Phyllodes Tumor Presenting with a Rare Widespread Abdominal Metastases: A Case Report and Review of the Literature

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Abstract

Phyllodes tumour is a rare breast neoplasia. The malignant variant is even rarer and it is unusual for abdominal metastases to occur sparing the more contiguous chest cavity and lungs. The patient presented with abdominal pain and distension, protracted vomiting and rapid weight loss five weeks after simple mastectomy for a bosselated, fungating right breast lesion. Exploratory laparotomy was carried out for the unexplained acute abdomen and it revealed an inoperable, generalized abdominal seedlings. This report illustrates a rare case of metastases at an unusual site from malignant phyllodes tumor occurring shortly after simple mastectomy.

Keywords: Phyllodes tumor; Simple mastectomy; Abdominal seedlings; Acute abdomen; Exploratory laparotomy

Introduction

Phyllodes tumor arises from the connective tissue stroma of the breast. Commonly, breast neoplasia involve the glandular and epithelial parts while the connective tissue component is rarely affected, consisting of about 0.5-1% of all breast lesions [1]. Phyllodes tumor is characterized by the formation of leaf like folds of breast tissues which give it the bosselated surface and which appear cystic on histo-pathological sections. This morphology made this tumor to be named albeit erroneously, cystosarcoma but the leaf like folds arrangement made the name phyllodes from phullon appropriate [1] and because majority of them (85%) are benign [2], the name phyllodes tumor is accepted as the standard nomenclature.

It is a rapidly growing breast lump that generate substantial tissue pressure, which causes skin necrosis and fungation [3]. About 15% of phyllodes tumor has malignant potentials [4], it hardly metastasize and when it does, common sites are the lungs, pleura and bone and this chance increases with larger masses [5,6]. Similarly, spread to the abdomen and peritoneum is rarer [6]. Very huge lesions that involve the whole breasts are seen in our environment because of late presentations and these are treated by simple mastectomies, with little concern for the ineffective adjuvant chemo-radiotherapies [7].

This is a case of histo-pathologically confirmed malignant phyllodes tumor, which presented with advanced, rare abdominal metastases shortly after a simple mastectomy.

Case Presentation

A 38 year old nulliparous lady presented with a right breast lump of 4 years, which started to increase in size in the past 6 months. She frequently felt weak, had good appetite and no weight loss. The increase in size was associated with fungation and subsequent erosion of the nipple. An initial serous breast discharge later became malodorous and purulent. A week prior to presentation, she started bleeding from the breast ulcer repeatedly and this compelled the relations to bring her to our level II surgical facility. Physical examination showed an apparently healthy looking lady, pale with a bosselated, fungating, bleeding lesion involving the entire right breast which was six times the size of the contralateral breast (Figure 1). The nipple was completely eroded but the breast was mobile on the chest wall. There was a 2 cm by 2 cm tender, soft, mobile level I right axillary lymph node. A preliminary diagnosis of malignant phyllodes tumor was made. The haemoglobin was 7.6 g/dl and leucocyte count showed neutrophilia.

Figure 1: A clinical photograph showing a grotesque right breast, bosselated, complete nipple erosion and contact bleeding.

Urinalysis and serum electrolytes urea and creatinine were normal. Chest radiograph was clear. Three units of blood were cross-matched. Because of the whole breast involvement and malodorous fungating...
lesion which was not attached to the chest wall, a simple mastectomy was carried out under general anaesthesia without a pre biopsy (Figure 2).

However, multiple sampling was taken from the whole breast specimen and sent to the pathologists. Having removed the smelling weighy grotesque breast, patient recovered swiftly with a good outlook to life.

She was discharged after three weeks but was readmitted two weeks later because of protracted vomiting associated with abdominal distension and pain. There was no fever, no cough but she had lost weight. Examination revealed pallor, healed mastectomy wound, no masses, no axillary node. The abdomen was distended and firm and mildly tender especially in the right iliac fossa. The chest was clear clinically and on radiographs. Erect plain abdominal radiograph showed multiple fluid levels. Abdomino-pelvic ultrasonography showed moderate ascites and echo changes in the right iliac fossa. A computerized tomography scan was not affordable. Serum electrolyte showed hypernatraemia, elevated urea but normal creatinine. Her urine output was adequate. Meanwhile, the histopathology report of the multiple samples confirmed malignant phyllodes tumor.

After adequate rehydration and under general anesthesia and a right para-median approach, we noted a hemorrhagic ascites and hyperemic intestines. The intestines and the mesenteries were adherent to the posterior abdominal wall. Further exploration showed mesenteric edema with widespread bubble like seedlings along the mesenteric border of the intestines (Figure 3).

The lesion was judged inoperable and some seedlings were taken for histopathology. The intra-operative findings were discussed with the patient and relations and ten days later, patient was discharged home on request. The histopathology also showed malignant phyllodes tumor.

Discussion

Phyllodes tumor is a neoplasia that originates from the connective tissue stoma of the breast and almost exclusively in the female breast [1]. It is rare when grouped with all breast neoplasias constituting 1%, but it is the commonest occurring non-epithelial tumor of the breast [1]. The aetiology is unknown but risk factors like female gender, fourth and fifth decades of life, previous breast lesions have been reported [1]. It is a fairly large tumor which could be confused with giant fibroadenoma but massive sized phyllodes tumors like in our index case have been reported, also is bilateral occurrence [1,7,8]. Our case involved the whole breast which was six times the size of the contralateral breast. The majority of phyllodes tumors are benign but the possibility of malignant variant should always be considered by clinicians especially in our environment where patients present very late. However, we were unable to link the protracted vomiting with the primary diagnosis in our patient initially because the abdomen is a rare site for metastasis and the patient did not show any sign of metastatic disease on the first presentation rather she showed progressive post-operative recovery up till the time of the initial discharge. Sometimes, distinguishing benign from malignant type is not always possible clinically [8] but the large size, nipple erosion and bleeding correlate positively with malignancy. Also, histopathology shows an unusual similarity among intra-canalicular fibroadenoma, benign phyllodes tumor and cystosarcoma and this could cause confusion even for the experienced pathologist [9]. A large mass could also show a spectrum of the disease with some areas benign, some borderline and some malignant [2]. More elucidation of the diagnosis could be done with immunohistochemistry where available.

