Aesth. Plast. Surg. 27:112–115, 2003
DOI: 10.1007/s00266-003-0108-8

Aesthetic Plastic Surgery

Mild Degree of Poland's Syndrome Reconstruction with Customized Silicone Prosthesis

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Abstract. Poland's Syndrome consists of unilateral absence of the pectoralis major muscle, ipsilateral symbrachydaktylia, and occasionally associated other malformations of the anterior chest wall and breast. Mild Poland's Syndrome is characterized by asymmetry of the breasts with partial absence of the pectoralis major muscle. This report describes a 19-year-old man with unilateral hypoplasia of the breast, absence of the anterior axillary fold, and absence of the pectoralis major muscle. The patient's chest wall was reconstructed with a customized silicone prosthesis. This reconstructive technique is a simple alternative to more complex surgeries or muscle transposition.

Key words: Pectoralis major muscle—Poland's Syndrome—Customized silicone prosthesis

The absence of some muscles, such as the pectoralis major, serratus anterior, and quadratus femoris, have been described [9]. However, deficiency of the pectoralis major muscle (PMM) is relatively common, agenesis of the PMM is most often partial [5,11,13]. This anomaly is often one component of a syndrome associated with ipsilateral upper limb (Poland's Syndrome) and thoracic anomalies [4]. Deformity upper limb and ipsilateral side of the thoracic cavity on the same side as the missing PMM have been described as components of Poland's Syndrome. Genitourinary, spine, cardiovascular, and other abnormalities sometimes coexist in this syndrome, making it a polymorphous syndrome with differing degrees of severity [18]. Severe forms that affect thoracic cavity and upper extremity may be detected at birth and require constructive surgery. But mild forms may go undiagnosed. Hypoplasia of one breast or anterior axillary fold may be sole clinical manifestation [15]. Unilateral breast hypoplasia and the absence of anterior axillary fold cause a contour deficit of the anterior chest wall [7]. Using customized silicone prosthesis, accurate contour restoration can be achieved with less morbidity and without donor defects.

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Case Report

A 19-year-old man was complaint of asymmetry of chest wall. On physical examination, his right anterior chest wall was flat and the right anterior axillary fold was absent. The patient had smaller, asymmetric nipples (Fig. 1). The right axillary hair was sparser than that on the left side. There was no visible anomaly for right upper extremity. His past and family history were not remarkable. Morphological and functional abnormalities in the ipsilateral upper limb were not exhibited. In the posteroanterior chest radiogram, the soft tissue volume was smaller for the right axilla than on the other side and a hypoplasic second rib was seen on the right side (Fig. 2A). Computerized tomography findings in the patient also demonstrated the absence of the sternocostal fascicle and the presence of the sternoclavicular fascicle (Fig. 2B).

A plaster mold of the chest and breast deformities was made of the patient prior to the surgery. Plaster was added to the skin surface of the affected side until it closely matched the normal, contralateral side. We tapered the plaster moulage at the edges to camouflage the implant in its subcutaneous position. The cast was sent away to a company that made a customized prosthesis based on the moulage.

The customized prosthesis, which was produced within one month, was placed into a subcutaneous

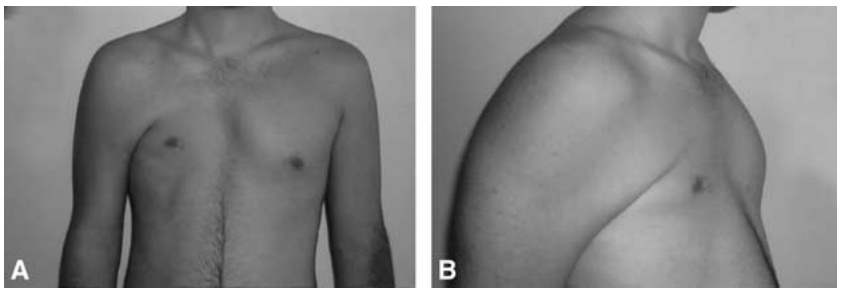

Fig. 1. (A) Preoperative anterior view of the patient. **(B)** Preoperative lateral view of the patient.

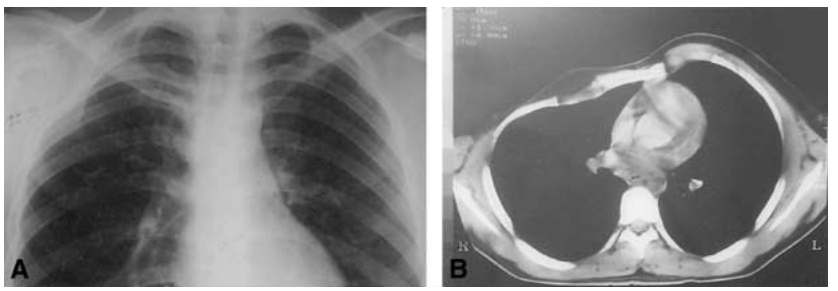

Fig. 2. (A) Chest radiographic view of the patient. **(B)** CT view of the patient.

