Recurrent Malignant Phyllodes Tumor: A Rare Entity

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Abstract

Phyllodes tumor of breast is a rare tumor of fibroepithelial origin forming only about 0.3-0.5% of all breast malignancies. Malignant phyllodes tumor is even more rarely encountered lesion of very unpredictable clinical outcome with high chances of recurrence and distant metastasis. Accurate preoperative pathological diagnosis is mandatory to allow correct planning of the surgical treatment which can be either wide excision or mastectomy with an intent to have a negative surgical margin of at least 1 cms. This rare and enigmatic tumor should be kept in mind while dealing with breast lumps. Here, we present a case of recurrent malignant phyllodes tumor in a young female alongwith review of literature of this rare entity.

Aim and Objective: To present a case of recurrent malignant phyllodes tumor in a young female alongwith its cytology images and to have a review of literature of this enigmatic entity.

Keywords: Malignant; Phyllodes; Young; Recurrent; Female

Introduction

Phylloids tumor is a rare fibroepithelial lesion constituting only about 0.3-0.5% of the breast malignancies [1]. Malignant phyllodes tumor is an entity of unpredictable behavior owing to its high chances of recurrence and distant metastasis [2]. Prompt diagnosis is mandatory for appropriate management.

Case Presentation

Our attention towards this topic was drawn by a case of recurrent malignant phyllodes tumor in a young female who presented to our institution. A 22 year female presented with swelling of size approximately 4×3 cm over a surgical scar mark on the right side of chest. The patient gave a history of right mastectomy for a breast lump around 3 years back. Her previous histopathological reports stated the lump to be a malignant phyllodes tumor. On examination, the lump was hard with restricted mobility. The FNAC of the swelling over the scar was performed. Smears revealed hypercellular stromal fragments along with large number of highly pleomorphic, singly scattered, round to oval to spindle cells with retained cytoplasm at places. Numerous atypical mitotic figures were also evident. Diagnosis of malignant phyllodes tumor was given which was confirmed on histopathology. The wide surgical excision of lump was done. The surgical margins were free of tumor infiltration on histopathological examination. No axillary lymphnodes were detected on palpation and imaging. The CT scan of chest and abdomen revealed no tumor metastasis. Patient has been administered two cycles of radiotherapy and is lost to follow up at present (Figure 1).

We would like to proceed with the discussion of this rare and aggressive malignancy, presenting the cytological images of the above said case.

Discussion

An extensive data search was made on the internet to gather all important information regarding this rare aggressive malignancy of variable and unpredictable behavior.

Phylloids tumor is a rare fibroepithelial tumor with the majority of them, been described as benign (35% to 64%), with the remainder divided between the borderline and malignant subtypes [3]. Generally, it presents as a rapidly growing and clinically benign breast lump (usually >3 cms) in females within the fourth or fifth decade of life. Only a few cases have been reported in young age group as was in our case. The average size of this tumor is around 4 cms, however approximately 20% attain a size larger than 10 cms. These tumors can
reach sizes up to 40 cm in diameter. Mammography and ultrasonography constitute the chief imaging modalities. Wurdinger et al. showed that round or lobulated shape, well-defined margins, heterogeneous internal structure, and nonenhancing internal septations are more common findings in phyllodes tumors than in fibroadenomas [4]. It has been seen that the chances of malignancy are greater in older patient however it should be kept in mind that even the young patient can have malignant phyllodes tumor. As opposed to its benign counterpart, it is characterized by an unusually aggressive course and has been reported to recur locally and have the capacity for dissemination [2]. Komenaka et al. found the sensitivity of core needle biopsy to be 99% and negative predictive value and positive predictive value 93% and 83%, respectively, for the diagnosis of phyllodes tumor [5]. Histopathological features suggestive of malignancy include marked degree of stromal growth and atypia, ≥10 mitotic figures per high power field and infiltrative tumor margins.

Risk of recurrence is always associated with phyllodes tumors, stated to be around 20-30% for the malignant one. In a retrospective study of 352 cases of PT from 1954 to 2005, positive margins, fibroproliferation rate, stromal pattern, stromal cellularity, frequency of mitosis, and necrosis were associated with an increase in local recurrence rates [6]. Distant metastasis, mostly through hematogenous route is found in approximately 25% of patients with malignant phyllodes tumor and the dissemination is chiefly determined by the biological behavior of the tumor. The commonest sites for distant metastasis are the lungs (66%), bones (28%), and brain (9%) and in rare instances, the liver and heart [7]. Metastasis to axillary lymph nodes occurred in only 2% [8]. Various other malignancies as angiosarcoma, chondrosarcoma, osteosarcoma, liposarcoma, leiomyosarcoma, and rhabdomyosarcoma, are rarely encountered in malignant phyllodes tumors.

Main treatment modality includes wide excision with the aim of obtaining negative surgical margins of >1 cm. Since an excision with the required margins is often impossible in giant phyllodes tumors, mastectomy should be reserved for larger tumors and should be considered in recurrent tumors, especially of the malignant histotype [8]. Depending on the size of the breast and the location of the phyllodes tumor, mastectomy may also be required for tumors that are between 5 and 10 cm in diameter [9]. A retrospective review of malignant phyllodes tumor in 478 patients was conducted to determine local control rates based on tumor size and type of surgery performed. Five-year local control rates were higher for patients treated with mastectomy compared with lumpectomy [6]. Belkacemi et al. reported that radiotherapy was associated with superior local control rates at 10 years, from 59% to 86% for both borderline and malignant phyllodes tumors [10]. Systemic chemotherapy and hormonal therapy is debatable in phyllodes tumor. To date there is no double blinded, multicenter study on this topic [11].

Regular follow up of the patient is necessary, 6 monthly during the first two years after treatment and then on yearly since this tumor is very unpredictable in recurrence and metastatic activity.

**Conclusion**

Phyllodes tumor should be kept as a differential diagnosis while dealing with the breast lump patients. Furthermore, malignant phyllodes is an aggressive breast neoplasm with high potential of recurrence and metastatic activity. So an appropriate diagnosis on time is mandatory for proper management.

**References**