Available data on phyllodes tumor are few due to its rarity and thus statistically significant inferences are difficult to make. Benign phyllodes tumors do not metastasize but they could have local aggressive growth and could reoccur locally after excision [1]. Malignant variant spread through the blood stream to the lungs, skeleton and mediastinum [5,10]. Apart from the benign and malignant variants, there is also a borderline phyllodes tumor which may appear benign on histopathology but behaves like malignant variant [4] clinically. Generally, the histo-pathological pattern of phyllodes tumor does not correlate with the biological behavior and therefore it is of limited predictive value for metastasis [1,8]. Impliendy, a biopsy confirmed benign tumour could reoccur after excision and
grow aggressively as if it is malignant. Metastatic disease could develop weeks and even years after treatment but spread to distant organs is rare [10]. A study approximated that 10% of phyllodes tumor develop distant metastases and this doubles to 20% if the tumour is histopathologically malignant [11]. Our index case started vomiting a few weeks following mastectomy, unfortunately, we did not associate this symptom immediately with abdominal metastases because it is rarer than involvement of more contiguous structures.

Phyllodes tumor could be diagnosed through astute clinical assessment. In our case, a massive, bosselated, fungating breast raised our suspicion. Furthermore, a pre-biopsy was not absolutely necessary because of the risk of uncontrollable haemorrhage [7] and also because of the whole breast involvement which was fungating, malodorous and needed to be removed anyway. A similar grotesque appearance could arise from tuberculosis of the breast but it is unlikely to be associated with nipple erosion. Clinically, cough, breathlessness, bone pain may be associated with metastatic lesions but in our patient, it was vomiting from abdominal spread. This breast tumour fungated through the skin [3] and unlike adenocarcinoma that could also fungate through the skin, phyllodes tumor is usually free on the chest wall. Smaller lesions on the other hand, may cause clinical diagnostic difficulties because they have distinct borders just like fibro adenoma [9] but phyllodes tumor occur in adult females who are 15 years or more, older than the age for fibroadenoma [8].

Giant fibroadenoma, adenocarcinoma, fibrocystic breast disease, pseudo-angiomatous stromal hyperplasia (PASH) and inflammatory breast cancer are possible differentials and distinguishing each lesion is by biopsy. Biopsy specimens must be a good representation of the lesion to improve diagnostic accuracy and multiple sampling is therefore very important. The histo-pathological distinctions are based on stromal cellularity, infiltration of the tumor edges and number of mitotic figures per high power field. Incisional biopsy for very large tumor or excisional biopsy for tumor 5cm and below is the standard practice. Because of heterogeneity and the risk of sampling errors, large core needle biopsy to a small extent and fine needle aspiration cytology to a large extent are not very reliable [12]. The benign tumor shows an increased number of regularly shaped fibroblast while the malignant type shows cellular atypia and increased mitotic figures. There are no useful biochemical tumor markers to strengthen the histo-pathological diagnosis of phyllodes tumor. They are not known to express estrogen receptors either and so hormonal manipulation is not useful in the treatment. However the expression of certain biological markers could help to determine the pattern of tumor activities and to distinguish the different variants. Consequently, immunohistochemistry for mitotic figures, Desmin and Vimentin determine malignancy while cluster of differentiation 10 (CD10) could predict metastasis [7,8,13]. Tumor genomic studies have shown chromosomal mutation, substitutions and deletions in the cell biology of the malignant variants [2,7]. Finally, phyllodes tumor cannot be diagnosed reliably on mammography or ultrasonography as there are no classic features with the use of these techniques [14].

The operative technique of shellling out a well encapsulated fibroadenoma is well known but this method of removal even when possible in phyllodes tumor is not advisable because it is associated with poor clearance and thus high chances of recurrence. The standard treatment for small phyllodes tumor 5cm or less is complete excision with a 1-2 cm cuff of normal tissue followed by histo-pathological confirmation and then long term follow up [15]. For large tumors or when cystosarcoma is confirmed, wide excision with 2 cm to 5 cm cuff of normal tissue is the treatment of choice [15-17]. When tumor load is massive or the whole breast is involved and the necessity for breast conservation is over-ridden like in our patient, simple mastectomy is indicated. Usually, there is no need for more radical operation or axillary dissection [18]. Malignant seeding in the lymph nodes is rare as spread is hematogenous and many enlarged lymph nodes are due to reactive hyperplasia from infection like in our case, and this expectedly resolved with antibiotics. Phyllodes tumor is not known to respond favorably to adjuvant chemo-radiotherapies or hormones or their combinations [7]. The prognosis for metastatic disease is therefore very gloomy as the systemic disease has no cure [18]. The rapid deterioration of our patient and the discovery of inoperable abdominal metastasis reinforce the poor prognosis of metastatic phyllodes tumor.

Conclusion

Huge phyllodes tumor has malignant potential and a rare distant spread to the abdomen like other systemic spread has a poor prognosis.

References


