Case Report

An Anomalous Collision Tumour in the breast! Angiosarcoma and Benign Phyllodes Tumour

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ABSTRACT

Breast Angiosarcoma is a very rare aggressive mesenchymal tumour which may represent only 0.04% of all breast malignant tumors, with few patients surviving long term. In the year 1887, Schmidt described the first case of Breast Angiosarcoma. Breast Angiosarcoma may be primary or secondary. Phyllodes tumour comprises 1% of breast malignancies, and is not a rare neoplasm. They were first considered as a kind of breast sarcoma, and show variable clinicopathological behaviour ranging from benign to malignant depending on histology. All forms of phyllodes tumours are regarded as having malignant potential, and the malignant change usually occurs within the hyperplastic stromal cells, resulting in sarcomatous lesions. Therefore pathogenesis of phyllodes tumour still remains an enigma.

We report a case of a 45 year old female patient suffering from recurrent bouts of pain and swelling in the breast, who was operated for a large palpable mass diagnosed as benign phyllodes tumour on histopathology three years back, which eventually recurred, and the final histopathology report was given as benign phyllodes along with angiosarcoma.

Key words: Angiosarcoma, Breast, Collision tumour, Phyllodes, Rare disease.

INTRODUCTION

Collision tumors represent a coexistence of two adjacent but histopathologically distinct tumors, without admixture in the same tissue or organ. They are rare in different organs, and rarer in breast. The collision tumors are diagnosed postoperatively because there are no specific features that aid their diagnosis preoperatively. Careful gross examination and extensive histopathological study from various parts of tumor is essential for proper diagnosis and further management. Breast angiosarcoma is a very rare aggressive mesenchymal tumour which may represent only 0.04% of all breast malignant tumors described first by Schmidt. [¹,²] A synchronous benign phyllodes and angiosarcoma in the breast is a rare entity, and only two cases to the best of our knowledge have been documented in literature so far. [³,⁴] Invasive ductal carcinoma and MALToma, primary cutaneous angiosarcoma with breast trauma, breast carcinoma with malignant phyllodes, and phyllodes tumour of the breast with malignant melanoma are the few examples of collision tumour in the breast. Thus, we report a phenomenal case of collision tumour consisting of benign phyllodes and angiosarcoma, presenting as a breast lump.

CASE REPORT

We came across a phenomenal case of a 45 year old female patient, with no relevant medical history, no family history of breast carcinoma, and without any history of thoracic radiotherapy in the past, who
presented with a palpable right breast lump in the year 2012, which on USG was suggestive of a cystic mass with irregular margins at 2 and 5'o clock positions. After three months, the right breast mass increased in size and histopathological examination of the tumour suggested a borderline Phyllodes tumour with fibrocystic change, following which a conservative surgery was performed, and the tumour was excised. After two years, the tumour recurred. An FNAC was done which suggested chronic mastitis of the right breast. After seven months, patient arrived with similar complaints. This time another FNAC was done, which suggested a benign proliferative breast disease-suspicious of malignancy. After one year, the patient came to the surgical Out Patient Department with complaints of diffuse enlargement of the breast, which was rapidly growing, and increased in size over a period of twenty days, with pain, and a small ulcerated area on the skin. Finally, an MRM (Modified Radical Mastectomy) was performed. We received the specimen, which on gross examination showed a skin covered fibrofatty piece of tissue measuring 22x11x8 cms, the size of the skin flap measuring 21x12x0.4 cms. A large fungating mass was seen protruding out from the lower quadrant measuring 13x8x4 cms. Also seen was an ulcerated area which measured 1.5 cms in diameter on the skin. The nipple appeared to be retracted, but no discharge was seen. On sectioning, a large tumour measuring 11x6x18cms having solid whitish areas, with few soft, yellowish, necrotic areas were seen in the retroareolar region, involving all four quadrants of the breast. The fungating mass appeared to be separate from the tumour proper, the cut section of which showed ill defined blackish necrotic friable areas. The deep resection margin seemed to be involved by the tumour proper. Axillary tail was present measuring 6x3x0.5cms. However, no lymph nodes were detected.

Histopathological examination from the deep resection margin (DRM) revealed an intracanalicular growth pattern of the glands with a leaf like architecture, on a background of highly cellular stroma. Therefore, a diagnosis of Benign Phyllodes was made. Sections from the fungating growth revealed closely packed, increased proliferation of thin walled anastomosing vascular channels lined by atypical endothelial cells, invading the breast parenchyma. The mitotic count was 8-10/hpf, showing atypical mitosis and marked nuclear pleomorphism, with some areas having slit like remnants of vascular channels, and a highly cellular stroma consisting of spindle cells, tumour giant cells and areas of necrosis. A diagnosis of recurrent Phyllodes with Angiosarcoma was made. Immunohistochemical studies were conducted to rule out other synchronous tumour such as malignant fibrous histiocytoma, leiomyosarcoma, extraskeletal osteogenic sarcoma, or carcinosarcoma. Neoplastic cells had a positive reaction for CD31, CD34 and Vimentin, while it was ER and PR negative. An IHC with CD31 was done because of its high sensitivity and specificity to endothelial differentiation to confirm Angiosarcoma, which showed that the endothelial cells lining the vessel walls were immunoreactive for CD31. Thus, the diagnosis of Angiosarcoma was confirmed.

