

Poland's Syndrome: a Cadaver Case Study

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ABSTRACT

We observed a rare variant case of Poland syndrome in a 55-year-old male cadaver in gross anatomy dissection room at department of Anatomy school of Medicine. His past and family history was not remarkable.

The outward appearance of anterior thoracic wall, deltoid and shoulder regions revealed obvious abnormalities. Upon dissection, the following features were observed: (1) On the right side, the pectoralis major and minor were completely absent and deltoid muscle was hypotrophied; therefore, the deltoid region in the right side was seen weaker in comparison with the opposite side. (2) A thick fatty layer about 3 cm was replaced instead of both the pectoralis major and minor muscles. (3) The distance between the neck and the upper part of deltoid region was seemed shorter on affected side (right side) ; in other words, the right clavicle bone was seemed a little shorter than the left clavicle bone. (4) The fingers of the right hand show webbing. Due to the absence of the pectoralis major and minor muscle that were accompanied with webbing and shortening in fingers of the hand of same side, the presented case was a rare variant of the Poland syndrome.

1. Introduction

Poland's syndrome is a rare congenital anomaly that is a very mysterious disorder; its prevalence in boys is 3 times more than girls (1) . The syndrome more often affects the right side of the body, and occurs in about one of every 30/000 live births (2) .

This syndrome is a frequent disease that its cause is relatively unknown, especially in its epidemiology and aetiopathogenetic aspects. The clinical features including

absence of the sternocostal portion of the pectoralis major muscle, hypoplasia or aplasia of the ipsilateral breast or nipple, hypoplasia of subcutaneous tissue, abnormalities of the rib cage, and upper extremity anomalies (3) .

The right side of the body nearby chest muscle (The serratus anterior and latissimus dorsi muscles) may also be missing; the breast on that side is also usually absent in girls (1) .

In addition, the fingers show webbing and shortening (Symbrachydactyly) on hand of the same side .

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For the first time, the syndrome was described by Alfred Poland in Guy's hospital in 1841.

2. Results

We presented a case study of a rare variant of the Poland syndrome in a 55-year-old male cadaver in dissection laboratory for medical students. The subject was relatively obese with about 90 kg weight, and adipose deposits concentrated in the abdominal wall, around his heart and kidneys. On the right side of the chest, the entire pectoralis major and minor muscles were absent (figure 1), and a thick fatty tissue layer was replaced instead.

The deltoid muscle on the same side was hypotrophied. The length of shoulder was seen a little shorter in the right side, and the length of right clavicle bone was also shorter than the left clavicle and the fingers were webbing. His kidneys had some abnormalities in the shape and veins, so that they were incompletely segmented and there were more than one vein, which deranged blood from kidneys. These veins formed a network near the hilum of kidneys and deranged to unusual places, such as testicular or lumbar veins (figure 2). The same contralateral muscles such as pectoral, deltoid and anterior serratus muscles were hypertrophied.

3. Discussion

The exact etiology of Poland's syndrome is unknown. Some studies have suggested that the primary defect in the development of the proximal subclavian artery with diminished blood flow to the affected sides leading to

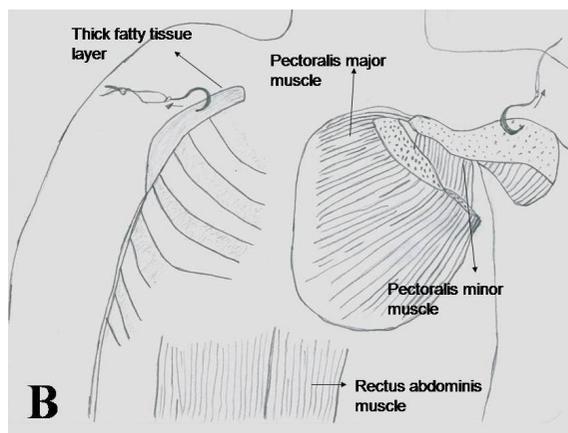
partial loss of tissue in that region. The others have proposed a mechanism that is inheritance, but this is not universally accepted (2). Poland syndrome as mentioned before consists of a deficiency of subcutaneous fat and muscles on one side of the body, it may include underdevelopment of the arm, hand, and fingers on the same side (4,5), and maybe associated with other conditions such as kippel- Feil syndrome (6).

Poland syndrome has several distinctive symptoms such as chest muscle deformities, absence of the pectoralis muscles, underdeveloped or missing ribs, underdeveloped arm, hand and fingers, and abnormally short and webbed fingers.

