



Case Report

Primary angiosarcoma of breast – an unusual breast malignancy

Saroj A. Bolde^a, Rajendra B. Madane^{*b}, Gopal A. pandit^c.

Dr. Vaishyampayan Memorial Government Medical College, Solapur, Opposite of Civil court, Civil Chawk, Solapur. Postal Code- 413003

ARTICLE INFO

Keywords:

Primary mammary angiosarcoma
Immunohistochemistry
CD 31

ABSTRACT

ABSTRACT- Primary angiosarcoma (De novo) of breast is an extremely unusual variant of breast malignancies and its incidence is about 0.05% of all primary breast malignancies. Here in, we are reporting an interesting case of Primary angiosarcoma (De novo) of breast in 84 yrs old female patient. Clinically patient presented with deeply located, non-tender, rapidly growing lump in breast. There was no history of breast surgery or radiotherapy. In the present case lumpectomy was done. On histopathological examination it was diagnosed as poorly differentiated angiosarcoma (grade III) and confirmed on immunohistochemistry.

©Copyright 2010 BioMedSciDirect Publications IJBMR - ISSN: 0976:6685. All rights reserved.

INTRODUCTION

Primary (De novo) mammary angiosarcomas arising from breast parenchyma are rare and the incidence is about 0.05% of all primary malignancies of the breast. Mammary angiosarcoma can be subdivided into 4 forms i.e. Primary (de novo) form in breast parenchyma, Secondary in skin and soft tissue of arm following ipsilateral radical mastectomy and subsequent lymphoedema – the Stewart Treves (S-T) syndrome, Secondary in skin and chest wall following radical mastectomy and focal radiotherapy, Secondary in skin or breast parenchyma or both following conservative treatment and radiotherapy [1].

Case Report

An 84 yrs old female patient presented with complaints of painless rapidly growing lump in right breast since 2 months. There was no history of previous surgery or radiotherapy. On clinical examination firm, non-tender lump which was fixed to the chest wall, measuring 10x8cm was seen. Overlying skin, nipple & areola were unremarkable and there was no evidence of bluish or red discoloration of overlying skin. There were no any palpable axillary lymph nodes. USG revealed benign neoplastic lesion suggestive of haemangioma, hence lumpectomy was performed and the specimen was sent to histopathology department.

Grossly, Specimen consisted of multiple pieces of soft, friable, spongy hemorrhagic mass, largest piece measuring 10x10x8cm. Cut surface was hemorrhagic with reddish brown discoloration. (Fig.I) Extensive sampling was done.

Microscopy of the mass revealed a tumour composed of inter anastomosing vascular channels lined by endothelial cells which showed prominent hyperchromatic pleomorphic nuclei exhibiting prominent nucleoli. Lumina of the neoplastic vessels were filled with RBCs. These vascular channels were intermingled with solid endothelial and spindle cell areas that showed necrotic foci &

numerous mitotic figures. Multinucleated highly pleomorphic giant cells were also seen. The solid cellular component was more than 50% of total neoplastic area. Areas of necrosis and large areas of haemorrhages were evident. The tumour was seen infiltrating adjacent muscle and fibrofatty tissue. Diagnosis of primary angiosarcoma of breast was made. (Fig. II and III)

On immunohistochemistry the tumour showed positivity for CD31 (fig.IV) .Thus, the case was finally diagnosed as primary angiosarcoma of breast and treated accordingly with Modified Radical Mastectomy followed by radiotherapy.

Fig. I - Gross specimen showing multiple pieces of soft, friable, spongy hemorrhagic mass, C/s - hemorrhagic with reddish brown discoloration.



