Breast Surgery

Case Report

Poland Syndrome and Breast Tumor: A Case Report and Review of the Literature

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Abstract

Poland syndrome is a rare congenital malformation. Hypoplasia of the sternocostal portion of the pectoralis major muscle is the most significant feature and is most frequently associated with homolateral breast hypoplasia. In this article, the authors present a case of bilateral phyllodes tumors in a 28-year-old woman with Poland syndrome and discuss (1) the relationship between the condition and breast cancer, (2) the modes of surveillance in patients with Poland syndrome, and (3) its impact on breast reconstruction.

Level of Evidence: 5

Keywords
asymmetry, phyllodes tumors, Poland syndrome

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Poland syndrome is a rare congenital malformation. The condition was mentioned for the first time by Lallemand in 1826, but it was Alfred Poland, a medical student and anatomist, who gave a precise description of the condition in 1841. Poland syndrome is associated with various degrees of thoracic and homolateral superior extremity anomalies, and the diversity of the clinical expression causes an underestimation of the frequency of Poland syndrome. The constant sign is agenesia of the sternocostal part of the pectoralis major muscle (PMM). Breast hypoplasia and thoracic wall deformities can also be associated with the condition. Breast asymmetry in women is frequently the sign, revealing the anomaly in puberty. The nipple-areolar complex can also vary in position, color, and dimensions in patients with Poland syndrome. Most cases are sporadic, and the etiology remains uncertain.

Breast cancer is the most frequent cancer in women. Diagnosis is most prevalent in patients between 55 and 60 years of age, but breast cancer is also found in younger women. The most common histological type is ductal carcinoma. Sarcomas account for less than 0.1% of malignant breast tumors. The clinical characteristics of the phyllodes tumor are close to those of fibroadenoma, with a more rapid progression. Phyllodes tumors can be classified into three histological types: Type 1 (benign tumor), Type 2 (borderline tumor), and Type 3 (corresponding to phylloide sarcoma). The basic problem in treating phyllodes tumors is their tendency to recur, which necessitates wide excision. Metastases are atypical, except in highly aggressive tumors.

Few reports in the literature discuss occurrences of breast cancer in patients who have Poland syndrome. In this case report, we describe bilateral phyllodes tumors in a young woman with Poland syndrome.

Case Report

A 28-year-old woman presented with breast asymmetry; her right breast was smaller than the left. A partial agenesia of the right PMM was easily established upon clinical examination. The right breast was smaller and less developed than the left. The nipple-areolar complex was also involved in the right breast, with a decreased diameter and color variation. Radiological examination of the chest revealed no other abnormalities or associated thoracic anomalies. The patient was referred to the Plastic, Reconstructive, and Aesthetic Surgery Department for further evaluation.

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examination (Figure 1). Multiple nodes were found in the right breast parenchyma. The patient had no thoracic anomaly and no obvious clinical anomaly of the right latissimus dorsi muscle. Her right upper extremity was hypertrophic, with brachymesophalangism and multiple syndactylies (Figure 2). The patient’s left breast was hypertrophic, with similar nodular lesions to the right breast. There were no pathological axillary lymph nodes bilaterally. Ultrasound and mammography of both breasts revealed five images typical of fibroadenoma in the left breast and three in the right breast (Figure 3).

In the first treatment procedure, the patient underwent a reduction mammaplasty of the left hypertrophic breast and fat grafting to the right breast. Fat tissue was harvested from the abdominal region, as described by Coleman.16-18 A 3-mm aspiration cannula, 15 cm long, with a rounded tip and double lateral openings was used to harvest the fat tissue. The cannula was attached to a 10-mL Luer-Lock syringe. The vacuum in the syringe was created manually and progressively to avoid any excessive depression on the adipocytes. Centrifugation was carried out for three minutes at 3000 rpm, and the fat was separated into three layers: superior or top (oil, containing triglycerides derived from the destroyed adipocytes), inferior or bottom (blood debris), and intermediary or middle (purified fat tissue). The top (oily) and the bottom (blood debris) layers were systematically removed. The purified fat tissue was transferred to 1-mL syringes.

