ISSN: 0976-3031

Available Online at http://www.recentscientific.com

CODEN: IJRSFP (USA)

International Journal of Recent Scientific Research
Vol. 8, Issue, 9, pp. 19969-19972, September, 2017

International Journal of Recent Scientific Research

DOI: 10.24327/IJRSR

Research Article

PHYLLODES TUMOR WITH SARCOMATOUS DIFFERENTIATION- A RARE AND DISTINCT ENTITY

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DOI: http://dx.doi.org/10.24327/ijrsr.2017.0809.0797

ARTICLE INFO

Article History:

Received 15th June, 2017
Received in revised form 25th July, 2017
Accepted 23rd August, 2017
Published online 28th September, 2017

Key Words:

Sarcomatous, phyllodes, malignant.

ABSTRACT

Phyllodes tumors are fibroepithelial neoplasms of the breast accounting of about 0.3- 0.9% of all breast tumors. They can be classified as benign, borderline or malignant according to World Health Organization with benign tumors comprising 54-85% of all cases. A 20 year old female presented with a right breast lump since 3 months. On physical examination lump was in the lower inner quadrant of right breast. Ultrasonography revealed a heterogeneous, lobulated mass. On Fine- needle aspiration cytology, a diagnosis of atypical cytology, breast (C3) was given. The patient underwent a right sided simple mastectomy. The specimen was sent for histopathological examination and a diagnosis of malignant phyllodes tumor with sarcomatous transformation was rendered. As considering about the rarity of this tumor a possibility should always be kept in mind in tumors of breast having similar characteristics so as to help in the early management of the patient.

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INTRODUCTION

Phyllodes tumors are fibroepithelial neoplasms of the breast accounting of about 0.3- 0.9% of all breast tumors.[1,2] They can manifest as benign lesions similar to fibroadenomas, or as malignant lesions with a potential to metastasize to distant sites. Histopathologically, they can be classified as benign, borderline or malignant according to World Health Organization[3]. With benign tumors comprising 54-85% of all cases.[4] They arise from periductal stroma and consists of both epithelial and stromal components. An association of phyllodes tumors (PT) with heterologous sarcomatous differentiation is quite rare and only five cases have been reported in the literature uptil now.[5,6,7,8]

Prognosis of these tumors is poor and mastectomy is indicated only for large tumors and those with persistent positive margins but axillary lymph node dissection is usually not indicated as phyllodes tumors rarely involve the regional lymph nodes. The role of adjuvant chemotherapy is unproven.[9] and should only be considered in patients with large, high grade malignant phyllodes tumor.[10]

We present a case of malignant phyllodes tumor with sarcomatous transformation in a young female.

Case report

A 20 year old female presented with right breast lump since 3 months. On physical examination 2x2.5cm mobile lump was noted in the lower inner quadrant of right breast. Ultrasonography revealed a 2.5x2x1.3cm heterogeneous, mildly hypoechoic, lobulated mass in the right breast. Computed tomography of the chest, abdomen and pelvis showed no evidence of metastasis. On Fine- needle aspiration cytology, a diagnosis of atypical cytology, breast (C3) was given. Following this the patient underwent a right sided simple mastectomy and the specimen was sent for histopathological examination. Grossly the specimen measured 15x11x8cm (Fig no.1) and overlying skin measured 14x10cm. On external surface, overlying skin showed multiple ulcerations.

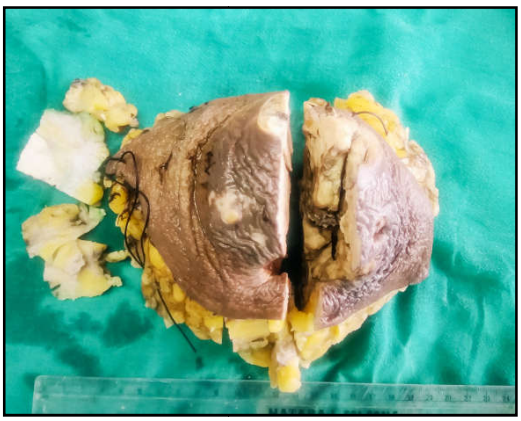

Fig 1 Gross photograph showing a mastectomy specimen measuring 15x11x8cm. Overlying skin shows multiple ulcerations

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On serial sectioning, a lobulated mass measuring 7x8x6.5cm was noted which was located 1cm from the deep resected margin. It was located 1cm from the superior margin, 2 cm from the inferior margin, 2.5cm and 3.5cm from the medial margins and lateral margins respectively. On cut section, mass was grey-white to grey-brown with areas of hemorrhage and necrosis (Fig no.2).