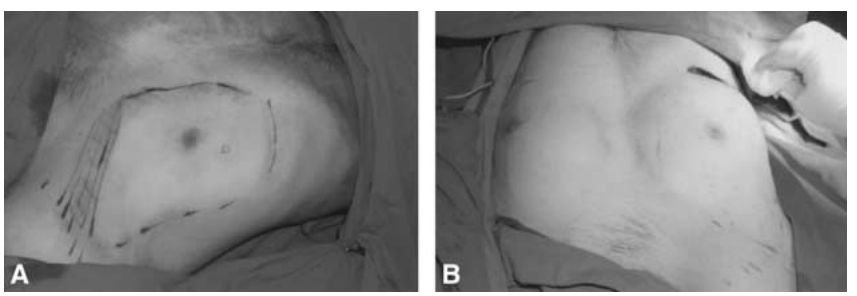

Fig. 3. (A) Intraoperative view of the patient. **(B)** Intraoperative view of the patient after prosthesis was inserted.

pocket through a 6-cm horizontal inframammary incision. The subcutaneous dissection extended beyond the anticipated limits of the PMM using the contralateral muscle as a guide (Fig. 3A). The sternoclavicular fascicle of PMM was seen and muscle origin was released and rotated inferiorly based on insertion. The muscle remnant was sutured to the periosteum of the fifth and sixth costae. The prosthesis was inserted in the pocket (Fig. 3B). A drain was placed into lateral side of the pocket. Subcutaneous and skin layers were approximated. A simple compressive bandage was placed around the implant for 72h.

Postoperatively, the drain was removed in three days. The implant appeared to be mobile within the pocket and a compressive bandage was placed lateral to the implant. On the first postoperative day, the implant was pushed from the lateral to the medial portions of the space every few hours. Prothesis was not malpositioned. Good contour was achieved (Figs. 4 and 5).

The patient is satisfied with aesthetic result of the treatment. He is continuing expansion exercises. No complications were seen in 13 months of followup. Fine aesthetic results were achieved.

Discussion

Deformities of the upper limb and ipsilateral side of the thoracic cavity on the same side as the missing PMM have been described as components of Poland's Syndrome, which was described by Alfred Poland in 1841 [15]. The etiology of this syndrome is unclear. Most of cases have been sporadic [4]. Disruption of the normal development of the subclavian artery, and defective migration of fetal mesodermal tissues are possible causes [1,2]. The incidence has been estimated as 1:16500 live births [6,13], the ration of men to women is 3:1 [14,18], and 75% of patients have the right side affected [13].

Clinical manifestation of this syndrome is polymorphic with differing degrees of severity. Minor manifestations have received less attention and may easily go unnoticed, but severe forms are detected at birth. Genitourinary, spine, cardiovascular, and other abnormalities are possibly associated [18]. The most consistent deformity is a deficiency or absence of the PMM. Hypoplasia of the breast, nipple, ribs, and chest skin may be seen with this syndrome [5,20].

Among the muscles congenitally absent, the pectoralis major is the most frequently involved. In the

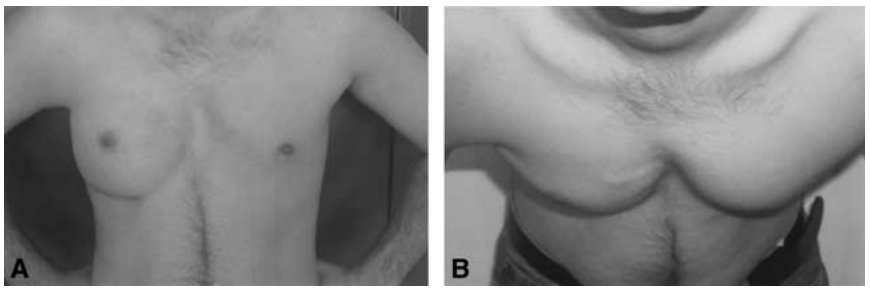

Fig. 4. (A) Anterior view of the patient, two months postoperatively. **(B)** View of the patient bending, two months postoperatively.

