

Bilateral pectoral musculature malformations with concomitant vascular anomaly

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We report on a unique combination of multiple variations concerning the pectoral muscles and the left external jugular vein. Specifically, a bilateral hypoplasia of the medial clavicular portion of the pectoralis major muscle was noticed along with the coexistence of total right pectoralis minor aplasia, substituted by loose connective and fatty tissue. Simultaneously, a supernumerary anterior-placed external jugular vein was found, which, after its supraclavicular course, pierced the interval between the left clavicular and the sternocostal head, and drained into the left jugular junction. The combination of the above anomalies constitutes an atypical pattern of Poland syndrome. We discuss the related embryological development and the relative literature. Attention was paid to the clinical importance for plastic surgeons, general surgeons, and radiologists, facilitating them with accurate interpretation of anterior thoracic wall findings. (Folia Morphol 2010; 69, 3: 187–191)

Key words: pectoral muscles, defects, vein anomaly, Poland syndrome, pectoralis

INTRODUCTION

Congenital malformations of the major and minor pectoralis muscles, either in combination or as sole variants, have an incidence from 0.009% [2] to 0.061% [21]. Pectoral musculature defects are of great value to the physician, since pectoralis major as well as minor muscle may serve as favourable musculocutaneous or osteomusculocutaneous free muscle flaps [25, 26]. Clinicians must be aware of such muscular defects in order to facilitate the selection of the preferable portion of the pectoralis major as free or turn-over flap since in various heads of the pectoralis major such as the sternocostal, developmental deficits usually appear [5, 25]. However, pectoral muscle defects can be associated with various muscular, skeletal, and vascular anomalies. The combination of the above anomalies constitutes Poland syndrome. The syndrome was firstly described by

a Guys Hospital student, Alfred Poland, who was occupied as an anatomy demonstrator [23]. The syndrome can coexist with various other congenital deficiencies such as Mobius syndrome and vocal fold paralysis [1]. Moreover, other reported anterior chest wall and shoulder girdle anomalies include the axillary arch [21], chondroepitrochlearis muscle [13], rectus sternalis muscle [12], and pectoralis minimus muscle [28, 32].

In the current study we describe a unique combination of bilateral deficits of pectoralis major muscles, aplasia of the right pectoralis minor muscle, and abnormal course of a supernumerary left external jugular vein. Familiarity with such anatomical variants could assist physicians in the diagnosis of atypical as well as typical patterns of Poland syndrome. Plastic surgeons should also be aware of this trait during total or segmental pectoral muscle free-flap transfer harvesting, and radiologists would also

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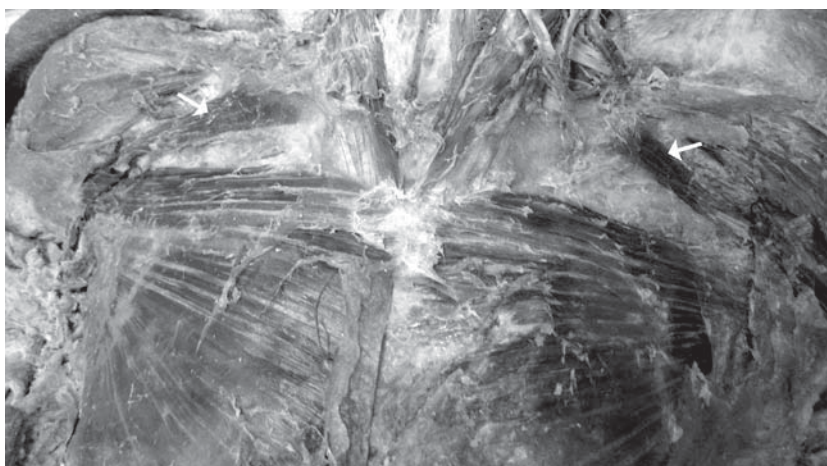


Figure 1. Bilateral hypoplasia of the clavicular heads of the pectoralis major muscle. On the right side, the inferior-medial half of the clavicular head is absent, while on the left side, the inferior-medial three-quarters of the clavicular head is aplastic. The residual clavicular heads on both sides are shown (arrows) as well as the wide gaps between the clavicular and sternocostal heads.

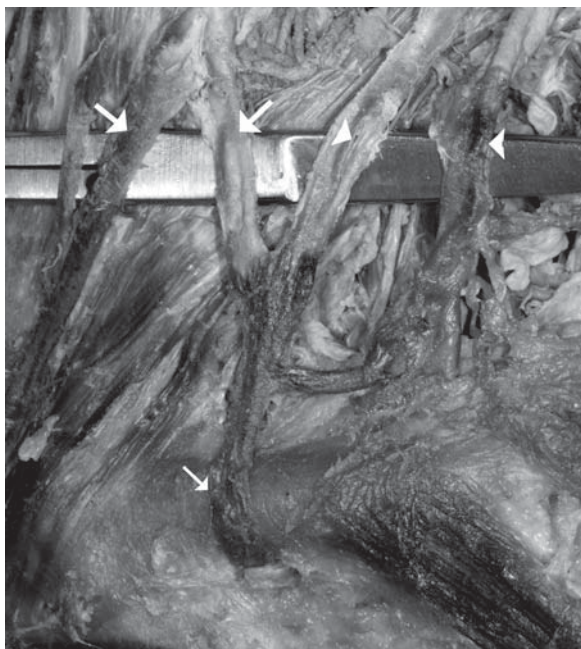


Figure 2. On the left side, two anterior (arrowheads) and two external jugular veins (large arrows) are shown. The anterior-placed accessory external jugular vein is demonstrated directing supraclavicularly (small arrows) and draining into the left jugular junction.

benefit from such knowledge in the precise study of anterior chest wall radiological imaging.

