

A Case of Ductal Carcinoma *In situ* of Breast with Poland Syndrome

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Abstract

Introduction: A 51-year-old woman was diagnosed with a rare case of multi-focal ductal carcinoma *in situ* (DCIS) of the breast associated with Poland syndrome. **Clinical Picture:** Physical examination showed mild hypoplasia of the left breast with microcalcifications on mammogram. Intraoperatively, there was complete absence of the pectoralis major, which precluded the insertion of a breast implant. **Treatment and Outcome:** She underwent a skin-sparing mastectomy and autologous microsurgical free flap reconstruction with deep inferior epigastric perforator (DIEP) flap. Postoperative recovery was uneventful with no evidence of recurrence at 6 months. **Conclusion:** This is the first reported case of DCIS of the breast with Poland syndrome. We performed a skin-sparing mastectomy with immediate DIEP flap reconstruction. Patient recovered well postoperatively with no evidence of recurrence at 6 months.

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Key words : Breast reconstruction, Ductal carcinoma *in situ*, Poland syndrome

Introduction

We report a rare case of multi-focal ductal carcinoma *in situ* (DCIS) of the breast associated with Poland syndrome. A skin-sparing mastectomy of the left breast with immediate reconstruction using ipsilateral deep inferior epigastric perforator (DIEP) flap was performed. The patient was a 51-year-old woman whose mammogram showed microcalcifications. Subsequently, a core biopsy revealed DCIS of the left breast. Physical examination showed mild hypoplasia of the left breast. Intraoperatively, a complete absence of the pectoralis major muscle with a less developed latissimus dorsi and pectoralis minor was found on the left side. Pathologic examination of the resected specimen showed multi-focal DCIS.

Case Report

A 51-year-old Chinese woman presented with suspicious microcalcifications in her left breast after a mammogram. Multi-focal calcifications were seen in the lower outer and inner quadrants. Ultrasound examination did not demonstrate any focal mass lesion. The left pectoralis major muscle was not seen on the medial-lateral-oblique view of the mammogram (Fig. 1). There was no medical or family history of note. Her menarche was at age 13 and she had had 4 children. Menopause had occurred 2 years ago. Physical examination revealed asymmetry of her breasts

with left-sided hypomastia (Fig. 2) and absence of the anterior axillary fold. Mammographic-guided core biopsies were performed which revealed multi-focal DCIS of the left breast.

A skin-sparing mastectomy was planned with reconstruction of her left breast. Reconstruction with ipsilateral DIEP flap was performed, with anastomosis of the left deep inferior epigastric artery and vein to the left internal mammary artery and vein. At surgery, the pectoralis major muscle was absent on the left side (Fig. 3) and the resulting chest wall defect was noted. The flap was larger than the resected specimen in order to achieve a better symmetry than preoperatively. The pectoralis minor and latissimus dorsi muscles were present. The contralateral right inferior epigastric artery and vein were found to be vestigial compared to the left side. The pathologic specimen revealed multi-centric and multi-focal DCIS in the lower inner and outer quadrants, measuring 7.4 cm in the largest dimension. The surgical margins were clear with no foci of invasive carcinoma. The patient made an uneventful postoperative recovery (Fig. 4). There was no evidence of recurrence on review at 6 months.

Discussion

Poland syndrome was first reported in 1841.¹ It describes a patient without the pectoralis major and minor muscles,

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Fig. 1.

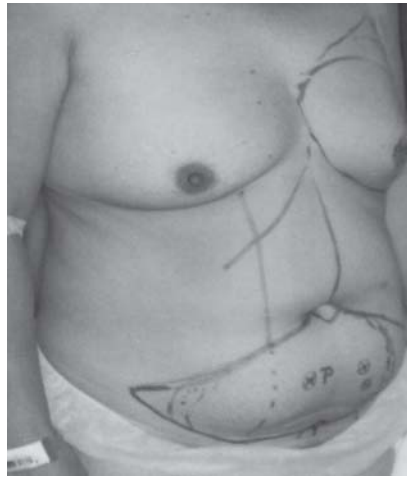


Fig. 2.

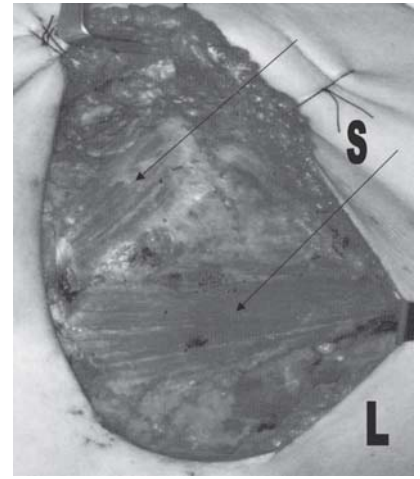


Fig. 3.