The fat was injected through blunt-tipped cannulas 2 mm in diameter, with a lateral opening. The graft was performed following the principles of structural fat grafting; all layers were grafted, from deep to superficial. The tunnels created by the cannula formed a three-dimensional grill in the tissue of the recipient site. When passing the cannula, a small quantity of fat was injected along the retraction trajectory. The fat was grafted in small aliquots with each passage. The grafts were placed only in the right infraclavicular region and the anterior axillary pillar; 240 mL of fat tissue was grafted in total (Figure 4).
Pathology of the reduction mammaplasty tissue from the left breast showed fibroadenomas with a phyllodes tumor (borderline, Type 2; Figure 5). After multidisciplinary consultation, we decided to excise the tumors in both breasts. Therefore, in a second procedure, wide excision of the bilateral tumors was performed, and the right breast was reconstructed with a round silicone implant. The patient was preoperatively marked with an echo-guided harpoon, and the right breast was approached via periareolar incision. The tumors were reached through subcutaneous dissection. When necessary, subglandular dissection was performed to provide comfortable accessibility. In fact, the procedure was similar to a subcutaneous mastectomy with preservation of the tumor-free parenchyma. In the left breast, the tumors were addressed through the mammaplasty incisions, by the deep surface. After excision, the breast was remodeled with an external pedicle glandular flap and an inferomedial pedicle glandular flap sutured together in the midline. Thus, the residual postsection parenchyma defects collapsed, were decreased, and were even sealed. Postoperative edema and blood clots probably contributed to additional filling of these cavities. As a result, a nice breast contour was produced without visible irregularities.

No lymph node dissection was performed. The pathologic examination of all other tumors revealed some phyllodes tumors in the resected parenchyma from the left breast, with a complete excision and a safety margin. Six months postoperatively, a second fat tissue transfer was performed to fill the patient’s right infraclavicular hollowness and the anterior axillary pillar. The areola was enlarged by spreading of the existing areola (Figure 6).

The results were evaluated as good by the patient, and she has been annually followed clinically and radiologically for four years at the time of this report.

DISCUSSION

The association between some congenital disorders and the development of cancer is well recognized.\(^\text{10,19}\) It seems that such associations are caused by mutations in the protooncogenes or in the suppressor genes of the tumor.\(^\text{20}\) Poland syndrome is frequently associated with Hodgkin disease, some types of leukemia or leiomyosarcoma, renal tumors, or lung tumors,\(^\text{20-23}\) but there are few reports on the association between Poland syndrome and breast cancer.

A literature search returned only seven reports on Poland syndrome and breast cancer (see Table 1). In all cases, adenocarcinomas were found. Fukushima et al\(^\text{9}\) reported two cases of Poland syndrome and a homolateral breast cancer, one of which they treated with mastectomy and one of which they treated conservatively. Havlick et al\(^\text{10}\) reported on a 33-year-old woman with a Poland syndrome and homolateral breast cancer; his patient also had upper limb malformations. The authors presented it as the first report published on the association of Poland syndrome and breast cancer. Katz et al\(^\text{11}\) reported on the same association in a 42-year-old woman. They insisted that hypoplasia of the breast did not decrease the development of breast cancer. The follow-up for their patient consisted of annual clinical examination and mammography. Ultrasound and magnetic resonance imaging (MRI) were
indicated only in cases of breast reconstruction. Okamoto et al.\textsuperscript{12} reported the case of a 59-year-old woman with a Poland syndrome evident in one breast and invasive ductal carcinoma in the contralateral breast. She was treated by quadrantectomy and lymph node dissection. This was the only case of breast cancer in the contralateral breast. These authors suggested that breast cancer was more frequent in Poland syndrome but concluded that an epidemiological study was necessary to prove their hypothesis. Wong et al.\textsuperscript{13} reported on a multifocal ductal carcinoma in situ in a 51-year-old woman with Poland anomaly. She was treated with subcutaneous mastectomy and reconstruction with a deep inferior epigastric perforator (DIEP) flap. Khandelwal et al.\textsuperscript{14} reported an invasive ductal carcinoma in a 71-year-old woman. They concluded that the hypoplasia of the breast evident in Poland syndrome did not render the discovery of the tumor easier. Tamioloakis et al.\textsuperscript{15} reported on a similar case in a 53-year-old woman and insisted on the importance of early screening for cancer in women with Poland syndrome. None of the seven published reports included a case of Poland syndrome with phyllodes tumors, so we believe our case report to be the first of its kind. The case presented herein is interesting because the breast tumor is not a glandular carcinoma but a phyllodes tumor.