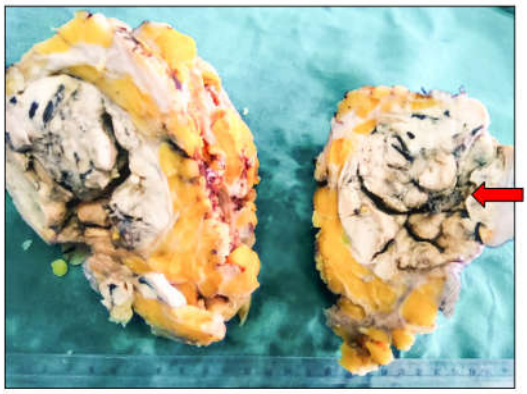

Fig 2 Cut surface showing a lobulated, well circumscribed, grayish-white mass. (arrow)

On microscopic examination a diagnosis of malignant phyllodes tumor with sarcomatous transformation was rendered characterized by a 'leaf like' growth pattern showing marked stromal proliferation (Fig no.3,4).

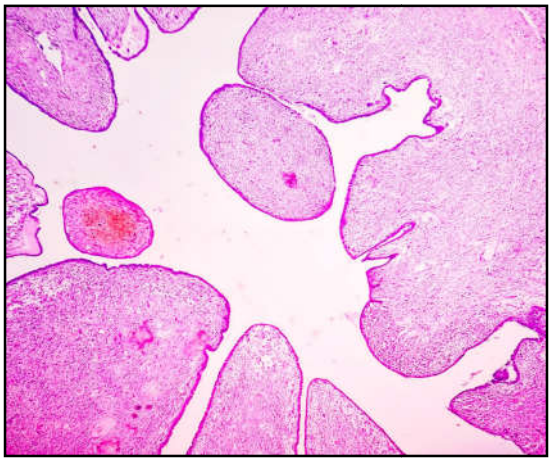

Fig 3 (H & E stain, 4x), Microphotograph showing the characteristic "leaf-like" growth pattern of a phyllodes tumor consisting of both epithelial and mesenchymal components

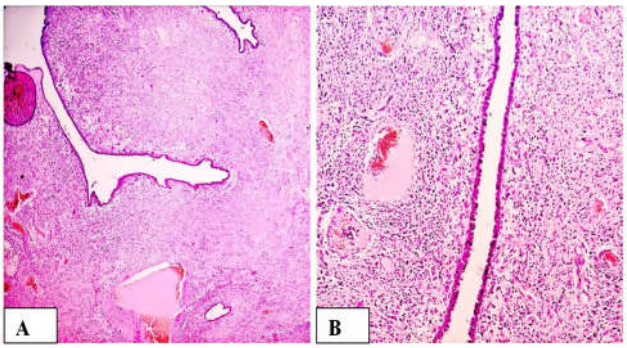

Fig 4(A, B)- (H & E stain, 4x), Stromal component showing hypercellularity

Within the phyllodes tumor, sarcomatous change was evident (Fig no.5).

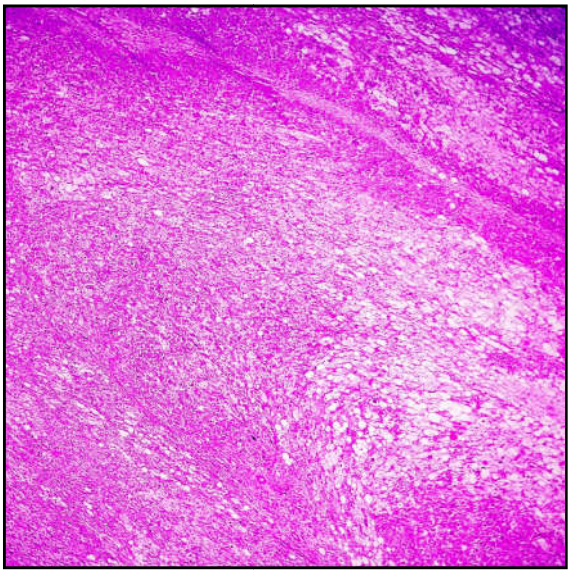

Fig 5 (H & E stain, 10x), Microphotograph showing broad areas of sarcomatous change with cells showing marked atypia

Cells showed marked pleomorphism, atypical nuclei with irregular cell borders and high mitotic acivity upto 15 mitotic figures in 10 high power fields (Fig no.6).

