A rare case of recurrent malignant phyllodes tumor of the breast in a young nulliparous woman

Zeeshanuddin Ahmad, Mahim Koshariya, Sameer Shukla, Vikram Vatti, Abhijeet Diwan
Department of Surgery, GMC, Bhopal, Madhya Pradesh, India

ABSTRACT
Phyllodes tumors (PT) are fibroepithelial neoplasm characterized by a combination of hypercellular stroma and cleft-like or cystic spaces lined by epithelium, into which the stroma classically project in a leaf-like fashion and have a potential to recur and metastasize. These tumors are more common in 3rd to 4th decade. Here we present a case of recurrent malignant PT of the breast in a young nulliparous woman.

Key words: Nulliparous woman, phyllodes tumor, recurrence

INTRODUCTION
Phyllodes tumors (PT) are a rare entity in the breast. PT are fibroepithelial lesions that account from 0.3% to 0.5% of female breast tumors[1] and have a potential for recurrence. Chelius in 1827 first described this tumor whereas Muller in 1838[2] coined the term cystosarcoma phyllodes; a misleading description as the tumors are rarely cystic and majority follow a benign clinical course. PT is the currently accepted nomenclature according to the World Health Organization. Surgery has been the primary mode of treatment for PT, with mastectomy being the common treatment of choice. However, breast conserving surgeries have become increasingly common in management of PT for cosmetic reasons. There is a relatively high incidence of local recurrence associated with PT and age, tumor size, surgical approach, mitotic activity, stromal overgrowth and surgical margin have been reported as prognosis-predictive factors related to local recurrence.[3] Herein we present a case of PT with rapid recurrence within 20 days of excisional biopsy, which is very early for PT in a young woman and is not the usual age group for the tumor.

CASE REPORT
The present case report is about a 20-year-old unmarried, nulliparous girl who presented with a lump, the size of a lemon in her left breast which was noticed while bathing. Over a period of 4 months, the lump increased in size gradually and progressively to about 6 cm × 6 cm. There was no family history of breast carcinoma. There was no palpable lymphadenopathy in either of the axillary basins. Mammography and ultrasound revealed a well-circumscribed mass in the lower outer quadrant of the left breast. An excisional biopsy was performed. Malignant PT was diagnosed on histology. Within 20 days of initial surgery, the patient returned with recurrence of the tumor. On examination, the whole left breast was occupied by a globular mass measuring about 15 cm × 15 cm, firm in consistency, with a smooth surface, not fixed to skin or chest wall. There was a single lymph node of 1 cm × 1 cm in left pectoral group of axillary lymph node. Nipple areolar complex was normal. The patient underwent a modified radical mastectomy (MRM). Grossly the MRM specimen showed a large gray-white invasive tumor with large areas of hemorrhage, in left upper outer quadrant of breast measuring 15 cm × 15 cm [Figure 1]. The histology showed a high grade neoplasm with large areas of...
Ahmad, et al.: Phyllodes tumor in young nulliparous

Clinical Cancer Investigation Journal | March-April-2014 | Vol 3 | Issue 2

necrosis [Figure 2]. The spindle cells were arranged in sheets and nests showing high grade of pleomorphism with brisk mitotic activity [Figure 3]. These findings were consistent with recurrence of malignant phyllodes. The skin, nipple, areola and single lymph node were negative for metastasis.

**DISCUSSION**

PT are fibroepithelial neoplasms with epithelial and cellular stromal components, the latter of which represents the neoplastic process, having a potential for recurrence and metastases. The incidence of PT is about 2.1/million, the peak is seen in women aged 45-49 years.[4] The tumor is rarely found in adolescents and elderly.[5] PT is divided into benign, borderline and malignant histotypes based on the microscopic appearance of the stromal component. Approximately 15-30% of all PT are classified as malignant.[6] Histologic appearance may not, however, correlate with clinical behavior, as both malignant and borderline tumors have been shown to be capable of metastasizing. The potential for PT to recur and metastasize was first recognized in the 1930s. Until date, local recurrence rates ranging from 10% to 40% have been reported, with most series averaging 15%.[7] A follow-up period of 30 months was used as the median time for recurrence is less than 24 months in almost all of the studies.[8] Local recurrence appears to be related to the extent of the initial surgery and should be regarded as a failure of primary surgical treatment; however in our case margins were clear. de Roos et al.[9] noticed that patients with recurrence have margin involvement on histological examination, but not all patients with the margin involvement developed recurrence. Whether malignant tumors have an increased risk of recurrence is unclear, but when it does occur it is invariably seen earlier than with benign tumors. Local recurrence usually occurs within the first few years of surgery and histologically resembles the original tumor. Occasionally, recurrent tumors show increased cellularity and more aggressive histological features than the original lesion. In most patients, local recurrence is isolated and is not associated with the development of distant metastasis as also in our case. In a minority of patients repeated local recurrence occurs, over a prolonged period with no survival disadvantage. This is often seen irrespective of either the histological type of the tumor or the extent of the specimen margins.

Local recurrence can usually be controlled by further wide excision and mastectomy is not invariably required. Mastectomy should, however, be considered for local recurrence after local surgery for borderline or malignant tumors. Kaprisi et al.[10] concluded that tumor size and surgical margins were found to be the principal determinant of local recurrence. Occasionally aggressive local recurrence can result in widespread chest wall disease with direct invasion of the underlying lung parenchyma. Isolated reports of good palliation in this situation with radiotherapy have been published.
Triple assessment by clinical, radiological and histological examination forms the fundamental basis for the evaluation of PTs. Current studies have found that new genetic mutation and intratumoral genetic heterogeneity can develop within the same tumor. These mutations could be the explanation of malignant behavior or recurrence of PT; for example, loss of expression of (P16INK4a) gene was found frequently in malignant PT, also activation mutations in and over expression of epidermal growth factor receptor gene are associated with progression in the grade.

CONCLUSION

This case has brought to light the need for a close follow-up in cases of PT and the importance of ensuring clear surgical margins for up to 1 cm. Metastatic deposits to axillary lymph nodes are rare, however metastasis to lung and bone are documented and should be looked for on follow-up visits.

REFERENCES