CONGENITAL ABSENCE OF PECTORAL MUSCLE: A CASE REPORT

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ABSTRACT: Congenital absence of the pectoralis muscles is unusual, although it is the most common of all congenital muscle anomalies. This entity is included under the syndromic manifestations of Poland syndrome, which was originally described by Alfred Poland in 1840.¹ This is a case report of a 25 year old female patient who presented with complaints of underdevelopment of right breast in our department. On imaging studies, this turned out to be isolated absence of pectoralis muscle with rib deformity.

KEYWORDS: Poland Syndrome, Congenital Absence of Breast, Pectoralis Aplasia.

HOW TO CITE THIS ARTICLE: Suresh Kumar, Mangilal Burman. "Congenital Absence of Pectoral Muscle: A Case Report." Journal of Evolution of Medical and Dental Sciences 2015; Vol. 4, Issue 94, November 23; Page: 16010-16012, DOI: 10.14260/jemds/2015/2335.

INTRODUCTION: Absent pectoral muscle, although the most common muscle anomaly has a rare incidence of about one in 30,000 to 50,000.² This entity is included under the syndromic manifestations of Poland syndrome, which was originally described by Alfred Poland in 1840.¹ Pectoral muscle is an important muscle in shoulder movements. Its absence can lead to functional disability of shoulder girdle and/or can be aesthetically displeasing, especially if associated with breast aplasia. Hence, diagnosing this entity with appropriate treatment is necessary.

KEY MESSAGES: Any patient presenting with flattening of chest should be completely evaluated to know which components forming the chest wall is hypoplastic or aplastic, as it is important in management. If there is partial absence of muscle, the remaining muscle can be developed by physical therapy. In case of complete absence of muscle and/or breast, the reconstruction surgery (Latissimus dorsi muscle flap and/or silicone breast implants) plays a role.

CASE HISTORY: A 25 year old female patient presented in Plastic Surgery Outpatient Department with complaints of undersized right breast since puberty. For further assessment, she was referred to our department for imaging purpose. On acquiring her detailed clinical history, she complained of deficient right breast since puberty leading to flattening of chest. However, she had borne two children whom she had breast fed from both breasts with equal frequency.

Four quadrant volumetric measurement of bilateral breast with high frequency ultrasound showed equal volumes of bilateral breast tissue. Corresponding quadrants of each breast had a volume between 18 to 24cc. However, there was near complete absence of pectoralis muscle beneath the breast parenchyma on right side with normally developed left pectoral muscle (Fig. 1). She was further subjected to computed tomography (CT) examination of chest for exact delineation of her incongruity.

Fig. 1: Ultrasound image showing Normal Pectoral Muscle (Black square) on Left Side, with Absent Pectoral Muscle on Right Side. The Breast Tissue (Red square) is Bilaterally Normal And Symmetric.

Scout scan revealed hypertransradiant right hemithorax with hypoplastic second to fourth right anterior ribs (Fig. 2). Serial helical scans of upper chest revealed that there was complete absence of both sternal and clavicular parts of right pectoralis major and pectoralis minor muscle. This was associated with hypoplastic, thinned anterior second to fourth ribs. As seen on high frequency ultrasound, the bilateral breast parenchyma was present and in equal volume (Fig. 3).

Financial or Other, Competing Interest: None.

Submission 13-11-2015, Peer Review 14-11-2015,

Acceptance 16-11-2015, Published 23-11-2015.

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DOI:10.14260/jemds/2015/2335.

Fig. 2: Scout image of the Chest showing Hypertransradiant Right Hemithorax with Hypoplastic Second to Fourth Ribs Ipsilaterally.

Fig. 3: CT image at Mid Thoracic Level showing Absent Pectoralis Muscle on Right Side.

There was no associated other congenital abnormality of limbs. The remaining muscles did not show compensatory hypertrophy. Hence, the diagnosis of congenital absence of pectoralis major and minor muscle with rib hypoplasia was made. Thereafter, she was referred back to the Plastic Surgery Department for reconstructive surgery.

DISCUSSION: Named after Sir Alfred Poland, Poland syndrome includes the features of ipsilateral breast and nipple hypoplasia and/or aplasia, deficiency of subcutaneous fat and axillary hair, absence of the sternal head of the pectoralis major muscle, hypoplasia of the rib cage, and hypoplasia of the upper extremity.¹ The main component of this anomaly is absence of the pectoralis major muscle.³

In 1841, Sir Alfred Poland described this chest wall anomaly in the Guy's Hospital Gazette while still a medical student based on findings of one cadaver dissection. In his original description titled "Deficiency of the pectoral muscles," he specifically noted absence of the stern costal

portion of the pectoralis major muscle with an intact clavicular origin, absence of the pectoralis minor muscle, and hypoplastic serratus and external oblique muscles.

Poland did not outline the breast hypoplasia or hand deformities in his original description. Additional features of Poland syndrome include hypoplasia or aplasia of any other muscle, total absence of anterolateral ribs and herniation of lung. Symphalangism with syndactyly and hypoplasia or aplasia of the middle phalanges can also be observed.⁴ The incidence of Poland syndrome is between one in 30,000 to one in 50,000,² and right side being affected more than left side.^{5,6} Some studies have reported the female-to-male ratio to be one:three,⁵ whereas other studies have suggested that both genders are affected equally.⁷

The theory put forth is interruption of early embryonic blood supply to subclavian arteries, the vertebral arteries and/or their branches. This critical vascular event known as subclavian artery supply disruption sequence (SASDS), occurs when the medial and forward growth of the ribs forces the subclavian vessel into a U-shaped configuration. A combination of the blockade of various branches could lead to Poland syndrome along with its variants.

These vascular disruptions could also lead to its relation with syndromes like Sprengel, Klippel-Fiel and Adams-Oliver syndrome.^{8,9} Because the functional disability in Poland syndrome is mild, patients usually present later for evaluation and discussion on aesthetic options. Preoperative evaluation should include a thorough history and examination.

During the examination, the stage of breast development and status of the latissimus dorsi muscle should be noted. Reconstructive surgery is the main course of treatment and includes latissimus dorsi muscle flap and silicone breast implants to give the chest a normal shape and aesthetic value.¹⁰ Also physical therapy may help to develop compensatory muscles of the shoulder girdle and preserve function of the shoulder girdle.

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