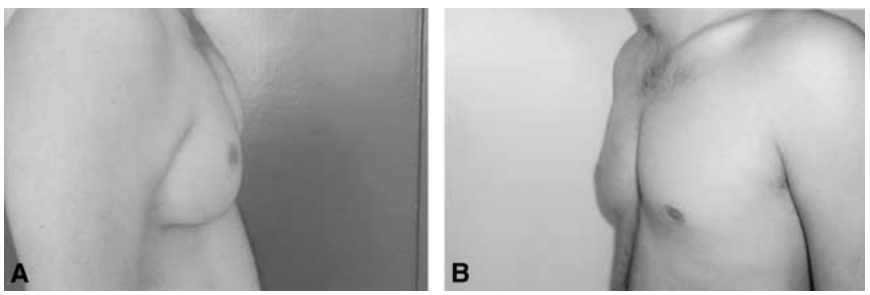

Fig. 5. (A) Lateral view of the patient from the right side, two months postoperatively. **(B)** Lateral view of the patient from the left side, two months postoperatively.

condition of congenital absence of PMM, there is no significant functional loss in ordinary activities because of compensation by surrounding muscles [11]. The anterior axillary fold is absent. PMM is composed of two fascicles, the sternocostal fascicle and the sternoclavicular fascicle. Generally, absence of PMM is incomplete and the sternocostal fascicle is often absent, but the clavicular fascicle is intact [5]. Other thoracic defects are the absence of the PMM, partial or complete absence of the serratus anterior and lattissimus dorsi muscles, and hypoplasia or aplasia of the ribs. The third, fourth, and fifth ribs are often affected in this syndrome [5,20].

In this case, our patient is male and his right side is affected. The clinical findings indicate the minor manifestation of Poland's Syndrome. Unlike cases in the literature, his second rib is hypoplastic.

In this syndrome, breast may be rudimentary and situated in a more cranial and lateral position. Amastia or athelia may be seen in this syndrome [5,13,20]. Noticeably sparse hair on the affected side and the lack of sweat glands is often associated [5].

Reconstruction of patients with Poland's Syndrome requires individualization by surgeon to address the particular degree of severity. In the literature, the latissimus dorsi musculocutaneous flap has been used to treat the congenital, unilateral absence of the PMM by stimulating the PMM and forming the anterior border of the axilla [14]. Disadvantages of the latissimus dorsi flap are the large scar on the back and the likehood of persistent fluid collection at the donor site. Atrophy of the transposed latissimus dorsi muscle has been also observed as a late occurrence [12].

Microvascular free flaps have become an increasingly popular option in reconstructing chest walls affected by Poland's Syndrome. It is possible that free tissue reconstructions with viable but denervated muscle experience late atrophy [10]. Many disadvantages of the tissue transfer exist, such as the following:

- Donor site scarring
- At least two operative stages and long surgical times
- Atrophy of the transposed muscle
- Long-term results that need to be reviewed

Silicone implants provide a means of correcting a strictly cosmetic deformity with much less morbidity. Minimal external scarring occurs if implant is placed through a submammary incision. Subcutaneous dissection must extend over the anticipated limits of the PMM using the contralateral muscle as a guide. The pocket should not be larger than necessary and should not be localized laterally. The risks of implant-related complications are infection, implant extrusion, and capsule formation [10]. Vinnik described a technique designed to preserve the dissected volume of the implant pocket after surgery by forcibly moving the prothesis within the cavity in a series of "expansion exercises" [19]. Beginning as early the first postoperative day, the implant is pushed from the lateral to the medial portions of the space every few hours. This movement, which is repeated frequently, avoids capsule development. Due to the absence of PMM, silicone prostheses cannot be placed in submuscular plane. The expansion exercises must be taught to the patient to prevent capsular contraction. The patients seek aesthetic treatment of mild Poland's Syndrome are generally young and they perform their expansion exercises.

A common problem with customized, nontextured prostheses is malposition [9,17]. For formation of anterior axillary fold and prevention of implant shifting to the lateral position, muscle remnants of PMM, which are sternoclavicular fascicles, can be transposed laterally based on its insertion. Strenoclavicular remnants of PMM have not yet been laterally transposed to form anterior axillary folds and to prevent the implant from shifting to the lateral position to treat Poland's Syndrome. We cannot find any information about this condition in the literature.

This patient with minimal chest deformity obtained satisfactory symmetry. Muscle transposition may not have been the procedure of choice in this patient. Customized implants have an advantage over autogenous materials in that very accurate contour restoration can be achieved with less morbidity and without donor defects. This is also a simple and timesaving procedure.

Conclusion

The cosmetic defects of the absence of PMM and mammary hypoplasia can be corrected with a customized silicone implant. This surgical approach should be given consideration as an effective and reasonable alternative to other accepted techniques for reconstruction of Poland's Syndrome chest deformities. This procedure may be used as a simple and safe alternative method for mild Poland's Syndrome.

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