Unilateral defect of pectoral muscle and ipsilateral syndactyly constitute Poland syndrome.

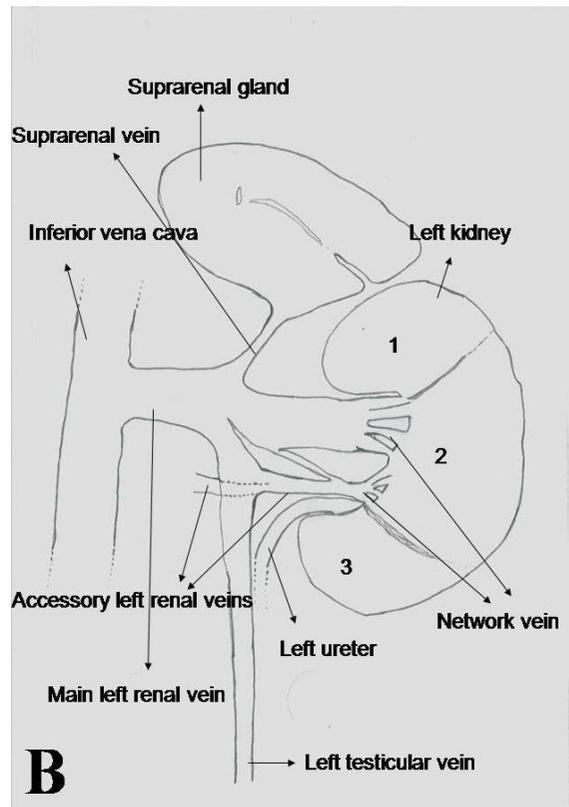
If Poland syndrome is identified in individuals, they should be examined for presence of other syndromes that mentioned above.

The combination of a lack of the pectoralis major and minor with skeletal, vascular, and surface feature anomalies in the ipsilateral upper limb has been referred to as Poland's syndrome. Upon rare occasions, the Poland's syndrome is associated with more severe finger and arm involvement or vertebral or kidney problems (7). Due to some symptoms in our case study, observations in figures, absence of pectoral muscles in right side and abnormalities of kidneys are similar to a Poland's syndrome; so, we named it as a rare variant of Poland's syndrome. In this case, the hypertrophied contralateral muscles may be caused secondarily by more using them



ANATOMICAL SCIENCES

Figure 1. a: Dissection of the pectoral region revealed the absence of the pectoral muscles in the right side and they were replaced with a thick fatty tissue layer. b: Schematic illustration of the defects shown in previous figure (figure a) depicts the considered parts. In unaffected side (left side), pectoralis major muscle has been cut to show pectoralis minor muscle lying in under of it.



ANATOMICAL SCIENCES

Figure 2. a: Dissection of the abdominal cavity revealed some abnormalities in the kidneys, such as incomplete segmented kidneys and in addition to main renal vein, each kidney had two or three accessory renal vein which deranged to unusual places, for example to testicular vein or lumbar veins. b: Schematic illustration of the abnormalities of the left kidney. 1, 2 and 3 show incomplete segments of the kidney. Accessory renal veins link to testicular or lumbar veins.

in comparison with the involved side (right side) and their compensatory role in some activities.

References

1. Mosconi T, Kamath S. Bilateral asymmetric deficiency of the pectoralis major muscle. *Clin Anat.* 2003 Jul; 16 (4) :346-9.
2. Allam SR, Yadav R, Meziane M, Mehta AC. A middle-aged man with asymptomatic chest wall asymmetry. *Cleve clin J Med.* 2006 Aug; 73 (8) :754-6.
3. Hsieh WB, Leung C. Poland syndrome in a neonate: report of one case. *Acta paediatr Taiwan.* 2005 Jul - Aug; 46 (4) :239-43.
4. Legbo JN. Poland's syndrome: report of a variant. *J Natl Med Assoc.* 2006 Jan; 98 (1) :97-9.
5. Riyaz N, Riyaz A. Poland syndrome (anomaly) with congenital hemangioma: a new association. *Indian J Dermatol venereol Leprol.* 2006 May- Jun; 72 (3) :222-3.
6. Özçakar L, Kaymak B, Vanderstraeten G. Klippel-Feil Syndrome presenting with bilateral thoracic outlet syndrome. *Spine.* 2005 Jun 15; 30 (12) :1473-4.
7. Seyfer AE, Icochea R, Graeber GM. Poland's anomaly. Natural history and long-term results of chest wall reconstruction in 33 patients. *Ann Surg.* 1988 Dec; 208 (6) :776-82.