IOSR Journal of Dental and Medical Sciences (IOSR-JDMS)
e-ISSN: 2279-0853, p-ISSN: 2279-0861.Volume 15, Issue 12 Ver. II (December. 2016), PP 32-35
www.iosrjournals.org

Primary Angiosarcoma of Breast: A Rare Entity with Review of Literature

Dr. Iffat Jamal[1,] Dr. Kumari Bashundhara Lata[2,] Dr Surya Kant Nirala[3]
[1]*Tutor ,Department Of Pathology All India Institute Of Medical Sciences, Patna*
[2]*DNB Trainee,Mahavir Cancer Sansthan ,Patna*
[3]*Tutor ,Department Of Pathology All India Institute Of Medical Sciences,Patna*

Introduction: *Primary angiosarcoma is a rare malignant tumour that arise from endothelial cells lining the vascular channels.It is an aggressive tumour with a high propensity for metastasis. Primary angiosarcomas account for <1% of all breast cancer cases . The other more frequent histological subtypes encountered are malignant fibrous histiocytoma, fibrosarcoma and liposarcoma.[1] Most angiosarcomas are secondary to radiotherapy treatments for breast cancer or to an arm lymphoedema subsequent to a modified radical mastectomy.*
Keywords: *Primary,angiosarcoma,malignant, endothelial,vascular ,lymphoedema*

I. Case Report

We report a case of 36 year old female coming with a large fungating breast mass of size 20x18 cms occupying whole of right breast for 1 year which was hard in consistency , fixed to overlying skin and underlying breast parenchyma and chest wall.Her biopsy from the right breast mass suggested Phyllodes tumour .Simple mastectomy was performed and the mastectomy specimen was sent for histopatholgical examination for further confirmation.The simple mastectomy specimen measured 23x18x16 cm which showed a cauliflower like growth on the overlying skin .The overlying skin measured 18x10 cm.No nipple areola complex was identified grossly.On cutting a large tumour mass of sixe 20x18 m was noted with large areas of haemorrhage and necrosis.No normal breast tissue was identified grossly. An adjacent rim of pectoralis major muscle was attached to the breast mass which was seen grossly infiltrated by the tumour.(Figure 1 &2)

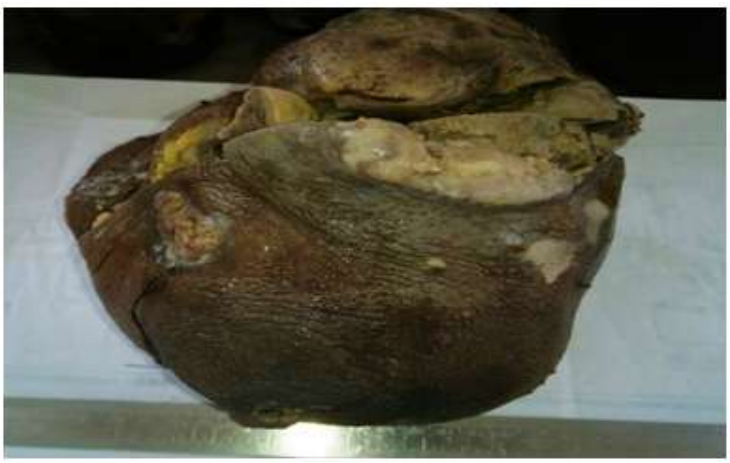

Figure 1: Gross specimen of right simple mastectomy showing large fungating tumour infiltrating the overlying skin with destruction of nipple and areola.

Histopathological examination showed that the mass was composed of vascular channels with a distinctive dissecting pattern between collagen bundles.The cells had high nuclear grade and were arranged in sheets. The vascular channels were lined by variably pleomorphic,hypechromatic endothelial cells with multilayering and papillary growths. The tumour showed high mitotic count (25/10 HPF), high nuclear grade and necrosis.

There was dermal invasion and ulceration and associated inflammation was found in the overlying epidermis.(Figure 3,4 & 5) . Immunohistochemistry revealed nuclear positivity of Fli-1 in endothelial cells which confirmed it to be of vascular origin.(Figure 6).

Figure 2: Cut section of mastectomy specimen showing large haemorrhagic and necrotic areas obscuring the breast parenchyma.

