A Very Large Malignant Phyllodes Tumor with Skin Ulceration and Nipple Areola Complex Involvement—Still a Reality!!!

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Abstract Cystosarcoma phyllodes is an uncommon neoplasm of the breast, constituting 0.3–0.9 % of all breast tumors. The incidence of malignant phyllodes tumor is even lower. The tumor is similar to fibroadenoma in structure, but it is distinguished from it histologically by large leaf-like projections of stroma with increased stromal cellularity (Dyer et al. *Br J Surg* 53:450–455, 1966). Although surgical removal is the mainstay of treatment, the extent of surgery required (excision vs. mastectomy) and the need for additional local therapy, such as radiotherapy, are unclear (Chaney et al. *Cancer* 89(7):1502–1511, 2000). We report a case of a 52-year-old married woman who presented to our outpatient department with 45×35×20 cm ulcerative, foul-smelling, huge right breast mass weighing 12 kg, and involving nipple areola complex, which turned out to be a malignant phyllodes tumor. The malignant variant of phyllodes tumor is indeed a very rare mammary tumor.

Keywords Breast · Phyllodes tumor · Malignant · Excision

Introduction

Cystosarcoma phyllodes is a rare, predominantly benign tumor that occurs almost exclusively in the female breast. Its name is derived from the Greek words sarcoma, meaning fleshy tumor and phyllon, meaning leaf. Grossly, the tumor

displays characteristics of a large, malignant sarcoma, takes on a leaf-like appearance when sectioned, and displays epithelial, cyst-like spaces when viewed histologically (hence the name). Because most tumors are benign, the name may be misleading. The World Health Organization in 1981 adopted the term phyllodes tumor, as this does not imply any biologic behavior, and it has gained wide acceptance [3]. To prevent confusion, the WHO classification should be strictly adhered to, so as to standardize treatment results and compare results from different centers. Thus, phyllodes tumors may be malignant (25 %), benign (60 %), or indeterminate, the so-called borderline lesion (15 %). Phyllodes tumors comprise less than 1 % of all breast neoplasms and are unique in their occurrence exclusively in the female breast and appearance in no other site in the body [4] (Figs. 1, 2, 3 and 4).

Case Report

A 52-year-old married multiparous lady presented to our outpatient department with a history of lump in her right breast for a period of 1 year. The lump, though initially small, started to increase rapidly in size, in the past 2 months, to become congested and heavy. The lump had burst open over the previous 15 days with initial discharge of seroanguineous and then pus, with foul odor. She did not complain of fever, jaundice, abdominal lump, or any bony pains.

On examination, there was pallor. Pulse and blood pressure were normal. There was a foul-smelling ulcerated mass measuring 45×35×20 cm, almost replacing the right breast. The nipple areola complex was also involved and the central one-third of breast was ulcerated, which exuded blood and pus with bad odor. The skin over the remaining mass was stretched and revealed dilated veins in the subcutaneous

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Fig. 1 Phyllodes tumor, showing ulceration. And fungation, with involvement of nipple—Areola complex

plane. The skin was mobile around the periphery and at the base. The whole tumoral mass and whatever little of breast that was left was mobile over the underlying chest wall. Axillary lymph nodes were palpable, two in number, largest measuring 1×2 cm and mobile. The left breast and axilla were normal. The respiratory and abdominal examination revealed no abnormality.

Since the mass had already ulcerated, we proceeded with incisional biopsy, which was reported as malignant phyllodes tumor. All blood investigations (except for increased total leukocyte count), chest X-ray, and abdominal ultrasound were normal. A right mastectomy was done. We took 2 cm margin from the periphery of the clinically palpable tumor; we did not dissect the axilla formally, but removed the low-lying palpable lymph nodes. As the patient was elderly and cosmesis was not our primary aim, so flap coverage was not done. Primary closure of the defect was done and suction drain placed after mobilizing the skin flaps. Postoperatively, patient made a fairly uneventful recovery, and was discharged on the tenth postoperative day. At discharge, the patient was very happy

Fig. 3 FNAC showing pleomorphic spindle cells

and relieved, and thanked us for helping her get rid of the large, ulcerated foul-smelling mass. The final biopsy report revealed high-grade malignant phyllodes tumor. Axillary lymph node showed reactive hyperplasia. Apart from regular follow-up, we advised the patient adjuvant radiotherapy.

