Giant Phyllodes Tumor Resulting from Gross Negligence
(A Case Report and Review of Literature)

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Abstract

Phyllodes tumors are rare fibroepithelial lesions that make up 0.3 to 0.5% of female breast tumors. They are benign in 35-64% of cases and timely diagnosis and surgical intervention gives good results. But lack of knowledge and fear for surgery brings many patients with giant phyllodes tumor in Indian setting. Herein we report a case of a 47 year old lady with diagnosis of benign phyllodes tumor for 20 years and neglecting it, till it grossly enlarged and started bleeding actively. She came with hemoglobin of 3.3g%, but is now doing well after mastectomy. So awareness among the people and proper counselling is required to reduce such cases and their complications.

Keywords-Giant Phyllodes tumor, fibroepithelial lesion, cystosarcoma phyllodes, fungating right breast mass, monoclonal proliferation of stromal Cells

INTRODUCTION

Phyllodes tumors are rare fibroepithelial lesions that make up 0.3 to 0.5% of female breast tumors [1]. The incidence is about 2.1 per million women and common age group is 45 to 49 years [2,3]. Chelius in 1827 [4] first described this tumor and Johannes Muller (1838) was the first person to use the term cystosarcoma phyllodes. In 1981 the World Health Organization adopted the term phyllodes tumor and subclassified them...
histologically as benign (35-64%), borderline, or malignant [5]. When the size is > 10 cm, it is called giant phyllodes tumor.

CASE REPORT

A 47 yr female presented with huge fungating right breast mass and bleeding [Fig. 1]. She had noticed the lump 20 yrs back and was on some medication. There was a gradual increase in the size of the swelling, but because of fear of surgery she kept on neglecting it until there was rapid increase in size for last 5 months and bleeding for 2 weeks.

On presentation the pt. was pale (Hb 3.3g %), right breast was hugely enlarged with altered contour and active bleeding. Base was free and there was no axillary and supraclavicular lymphadenopathy.

The FNAC was phyllodes tumor. Because of acute emergency and compromised anaesthetic fitness, mastectomy was carried out with few basic investigations and simultaneous blood transfusion. The patient was still unwilling for surgery and had to be counselled many times before giving consent for surgery. Intra-operatively, pectoralis muscle was not infiltrated [Fig. 2] and primary closure could be done.

Tumor specimen measured 18x17x12 cm, weight 2.5 kg with all margins free. Microscopically it was benign phyllodes tumor [Fig. 3]. The post-op period was uneventful and the patient is doing well in follow-up since last 10 months.

The phyllodes tumor is caused by monoclonal proliferation of stromal cells. World Health Organization has divided phyllodes tumor into benign, borderline, and malignant categories based on the degree of stromal cellular atypia,

Fig. 1 Huge fungating right breast mass

Fig. 2 Intra-operative photograph

Fig 3. Histopathology (H & E stain 40X)

REVIEW OF LITERATURE

The phyllodes tumor is caused by monoclonal proliferation of stromal cells. World Health Organization has divided phyllodes tumor into benign, borderline, and malignant categories based on the degree of stromal cellular atypia,
mitotic activity per 10 high power fields, degree of stromal overgrowth and tumor margin [5].

It is diagnosed by clinical presentation, radiological investigations (which includes ultrasonography, mammography, MRI and color Doppler ultrasonography) and pathological assessment by FNAC and core needle biopsy. FNAC has an overall accuracy of about 63% [6] and core tissue biopsy has sensitivity of 99% and negative predictive value and positive predictive value 93% and 83% respectively [7]. Paddington Clinicopathological Suspicion Score outlines criteria to assist in the selection of patients for core biopsy, for use in conjunction with existing local protocols.

According to NCCN guidelines, surgery with > 1 cm margin is the treatment of choice. It can be accomplished by lumpectomy or mastectomy with or without immediate reconstruction. As malignant phyllodes tumors undergo mainly hematogenous spread, the proportion of patients with lymph node metastases are <1% (lymph node enlargement in about 10%) and routine axillary clearance is not recommended. Axillary dissection is required, when histologically positive for malignant cells. For recurrent disease, in non-metastatic setting re-excision with wide local excision and post-operative radiotherapy should be considered. In metastatic setting, management is according to the principles of soft tissue sarcoma. In MD Anderson Cancer Center, radiotherapy is recommended for positive or near-positive surgical margins and for whom further surgical procedures cannot be performed. Chemotherapy has no survival advantage and endocrine therapy has not been extensively studied.

Local recurrence (15 to 20%) is correlated with positive excision margins. Factors like stromal overgrowth, tumor necrosis, infiltrating margins, mixed mesenchymal components, high mitotic rate, and stromal atypia are important for recurrence but in isolation each appears to have a low predictive value. Higher p53, Ki-67 expression and microvessel density is associated with poor prognosis.

10% of patients develop distant metastases. The commonest sites for distant metastases are lungs (66%), bones (28%), brain (9%) and rarely, the liver and heart. So the patients should be followed up regularly at 6-month interval for the first two years and then on yearly basis.

CONCLUSION

Phyllodes tumor bears specific clinical characteristic and can be considered as a differential diagnosis for the breast lumps. Patient's lack of information and also fear of surgery seems to be the main reasons for patients presenting with giant phyllodes tumor. Nevertheless, if there is no malignant transformation, giant phyllodes tumor can be completely resected with excellent results.

REFERENCES