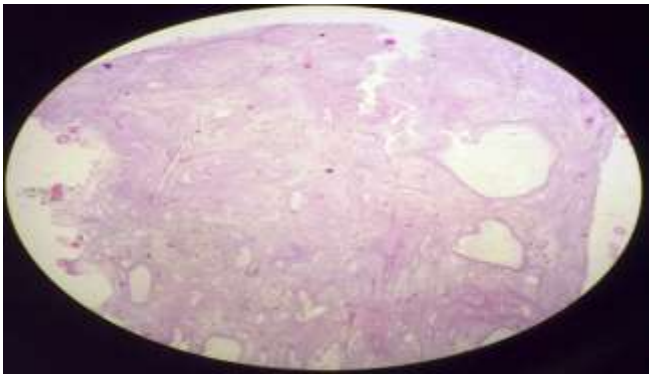

Figure 3. Microphotograph showing dilated and anastomosing vascular channels lined by atypical endothelial cells.(H &E;20 X)

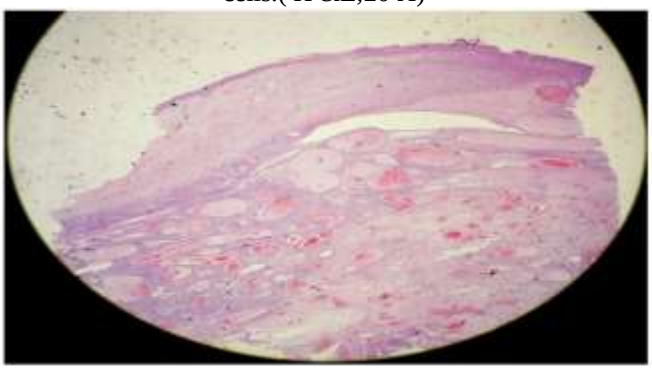

Figure 4: Microphotograph showing vascular spaces lined by atypical cells with large,hyperchromatic nuclei .(H & E;100X)

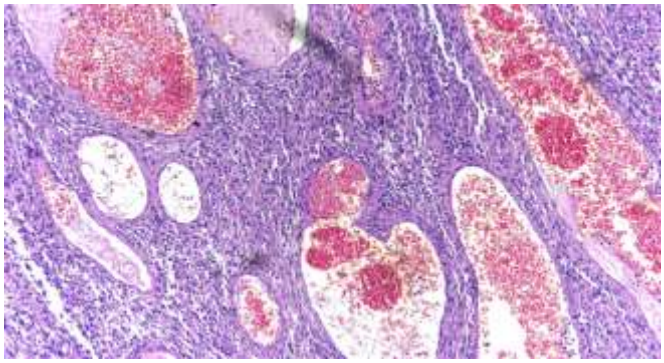

Figure 5: Microphotograph showing vascular spaces surrounded by spindle cells which are large,bizarre with presence of atypical mitiosis.(H & E;400X)

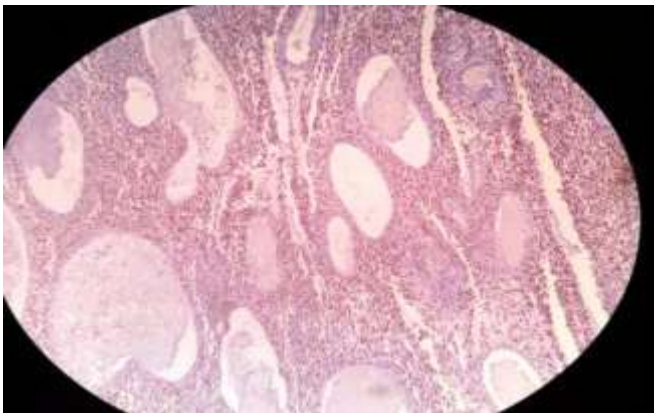

Figure 6 : Immunohistochemistry(IHC) showing diffuse nuclear positivity of Fli-1 in the endothelial cells.(100X)

