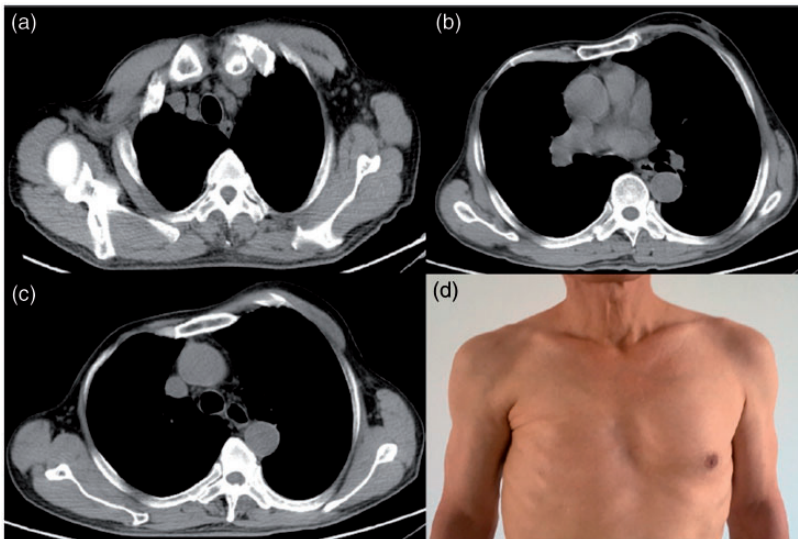


Figure 2. (a–c) Axial chest computed tomography images show an asymmetric chest wall at the level of the lower segment of the pectoral muscles. (d) A physical examination shows thinner soft tissue in the right chest region than in the left chest region, with the absence of the nipple.

was not observed. The patient had not visited the hospital for treatment because of normal function of the chest wall.

Discussion

PS, also known as “Poland anomaly”, was first reported by Alfred Poland in 1841.³ Patients affected by PS can be identified at birth, and most cases have been reported as sporadic. This condition is more frequent in boys/men (male to female ratio is 2:1). Ohn and Ohn reported a case of PS mimicking a traumatic chest injury.¹ The etiopathogenesis of PS is still unknown, but interruption in the circulation of the subclavian artery and its branches at the end of the sixth week of embryonic development may lead to PS. Aplasia or hypoplasia of unilateral pectoral muscles, the absence of ipsilateral subcutaneous fat, and the absence or hypoplasia of the nipple and breast on one side of the body are typical characteristics. PS is more frequently found in the right side of the thorax (75%),⁴ while patients with PS on the left side usually also have dextrocardia. Additionally, deformities of the chest wall soft tissue, the absence or hypoplasia of the pectoralis minor, deltoid, anterior serratus, external oblique, or latissimus dorsi muscle, bone and cartilage defects of the ribs, clavicle, and sternum, Sprengel deformity, scoliosis, and lung herniation may be observed. PS may be associated with hand deformities, including partial or complete syndactyly and brachydactyly. The absence or hypoplasia of the carpal bones or metacarpals, absence or attenuation of flexor or extensor tendons, shortening of the humerus, radius, or ulna, constriction bands, or polydactyly nail agenesis may also be observed.⁵

The surgical treatment of PS should be performed as early as possible, and this is recommended between 18 and 24 months after birth to avoid severe functional injury. Functional rebuilding and cosmetic

shaping are the main purpose of a surgical intervention. However, many patients with PS are not diagnosed at birth. A previous study reported that 68.42% (39/57) of patients were not diagnosed at birth, and the average age of patients with PS at diagnosis was 14 years.⁶ Although case 1 had visited an orthopedic clinic once for his digital deformity, he was not diagnosed with PS because the doctor was not informed that he also had ipsilateral hypoplasia of soft tissues of the chest wall. Similarly, case 2 was not diagnosed until he had a chest CT scan for a fever. The confidence and self-consciousness of the body in our two patients could have been improved if an early diagnosis and timely treatment had been performed.

An increased attenuation of the right upper chest is found in most patients with PS by posteroanterior radiography. Although surgical resection can be ruled out through inquiry, the real cause of the lower density of the unilateral chest is not identified if radiologists are not familiar with PS. With the development of imaging technology, low-dose chest CT is widely used in detecting respiratory diseases and for regular medical examinations. Chest CT also enables a clear diagnosis and can guide treatment of PS through observing axial images or any directional reconstructed images symmetrically. Chest CT can detect the presence of the pectoralis major, pectoralis minor, or anterior serratus muscle to identify changes in chest wall soft tissue after surgical resection. The size, position, origin, and termination of bilateral pectoral muscles can also be determined with CT images to identify the severity of chest wall soft tissue dysplasia.

In conclusion, we report two patients who were diagnosed with PS on the basis of dysplasia of the chest wall, which was accidentally detected by chest CT images. Axial chest CT images showed an asymmetric chest wall with the absence of the right

nipple and agenesis of the right pectoral muscles. One of the two case had agenesis of all fingers, except for the thumb, and syndactyly of the third and fourth fingers. Our findings suggest that radiologists should pay more attention to this rare congenital anomaly in clinical practice. An early diagnosis and timely treatment are helpful to improve the quality of life in patients with PS.

Declaration of conflicting interest

The authors declare that there is no conflict of interest.


Ethics statement

The study protocol was approved by Hebei Medical University Ethics Committee (approval number: 2020-168). Written consent was obtained from the patients or the patients' parent/carer.

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