Discussion

Phyllodes tumor is the most commonly occurring nonepithelial neoplasm of the breast, although it represents only about 1 % of the tumors in the breast [4]. The tumor was first described by Muller in 1838 [5]. Since then, numerous small and large series have appeared in the literature. There is still considerable uncertainty about the biological behavior of the disease. It has a smooth, sharply demarcated texture and typically is freely movable. It is a relatively large tumor with an average size of 5 cm. However, lesions of more than 30 cm have been reported. Although the benign tumors do not

Fig. 2 Cross section showing multiloculated cyst with necrosis

Fig. 4 Biopsy showing pleomorphic. Spindle cells without ductal involvement

metastasize, they have a tendency to grow aggressively and can recur locally [2]. Similar to other sarcomas, the malignant tumors metastasize hematogenously. Unfortunately, the pathologic appearance of a phyllodes tumor does not always predict the neoplasm's clinical behavior. Recurrent malignant tumors seem to be more aggressive than the original tumor. The lungs are the most common metastatic site, followed by the skeleton, heart, and liver. Most patients with metastases die within 3 years of the initial treatment [6].

Depending on the cellularity, invasiveness of the tumor, cytological atypia, and mitotic count, phyllodes tumor is classified into benign and malignant. Malignant is further divided into low-grade and high-grade based on histology. Pseudoangiomatous stromal hyperplasia occurs in patient with phyllodes tumor, and in some instances it is a prominent feature of the lesion.

There have been few molecular studies of phyllodes tumor. Comparative genomic hybridization analysis of 18 whole, fresh-frozen phyllodes tumors revealed that loss of material on 3p and gain of 1q were the two most common chromosomal abnormalities [7]. Recent studies have suggested that Ki-67, a proliferation marker, and p53, a tumor suppressor gene, may be implicated in predicting the behavior of the breast carcinomas in general and of phyllodes tumor in particular [8–10].

Clinically, a firm, mobile, well-circumscribed, nontender breast mass is appreciated. Overlying skin may display a shiny appearance and be translucent enough to reveal underlying breast veins. Physical findings (i.e., the occurrence of mobile masses with distinct borders) are similar to those of fibroadenoma [11].

For benign lesions, excision with a 1 cm minimum negative margin is advocated. For malignant tumors, if adequate margins are achieved with breast conserving surgery, mastectomy is not required. Formal axillary dissection seems to be unnecessary, but removal of low axillary lymph nodes cannot be criticized, especially if patients have palpable lymphadenopathy [4].

Adjuvant radiotherapy may be considered for high-risk phyllodes tumors, including those greater than 5 cm, with stromal overgrowth, with more than 10 mitoses per high power field, or with positive margins [12]. Radiotherapy is used as an adjuvant for acquiring maximum local control. Radiotherapy after local wide excision is thought to decrease the local failure rates; however, it has been found to have no significant effect on survival [13]. There is no clear indication for adjuvant chemotherapy for patients with phyllodes tumors [14].

Local recurrences are usually seen within the first few years of surgery, at the site of original excision, and are inversely proportional to the width of the negative resection margin. Most patients with local recurrences are treated with total mastectomy.

Most metastatic phyllodes tumors spread hematogenously to lung, bone, abdominal viscera, and mediastinum. There are no reports of long-term survivors. The optimum palliative treatment of metastatic phyllodes tumor has not been found. Cyclophosphamide, doxorubicin, cisplatin, and etoposide have been used with limited success. Radiation in symptomatic metastasis may be helpful. The majority of tumors contain estrogen or progesterone receptors, although palliation with hormone manipulation has not been extensively explored [4].

Conclusion

Malignant phyllodes tumor is an extremely rare breast tumor that usually does not ulcerate the skin or involve the nipple areola complex. In our case, the lady presented with a very large malignant phyllodes tumor (45×35×20 cm) weighing 12 kg, with skin ulceration and involvement of nipple areola complex, which is rarest of rare presentation. There is no reported literature of such case. This case gives an insight into the natural history of progression of phyllodes tumor, which may involve the skin and nipple areola complex in late stages, if left untreated.

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