Figure 1: Gross examination of the right breast showing a fungating growth protruding out from the lower quadrant of the breast, and an ulcer (right side) measuring 1.5 cms in diameter.
Sanchari Saha et al. An Anomalous Collision Tumour in the breast! Angiosarcoma and Benign Phyllodes Tumour

Figure 2: Cut section of the breast shows a well demarcated mass composed of two separate tumorous lesions.

Figure 3: Photomicrograph showing the anastomosing vascular channels lined by atypical endothelial cells.

Figure 4: Photomicrograph showing endothelial cells lining the vessel walls are strongly immunoreactive for CD31.

Figure 5: Photomicrograph showing angiosarcoma of the breast (10 xs)

Figure 6: Photomicrograph showing Benign Phyllodes tumour of the breast, (10x)

Figure 7: Photomicrograph showing the collision of Angiosarcoma and Benign Phyllodes Tumour in the same field.
DISCUSSION

This case highlights the huge variation in behaviour and histopathology of these two tumours in the same breast. On review of literature, we found three cases of true collision tumour of the breast, where the tumours were composed of two different components in a separate manner. Our present case showed distinct presentation of the two separate tumours in the same site without any interminglement grossly as well as microscopically. Phyllodes Tumour accounts for approximately 0.3-0.5% of all breast tumours. They have a propensity to be fast growing. They are fibroepithelial elements projecting into a hypercellular stroma in a leaf like fashion. Their behaviour from benign to malignant depends on the histological features like, stromal cellularity, cytologic atypia, infiltration at the tumours edge and mitotic activity. Recent studies have suggested that the epithelium exerts a field effect that potentiates stromal overgrowth. Malignant change occurs usually within the hyperplastic stromal cells resulting in sarcomatous lesions. Recurrence is very common in these tumours, especially in involved resected margins and wide excision remains the mainstay of treatment. The clinical prognosis of collision tumour may be influenced by the subtype and pathologic stage of the more aggressive tumour in the breast. The incidence of Breast Angiosarcoma is 17 new cases per million women. [5] Borrman in 1907 first documented a case of angiosarcoma. [6] Studies reveal that given the endothelial origin of Breast Angiosarcomas, angiogenesis inhibition represents potentially attractive therapeutic modality. They are rare malignant tumours arising from the endothelial cells lining vascular channels, having high local recurrence and poor prognosis, but lymph node metastasis is rare as reported by Sher et al. It tends to occur in other sites such as, Liver, skin and deep soft tissues, and spreads hematogenously to lungs. [7] Breast Angiosarcomas may be primary or secondary. In comparison, primary angiosarcomas are relatively rarer, they typically occur in younger women, arise from breast parenchyma; and Secondary angiosarcomas usually occurs in elderly women, arise from skin, and show a pattern of infiltration into breast parenchyma. Patients with BAs may present with a palpable, rapidly growing breast lump that is insidious in onset. In a study of 24 breast angiosarcoma cases, the mean tumour size of the mass at presentation was 5.9 cms. [8] Imaging studies do not contain characteristic features of an angiosarcoma. Harden et al previously reported a case of a Borderline Phyllodes tumour subsequently developed cutaneous Angiosarcoma of scalp, which later metastasized widely. However, the link between the two tumours could not be established by him. [9] In our case, the exact reason behind the appearance of angiosarcoma along with recurrent phyllodes, could not be established as well. Angiosarcoma stratifies three grades, depending upon their histopathological features. The prognosis for patients with BA depends on tumour size, presence of a residual disease and cellular pleomorphism. According to Nascimento et al, grade of the tumour may not be an important prognostic factor, and for a definitivediagnosis, tissue biopsy is needed with immunostaining, for the presence of blood vessel, with endothelial markers CD31 and CD34. MRM is the preferred method for surgical treatment, whereas immunohistochemistry (IHC) studies are required for the confirmatory diagnosis of this disease.

CONCLUSION

We reported a case of a true collision tumour presented as BenignPhyllodes and Angiosarcoma. A large metabolically active vascular mass in breast should always be considered at first sight to be an angiosarcoma until proven otherwise. A definitive diagnosis is based on histopathology with immunostaining with the endothelial markers CD31 and CD34. Total mastectomy is the preferred
surgical treatment, and further investigation with adjuvant therapy is required for the aggressive nature of this disease, to prevent its recurrence after surgery, though literature contains few data concerning adjuvant treatment, and there is no generally agreed course of action.

REFERENCES


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