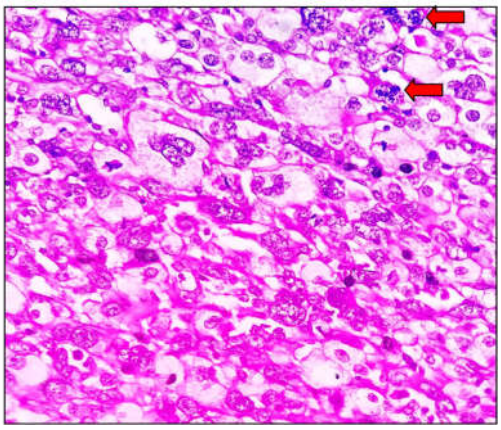

Fig 6 (H & E stain, 40x), Microphotograph showing cells with highly pleomorphic nuclei, nuclear atypia and numerous mitotic figures are seen as shown by the arrows

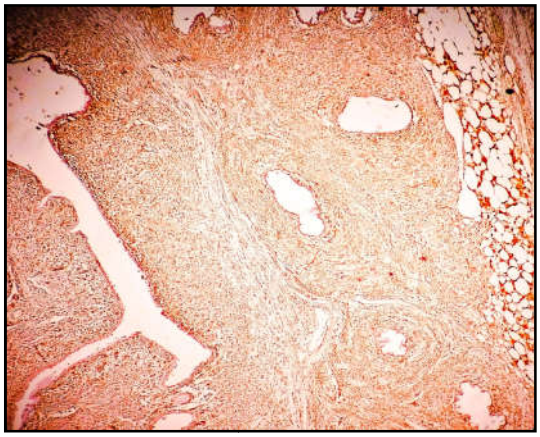

Fig 7 (Vimentin, 10x), Stromal cells showing positivity for vimentin demonstrating the mesenchymal origin of the tumor.

Sections from the skin, nipple- areola and deeper plane of resection along with all margins were free of tumor. Adjacent

breast showed features of fibrocystic change with moderate epithelial hyperplasia of usual type. On immunohistochemistry tumor cells showed positivity for vimentin (Fig no.7). Tumor cells were negative for Cytokeratin, EMA, S100 and CD-68.

DISCUSSION

The term Phyllodes was firstly proposed by Johannes Muller in 1838[11] as 'Cystosarcoma Phyllodes'. Depending upon histological features, PTs are classified as benign, borderline or malignant. Microscopically they are characterized by having a typical leaf- like pattern formed by the projections of hypercellular stroma into epithelium- lined cystic spaces. Components of both epithelial and stromal cells should be there for the diagnosis of PT.

Many histological grading systems have been proposed for PTs, with revisions of the classification first described by Treves and Sutherland in 1951.[12] Most of the systems used for grading are three- tiered : benign, borderline or malignant or low, intermediate or high grade and use the histological parameters including margins characteristics, cellular atypia, stromal overgrowth, stromal cellularity and mitotic activity.[13] A heterogeneity of malignancies can be associated with PTs, with its dual population of cells.[6] The ductal component can become malignant whereas the mesenchymal component can demonstrate sarcomatous differentiation as was seen in our case. It is quite rare to find heterologous sarcomatous transformation in PTs and till date only five cases have been reported in the literature, all of which were associated with malignant PT.

Many prognostic factors have been assessed for the identification of these tumors with aggressive features which include grade,[4] infiltrative borders, marked stromal overgrowth,[14] stromal atypia, high mitotic count,[14,16] pseudoangiomatous stromal hyperplasia,[17] heterologous stromal elements, fibroproliferation in the surrounding breast tissue and tumor necrosis.[16,18]

The prognosis of phyllodes tumors is not well defined with local recurrence rates ranging from 10-40% (average 15%) and distant metastases in 10% of all phyllodes tumors and in upto 20% of malignant phyllodes tumors.[13] Survival is poor after metastatic disease with median survival of 4 to 17 months documented in the literature. Reinfuss *et al* reported 5 year disease free survival rate of 96% in benign phyllodes tumor and 66% in malignant phyllodes tumors.[4]

CONCLUSION

As considering about the rarity of this tumor accompanied with sarcomatous component, there is a paucity of data available regarding the presentation, diagnosis and treatment therefore a possibility should always be kept in mind if a tumor of similar characteristics is noted in the breast and a careful approach should be commenced early so as to help in the proper management of the patient.

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How to cite this article:

Priyanka Anand and Namrata Sarin.2017, Phyllodes Tumor With Sarcomatous Differentiation- A Rare And Distinct Entity. *Int J Recent Sci Res.* 8(9), pp. 19969-19972. DOI: http://dx.doi.org/10.24327/ijrsr.2017.0809.0797

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