CASE REPORT

During a routine dissection in our Gross Anatomy Laboratory of a 76 year-old female formalin/phenol/ /alcohol embalmed cadaver used for educational purposes, we noticed the presence of the following mul-

iple variations: 1) An extended gap situated between the hypoplastic clavicular and the normal sternocostal head of the right pectoralis major muscle, the latter being covered by thick fibrofatty tissue. No vessel or nerve was noticed perforating this tissue, and the gap of the absent portion of the clavicular head concerned the inferior half of a normal sized clavicular head; 2) A greater interval than the previous one was noticed between the atrophied clavicular head and the sternocostal head of the left pectoralis major muscle. The atrophied clavicular head occupied the lower three-fourths of a normal sized clavicular head (Fig. 1). The interval between the mentioned muscular heads was covered by dense fibrofatty tissue pierced just inferior to the midpoint of the clavicle by a left anterior-placed accessory external jugular vein deriving from a pair of left external jugular veins. The supernumerary external jugular vein drained into the left venous junction after a short supraclavicular course (Fig. 2); and 3) After resection of the outer portion of the right pectoralis major muscle, a total aplasia of the underlying right pectoralis minor muscle was observed (Fig. 3). Only loose connective and fatty tissue without any nervous branches was found replacing the missing muscle. After removal of the fibrofatty tissue, the underlying vessels and nerves of the axillary cavity were exposed. It is worth mentioning that the cadaver appeared without any visible chest wall and axilla malformations nor any pathological condition in both upper limbs. We knew from the cadaver's medical history that no movement disorders in the shoulder region had existed.

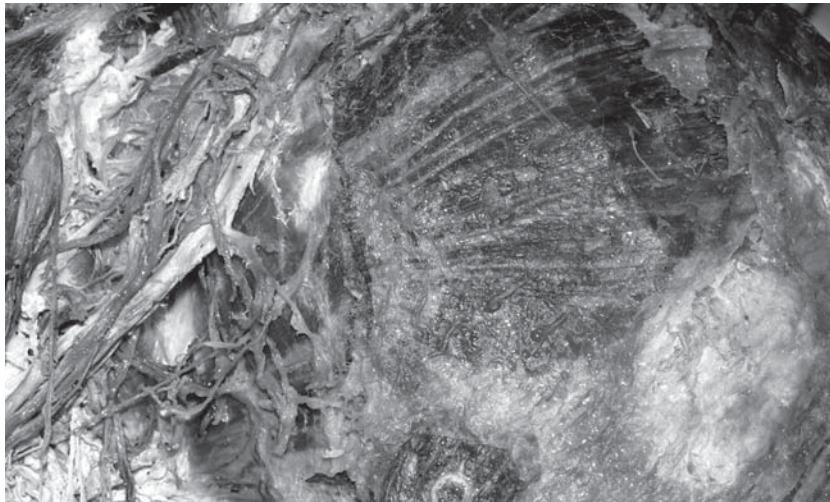


Figure 3. Absence of the right pectoralis minor muscle after resection of the outer portion of the right pectoralis major muscle. The underlying axillary artery and brachial plexus divisions are illustrated after resection of the fibrofatty tissue covering them.

DISCUSSION

Among the ventral muscles of the shoulder girdle, the pectoralis major and minor muscles are the most important. The sternocostal head of the pectoralis major converges to the upper humerus by a trilaminar tendon. The clavicular head originates from the median half of the clavicle and courses almost horizontally while laterally it lies in a groove at the manubrial part of the muscle. The sternocostal head arises from the lateral portion of the anterior surface of the sternum as well as by a series of fibres from the upper six costal cartilages. Moreover, an abdominal slip or head originates from the aponeurosis of the external oblique muscle. The pectoralis minor arises from the 3rd, 4th, and 5th ribs just beneath the pectoralis major muscle and converges in a triangular pattern before insertion into the coracoid process [18].

Pectoral muscles derive from the outer of the three primitive body wall sheets. During pectoralis major embryological development, as well as in most shoulder girdle muscles, the exact proportion of the upper limb or trunk myotome migration is still not assured [5, 18]. In the 5-week embryo, the pectoral pre-muscle mass derives from the 2nd rib and from the proximal portion of the humerus. The median portion is attached to the clavicle and corresponds to the bases of the origin of the clavicular and sternocostal heads [15, 31]. The pectoralis minor in the 5-week embryo arises from the same previously mentioned pre-muscle mass along with the pectoralis major anlage. The pectoralis minor muscle is separated into its three bellies, while the pectoral pre-muscle mass growth is directed inferiorly [15].