Fig. II- Low power view showing inter anastomosing vascular channels intermingled with solid endothelial and spindle cell areas

* Corresponding Author :
drrajmadane@gmail.com

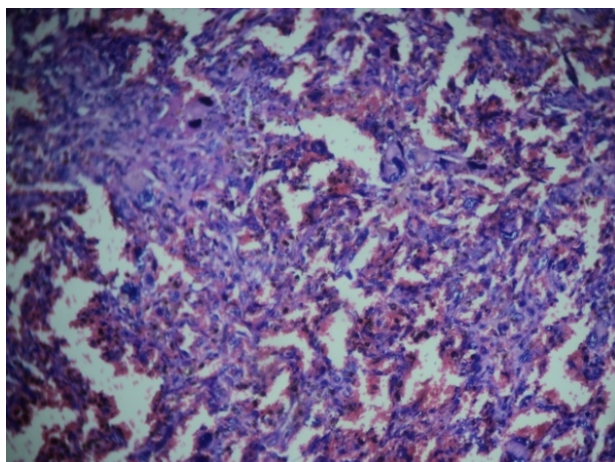


Fig.III- High power view showing highly pleomorphic giant cells.

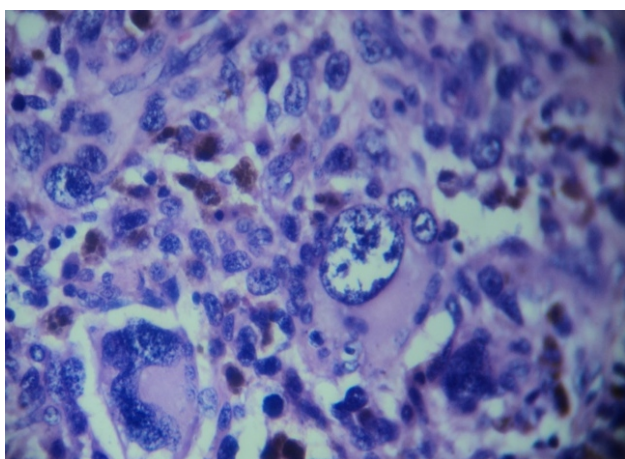
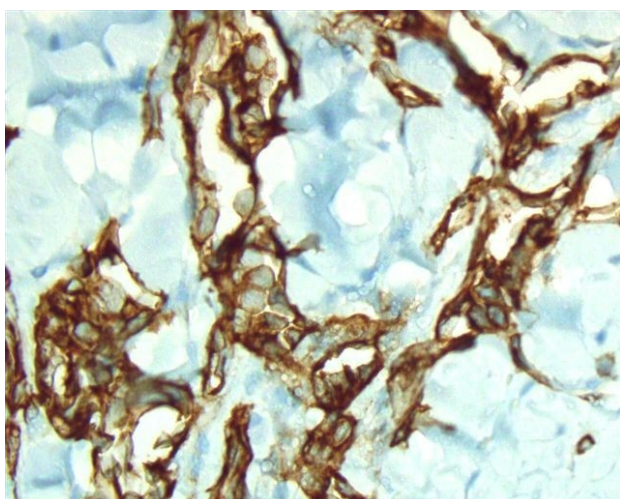


Fig. IV -Showing endothelial cells with immunopositivity for CD 31



Discussion:

Of all the cancers of the breast, carcinomas form the overwhelming majority while sarcomas are of negligible incidence. The breast sarcomas include fibrosarcoma, malignant fibrous histiocytoma, angiosarcoma and liposarcoma [1, 4]. Primary soft tissue sarcoma of the breast is a rare disease that accounts for less than 1% of breast malignancies [3, 4]. The estimated incidence of secondary (postirradiation) soft tissue sarcomas of the breast ranges from 0.01% to 0.02% per year, whereas the overall incidence of angiosarcoma of the breast is estimated to be between 0.002% and 0.05% per year [4]. Angiosarcoma was formerly known as hemangiosarcoma, hemangioblastoma or lymphangiosarcoma. But at present there are no reliable criteria to make a histological distinction between tumours derived from endothelium of blood vessels and lymphatic vessels [1]. Mammary angiosarcoma can be subdivided into 4 categories-

- 1- Primary (de novo) forms in breast parenchyma
- 2- Secondary in skin and soft tissue of arm following ipsilateral radical mastectomy and subsequent lymphoedema – the Stewart Treves (S-T) syndrome.
- 3- Secondary in skin and chest wall following radical mastectomy and focal radiotherapy.