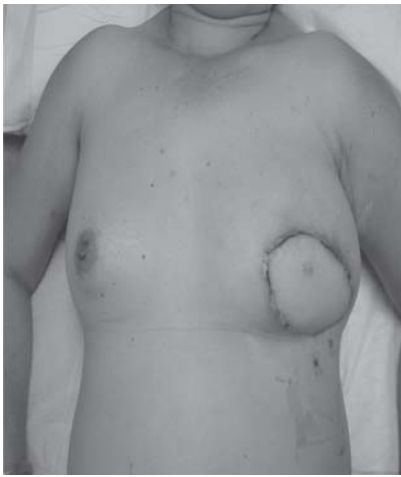


Fig. 4.

as well as with malformations of the ipsilateral upper limb. Female patients with Poland syndrome usually present with asymmetry, hypoplasia and a smaller nipple-areola complex on the affected side. Further evaluation will ascertain the degree of lack of muscular development of the pectoralis major and associated chest wall deficiency. Other commonly described findings include deficiency or absence of the subcutaneous tissue, breast or nipple, abnormalities of the anterior ribs, subclavian vein and its tributaries, absence or maldevelopment of other major muscles of the shoulder girdle, and anomalies of the ipsilateral upper extremity.²⁻⁴ The estimated incidence of this anomaly varies from 1 in 20,000 to 1 in 32,000 births.⁵⁻⁷

Although several cases of Poland syndrome associated with malignancy⁸⁻¹⁰ and a few cases of breast cancer related to Poland syndrome have been reported,¹¹⁻¹³ this is the first case of multi-focal DCIS. As Poland syndrome is more common in men than women,² there have been few reports of breast cancer in Poland syndrome in women.

Fig. 1. Medial-lateral-oblique view of mammogram demonstrating absence of pectoralis major muscle and suspicious microcalcifications in the left breast.

Fig. 2. Oblique preoperative view with markings of perforator locations (P) as well as hypoplastic left breast with less prominent inframammary crease. Right breast more developed with greater ptosis.

Fig. 3. Intraoperative view with absent pectoralis major and deficient pectoralis minor with remnant slips (arrows). S: superior aspect of mastectomy site; L: lateral aspect of mastectomy site.

Fig. 4. Two-week postoperative view showing reconstructed left breast with absence of left anterior axillary fold due to absence of pectoralis major muscle.

Consequently, reports of reconstruction after mastectomy are limited.

This patient was found to have an absence of the left pectoralis major muscle, hypoplasia of the left breast and vestigial contralateral right inferior epigastric vessels. In patients with Poland syndrome, the anterior chest wall asymmetry requires evaluation of the degree of muscular underdevelopment of the pectoralis major and associated chest wall asymmetry. Reconstruction in such patients presents a unique challenge; in some cases, the chest wall deficiency may be severe enough to require customised and prefabricated chest wall implants. A muscle flap, such as the latissimus dorsi, is often required to provide sufficient cover for the breast implant. This patient, however, had declined implant reconstruction.

In autologous reconstruction, the flap is designed to be larger than the size and volume of the resected breast. This is due to the deformity caused by the absence of the pectoralis major muscle and its corresponding loss of soft-tissue contour in the upper chest wall. Besides chest abnormality, a natural-looking breast is a reconstructive challenge when trying to match the contralateral normal breast. Failure to recognise the presence and significance of such a deformity can lead to suboptimal results in breast reconstruction.

The lack of pectoral sweep of the pectoralis major is rarely of concern to the patient and often goes unnoticed, although this problem has been addressed by authors who used latissimus dorsi muscle transfer.⁴ Transfer of the latissimus dorsi, preferably using endoscopic techniques, can certainly re-establish lateral sweep, although the muscle may be deficient and is prone to atrophy. The volume of the transferred latissimus was inadequate in this case to provide chest wall projection.

The chest wall projection is generally dealt with by using a customised or prefabricated chest wall implant, as suggested by Hodgkinson.¹⁴ However, a microvascular free flap reconstruction was chosen in this case due to the absence of the pectoralis major muscle and skin defect from the mastectomy. The use of autologous tissue from the ventral abdominal wall to reconstruct this defect also avoids the complications associated with an implant, which are reported to be between 15% and 100% in Poland syndrome. Seyfer et al³ removed 80% of the prostheses placed for chest wall/breast reconstruction because of implant migration or unsatisfactory cosmesis. The use of a DIEP flap in our case avoids the short- and long-term sequelae of implants while providing ample fat and versatility with less donor site morbidity, since no muscle was transposed compared to the pedicled or free transverse rectus abdominis myocutaneous flap. The ability to customise autologous tissue to reconstruct the contour deformities associated with accompanying pectus and rib deformities also provides an added advantage. De-epithelialised skin and fat helped to fill the infraclavicular hollow in our case.

Conclusion

This is the first reported case of DCIS of the breast in a patient with Poland syndrome. Implant reconstruction was not possible due to patient preference and an unacceptable rate of complication with subcutaneous placement. We did a skin-sparing mastectomy with immediate reconstruction with ipsilateral DIEP flap. Patient recovered well postoperatively with no evidence of recurrence at 6 months.

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