The wide excision with clear margins (superior to 10 mm) we performed is the treatment recommended in cases of phyllodes tumors since they have a high recurrence rate and risk of transformation in phylloide sarcoma.\textsuperscript{24} For patients with Type 3 phyllodes tumors, a mastectomy is recommended.\textsuperscript{25} The role of adjuvant treatments is unproven and must be considered on a case-by-case basis.\textsuperscript{25} Mammography and breast ultrasound were reported as unreliable for differentiating phyllodes tumors from fibroadenomas. Frozen section studies also seem to be of limited value.\textsuperscript{24} This information was proved in our case,

\begin{figure}[h]
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\includegraphics[width=\textwidth]{figure5.jpg}
\caption{Histological sections of phyllodes tumors (coloration, Hematoxylin-Eosin-Saffron [HES]). (A, B) Magnification $\times2.5$; nodule limited, polylobed with fibroepithelial proliferation. (C, D) Magnification $\times5$; stromal hypercellularity, which characterizes phyllodes tumors versus fibroadenomas, without atypia or mitosis (low grade).}
\end{figure}
where preoperative mammographies and ultrasound could not differentiate the tumors from fibroadenomas. Thus, the most reliable diagnosis remains the histopathological examination. No axillary lymph node dissection was performed since routine lymph node dissection is not recommended. This is because metastasis from phyllodes tumors seems to be uncommon, especially in cases with “normal” lymph nodes at the clinical examination. It is impossible to establish any direct correlation between phyllodes tumors and Poland syndrome since there are no similar cases in the literature. The low incidence of Poland syndrome (one in 30,000 live births) and the sex ratio (boys to girls = three to one) also make statistical correlation between Poland syndrome and breast cancer difficult. Nevertheless, it seems that patients with Poland syndrome are not at decreased risk of cancer, despite the breast hypoplasia signs. In this sense, it seems that the screening recommendations should follow those of the general population. A mammography (along with ultrasound and MRI, if indicated) seems necessary before any decision is made for breast reconstruction.

Various techniques for breast reconstruction in patients with Poland syndrome were reported in the literature, including breast implant placement, prefabricated thoracic and mammary implant placement, fat grafting, latissimus dorsi muscle flap dissection, and combinations of these. Obviously, the incidence of breast cancer in patients with Poland syndrome should cause us to proceed cautiously with fat grafting in these patients. In this case, fat grafting was used to treat only the right infraclavicular region and the anterior axillary pillar.27 Since these regions are hypoplastic in patients with Poland syndrome (due to PMM hypoplasia) and they are deprived of breast parenchyma, fat grafting can be easily performed with satisfactory correction of these deformities. No fat was injected directly in the breast parenchyma. All effort was made to inject it in the deep muscular plane, and thus all grafted tissue remained outside the breast parenchyma. The fat injection in this case was completed only after the patient signed a detailed written consent form. Fat grafting in patients with Poland syndrome should be carried out only after a careful pre- and postoperative examination (at three months) consisting of mammography, echography, and MRI, if indicated. As stated earlier, fat grafting should be reserved for the restoration of infraclavicular defects and the anterior axillary pillar.27

Since the patient’s imaging studies showed benign tumors (fibroadenoma), we did not plan to resect them in the hypoplastic breast. Clinical and radiological observation seemed sufficient. That is why the initial strategy included only an effort to create symmetry of the breasts—specifically, reduction of the left breast and correction of the Poland irregularities in the right breast through fat grafting. The decision to perform bilateral tumor resection was made after the final histological results showed Grade 2 phyllodes tumors in the left breast. Thus, tumor resection and breast reconstruction of the right breast with a round silicone implant took place at a second stage.
CONCLUSIONS

Patients with Poland syndrome have the same risk of breast cancer or borderline tumors as the general population. Therefore, when a patient with Poland syndrome presents for treatment, a bilateral mammography and ultrasound should be performed before proceeding with breast reconstruction. The small number of reported cases of concurrent Poland syndrome and breast cancer does not allow us to draw conclusions about etiology or predisposing factors. The possible development of breast cancer in patients with Poland anomaly should influence the surgeon’s choice of the techniques for reconstruction, including fat grafting.

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REFERENCES