Secondary in skin or breast parenchyma or both following conservative treatment and radiotherapy [1]

Age of the patients with primary angiosarcoma ranges from 17 to 70yrs, median age being 38 yrs. The average age of patient with grade III angiosarcoma is 29yrs. In present case this tumour was encountered in 84yrs old patient. Breast angiosarcoma presents as a rapidly growing, painless, palpable mass [2]. The tumour was deeply located in the breast tissue. Approximately 12% patients present with diffuse breast involvement. Large or superficial tumours often present with purplish, ecchymosis-like skin coloration [2]. Angiosarcomas vary in size from 1 to 20cm, average size being 5cm [4] have spongy appearance and rim of vascular engorgement which corresponds to a zone of well differentiated tumour. Poorly differentiated tumors are ill-defined, indurated fibrous lesion. Angiosarcomas of breast are graded as grade I (well differentiated), grade II (intermediately differentiated), and grade III (poorly differentiated). In grade III, >50% of the total neoplastic area is composed of solid and spindle cell components without evident vascular channels [1]. The present case is of poorly differentiated (grade III) angiosarcoma.

Cell block immunocytochemistry and tumour cell labelling with endothelial markers are necessary for accurate diagnosis [6]. The most widely used antibodies that characterize endothelial differentiation are factor VIII, CD34 and CD31. CD31 seems to be the most sensitive and specific for endothelial differentiation [5]. These markers are present in all grade I and most grade II angiosarcomas but these markers may be lost in more poorly differentiated tumours. Our case showed positivity for CD31 and CD34 and focal positivity for pancytokeratin and EMA on immunohistochemistry. Diagnosis was confirmed as primary angiosarcoma of breast. Low grade and intermediate grade tumours should be differentiated from haemangioma [1].

Prognosis of well differentiated angiosarcoma is better, intermediate and high grade angiosarcoma is usually lethal [1].

Estimated probabilities of disease free 5 years survival after initial treatment are as follows: TypeI: 76%; TypeII:70% & TypeIII:15%. The median length of disease free survival also was related to tumour type (Type-I >15yrs, Type II- >12yrs, Type III-15mths) [7].

Metastasis is mainly to lungs, skins, contralateral breast, bone and liver. Very rarely axillary lymph nodes show metastasis. Metastasis was not seen in this case.

In conclusion, primary angiosarcoma (De novo) of breast is an extremely unusual variant of breast malignancies. We are presenting this case for its rarity and uncommon clinical presentation. Meticulous histopathological examination and immunohistochemistry is mandatory to arrive at the diagnosis.

References:

1. Angiosarcoma. In: Tumors of breast and female genital organs World Health Organisation Classification of Tumors, pathology and genetics, 94-96.
2. Desbiens C, Hogue JC, Levesque Y. Primary Breast Angiosarcoma: Avoiding a Common Trap. Case Reports in Oncological Medicine 2011.
3. Pandey M, Mathew A, Abraham EK, Rajan B. Primary sarcoma of the breast. Journal of Surgical Oncology, 2004;87:121-125.
4. Vorburger SA, Xing Y, Hunt KK, Lakin GE, Benjamin RS, Feig BW, Pisters PW, Ballo MT, Chen L, Trent J, Burgess M, Patel S, Pollock RE, Cormier JN. Angiosarcoma of the breast. Cancer. 2005;104:2682-2688.
5. Lim RE, Goei R. Angiosarcoma of the Breast1, RadioGraphics. 2007; 27, S125-S130.
6. Markidou S, Karydas I, Papadopoulos S, Christodoulidou I, Skarpidi E, Maounis N. Fine needle aspiration cytology in primary breast angiosarcoma: a case report. Acta Cytol. 2010;54:764-70.
7. Rosen PP, Kimmel M, Ernsberger D. Mammary angiosarcoma: The prognostic significance of tumor differentiation. Cancer, 1988;62:2145-2151.