II. Discussion

Breast angiosarcoma can be observed as a primary neoplasm or, more commonly, secondary to breast-conserving surgery combined to radiation therapy. Primary breast sarcomas and angiosarcomas represent, respectively, less than 1% and 0.04% of all breast cancers .[1] Breast angiosarcoma is more frequent in young women (20 to 50 years) with no previous cancer history or other known risk factors . Between 6 and 12% of primary breast angiosarcomas are diagnosed during pregnancy or shortly after, suggesting hormones involvement.[2] In most published cases, breast angiosarcoma presents as a palpable mass, without pain and with a fast growing rate . Large or superficial tumors often present purplish, ecchymosis-like skin coloration. In most cases, absence of pathognomonic characteristics specific to angiosarcomas will result in a wrong or delayed diagnosis . Diagnosis prior to surgery, either by FNA (Fine needle aspiration) or NCB(Needle core biopsy) , is always difficult.Radiological characteristics may help to establish right diagnosis, but, most often, as in the present case, mammography is unspecific and heterogeneous.[3] Sonography and MRI are useful in characterizing breast lesions, but again there is no distinctive features to angiosarcomas. As most soft-tissue sarcoma in most anatomical sites and of most histological subtype, angiosarcomas larger than 5 cm are associated to a shorter disease-free survival than angiosarcomas smaller than 5 cm. Indeed, tumors smaller than 5 cm are usually associated to a better prognosis, even in the presence of worsening factors.[4] With small tumors, recurrence is usually local and distant metastases are infrequent. However, tumors larger than 5 cm are associated to distant metastases, independently of local recurrence . There is a need for a close follow-up, including computed tomography scan at 6 months intervals. Sondenaa et al. conducted a survey of metastatic sites of all reported primary breast angiosarcoma. They concluded that the liver was the more frequent metastatic site, followed by lung, lymph nodes, bones, bone marrow and, less frequently, ovary, kidney, omentum, adrenal gland, stomach, pancreas, peritoneum, esophagus, and skin. Breast angiosarcomas are stratified according to three grades.Well-differentiated tumors (grade I) are composed of anastomosing vascular channels that surround breast ducts and infiltrate the adipose tissue. Blood vessels are lined by a single layer of endothelial cells with hyperchromatic nuclei showing little mitosis. No endothelial tufting is seen. Well-differentiated breast angiosarcomas are associated to a longer recurrence-free survival and to fewer distant metastases. Moderately differentiated angiosarcomas (grade II) are largely like well-differentiated tumors but show small foci of solid proliferation of spindle-shaped cells and more mitotic figures. Poorly-differentiated angiosarcomas present more solid and atypical cell proliferation and often show necrotic area and blood lakes. Grade III tumors are associated to decreased five-year survival rate and to increased metastases rate. Diagnosis prior to surgery, either by FNA or NCB, is at best difficult.[5] Chen et al. reported a percutaneous biopsy false-negative rate of 37% .[4,5] Differential diagnosis of this rare tumor include: benign hemangioma, cystosarcoma phyllodes, stromal sarcoma, metaplastic carcinoma, fibrosarcoma, liposarcoma, squamous cell carcinoma with sarcomatoid features, myoepithelioma, fibromatosis, and reactive spindle cell proliferative lesion.[6] Large-core macrobiopsies might be useful to improve diagnosis prior to surgery since a larger sample is taken. However, in the present case, the mass was so importantly hemorrhagic that a macrobiopsy would have been very difficult to perform.[7] Surgical resection and microscopic examination of sufficient sampling of the tumor are often necessary to render a final diagnosis. Immunohistochemical examination (factor VIII , CD31 and FLI-1 positivity) confirms the vascular nature of the tumor .[7,8] It is generally recognized that breast angiosarcomas have a bad prognosis. However, prognosis depends upon tumor grade (most important factor), tumor size at diagnosis, and margin status at surgery. Generally, 33% of patients with breast angiosarcoma, all grades together, are disease-free 5 years after initial diagnosis.[9] Most of patients with a grade I/III tumor are alive after 15 years. Grade III/III tumors are the most

aggressive, and the median disease-free survival is about 15 months.[10] It is now well recognized that complete surgical excision of breast angiosarcoma is the best course of action and that total mastectomy is the best option .[11] Since hematogenous dissemination is more than likely, axillary node dissection is not indicated . Chemotherapy and radiation therapy may be used as adjuvant treatment.[12] However, the role of adjuvant chemotherapy is ill defined, because of the rarity of primary breast angiosarcoma and of the lack of prospective studies . Most primary breast angiosarcomas are treated the same way as breast angiosarcoma secondary to radiation therapy.[13] In most reported cases with use of chemotherapy, authors prescribed cyclophosphamide, anthracycline, or an alkylating agents combined to a pyrimidine analogue. The endothelial cells show reactivity for several markers, including CD31, CD34 and von Willebrand factor (factor VIII).[14] Among them, CD31 is considered the most sensitive and most specific endothelial cell marker.[14-15]