The clavicular fibres are usually separated from sternocostal fibres by a slight cleft [30].

Various malformations of the pectoral musculature have been reported so far. There are publications discussing the attachments between pectoral muscles and costal cartilages as well as the number of costal attachments and the separation pattern of clavicular and sternocostal heads [4, 18, 34]. Congenital abnormalities concerning the pectoralis major with or without involvement of the pectoralis minor muscle are very rare, incidence in the European population being 0.013% and 0.061% in the Japanese population [21]. Other researchers report pectoral malformation incidence as 1:11000 [2] and 1:5000 cases [10]. From a review of the literature we found reports about at least 23 cases with pectoralis major agenesis: 12 out of 23 were Bing's cases [5], 3 cases by Takeya [30] and Yabashi et al. [35], and the remaining 8 cases were reported solely by others [3, 10–12, 14, 20, 23, 24].

The majority of investigators showed that the most common pectoralis major variant is hypoplasia or aplasia of the sternocostal head coexisting with hyperplasia of the clavicular head [1, 3, 5, 10, 11, 36]. Kitamura et al. [12] reported that it is impossible for clavicular head deficiency to occur. However, reported incidence of clavicular head defect by Tsukuda [31] was 4.3% and by Bing [5], 6.9%. Moreover, Takeshige et al. [29] presented a quite rare clavicular and abdominal head deficiency variant.

There are a few reports regarding pectoralis minor muscle malformations in the literature. Yamasaki [36] observed one case with pectoralis minor mus-

cle agenesis as well as another in which the muscle was substituted by a string-like muscle band. Mosconi and Kamath [20] commented on a case in which the pectoralis minor was poorly developed and represented as a fibrous fatty muscle band arising from the 2nd to 5th ribs and inserted into the coracoid process. The coexistence of pectoralis major and minor aplasia is very rare [11]. Irvine and Tilley [10] reported three cases of deficient pectoralis minor along with abnormal pectoralis major muscles. Yamasaki [36] noticed the absence of pectoralis minor, while the pectoralis major was substituted by a fatty membrane. Due to the absence of nerve branches within the membrane, Yamasaki [36] concluded that the membrane was not a degenerated pectoralis minor muscle. It seems that individuals with abnormal pectoralis muscles do not show any significant disorders during shoulder movement in daily activities [14].

To our knowledge, the reported case constitutes a valuable and probably unique case based on a combination of bilateral hypoplasia of the clavicular head of the pectoralis major muscle and the absence of the right pectoralis minor muscle and supernumerary left external jugular vein that after its supraclavicular course pierces the connective tissue in the cleft between the clavicular and sternocostal head of the left pectoralis major. The presence of accessory anterior and external jugular veins as well as the variability in their size is well documented in the literature [8, 27]. The superficial jugular veins appear at the 8th week of gestation as extraneous vessels that develop independently and attach secondarily [19]. The coexistence of pectoral muscle defects with other anomalies such as hypoplasia of the breast or the nipple, subcutaneous tissue hypoplasia, absence of the upper ribs, or brachysyndactyly has been described as Poland syndrome [22, 23]. The simultaneous appearance of all the above features of Poland syndrome in one patient is very rare [16]. In our case, no skeletal deformities were noticed, but the combination of pectoral muscle deficits along with vascular anomalies could be considered as an atypical pattern of Poland syndrome.

Regarding the likely model of embryological development of such anatomical variants, we consider that during the 8th gestational week the external jugular vein anlage may provide new formations, and thus multiple superficial jugular veins [19]. This may shift the vein's formation direction to a supraclavicular course, thus penetrating the upper part of the pectoralis major muscle. It is possible that

the existence of a supraclavicular accessory external jugular vein restrains the growth of the supraclavicular head of the pectoralis major. The incidence of bilateral hypoplastic clavicular heads is very rare since these heads are proximally positioned and develop earlier than the other portions of the pectoralis major muscle [20]. We believe that the intervention of the accessory external jugular vein explains such a deficiency in the left clavicular head, while the left head atrophies since it follows similar development to that of the contralateral side without an intervening vein.

Anomalies of the pectoralis major muscle are of prominent interest for plastic surgeons because that muscle is harvested during total, segmental, or turn-over flap graft removal for coverage of major sternal wound infections after cardiac surgery [7], breast reconstruction [6], or local mediastinal wounds [25] and may serve as treatment for a paralytic elbow [17]. Modern single-stage Poland syndrome management includes reconstruction of the anterior chest wall, augmentation mammoplasty, and transfer of a myocutaneous flap [33]. Moreover, both pectoralis major and minor muscles can be used as osteomusculocutaneous flaps for reconstructive surgery of the head and neck or other distant region microsurgery [26]. Furthermore, the pectoralis minor is useful as a free-flap in cases of facial palsy [9]. All surgeons advancing to the surgical exposure of the chest wall, as well as radiologists, should be aware of such malformations with respect to the precise anatomy of the shoulder girdle region.

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