III. Conclusion

A large and metabolically active vascular mass in the breast should always be considered at first sight to be an angiosarcoma, until proven otherwise. Primary breast angiosarcoma is a rare entity, very few cases have been reported so for which makes our case worth reporting. There is no general agreed course of treatment and no consensus has been achieved regarding advantage of adjuvat therapy. Hormonal treatment doesn't seem to be appropriate since these tumors usually do not express estrogen receptors. Even if these tumors have a bad prognosis, surgical treatment using total mastectomy is preferred; wide local excision may be selected depending on tumor size.

References

[1]. S. A. Vorburger, Y. Xing, K. K. Hunt et al., "Angiosarcoma of the breast," Cancer, vol. 104, no. 12, pp. 2682–2688, 2005.
[2]. X. Y. Wang, J. Jakowski, O. W. Tawfik, P. A. Thomas, and F. Fan, "Angiosarcoma of the breast: a clinicopathologic analysis of cases from the last 10 years," Annals of Diagnostic Pathology, vol. 13, no. 3, pp. 147–150, 2009.
[3]. J. S. Scow, C. A. Reynolds, A. C. Degnim, I. A. Petersen, J. W. Jakub, and J. C. Boughey, "Primary and secondary angiosarcoma of the breast: the Mayo Clinic experience," Journal of Surgical Oncology, vol. 101, no. 5, pp. 401–407, 2010.
[4]. M. Ohta, Y. Tokuda, S. Kuge et al., "A case of angiosarcoma of the breast," Japanese Journal of Clinical Oncology, vol. 27, no. 2, pp. 91–94, 1997.
[5]. T. Sher, B. T. Hennessy, V. Valero et al., "Primary angiosarcomas of the breast," Cancer, vol. 110, no. 1, pp. 173–178, 2007.
[6]. C. M. Johnson and G. A. Garguilo, "Angiosarcoma of the breast: a case report and literature review," Current Surgery, vol. 59, no. 5, pp. 490–494, 2002.
[7]. S. N. Georgiannos and M. Sheaff, "Angiosarcoma of the breast: a 30 year perspective with an optimistic outlook," British Journal of Plastic Surgery, vol. 56, no. 2, pp. 129–134, 2003.
[8]. L. D. Britt, P. Lambert, R. Sharma, and L. E. Ladaga, "Angiosarcoma of the breast: Initial misdiagnosis is still common," Archives of Surgery, vol. 130, no. 2, pp. 221–223, 1995.
[9]. K. N. Glazebrook, M. J. Magut, and C. Reynolds, "Angiosarcoma of the breast," American Journal of Roentgenology, vol. 190, no. 2, pp. 533–538, 2008.
[10]. Y. Kikawa, Y. Konishi, Y. Nakamoto et al., "Angiosarcoma of the breast—specific findings of MRI," Breast Cancer, vol. 13, no. 4, pp. 369–373, 2006.
[11]. W. Zeng, T. M. Styblo, S. Li, J. N. Sepulveda, and D. M. Schuster, "Breast angiosarcoma: FDG PET findings," Clinical Nuclear Medicine, vol. 34, no. 7, pp. 443–445, 2009.
[12]. R. C. Fields, R. L. Aft,W. E. Gillanders, T. J. Eberlein, and J. A. Margenthaler, "Treatment and outcomes of patients with primary breast sarcoma," American Journal of Surgery, vol. 196, no. 4, pp. 559–561, 2008.
[13]. K. Sondenaa, R. Heikkila, A. Nysted et al., "Diagnosis of brain metastases from a primary hemangiosarcoma of the spleen Case Reports in Oncological Medicine 5 with magnetic resonance imaging," Cancer, vol. 71, no. 1, pp. 138–141, 1993.
[14]. K. T. K. Chen, D. D. Kirkegaard, and J. J. Bocian, "Angiosarcoma of the breast," Cancer, vol. 46, no. 2, pp. 368–371, 1980.
[15]. L. Zelek, A. Llombart-Cussac, P. Terrier et al., "Prognostic factors in primary breast sarcomas: a series of patients with long-term follow-up," Journal of Clinical Oncology, vol. 21, no. 13, pp. 2583–2588, 2003.