

Angiosarcoma of Omental Mass: A Case Report

Sujata S. Giriyan¹, R. Lakshmi²

¹Professor and Head, Department of Pathology, Karnataka Institute of Medical Sciences, Hubballi.

²Postgraduate Student, Department of Pathology, Karnataka Institute of Medical Sciences, Hubballi

Abstract: *Angiosarcomas are rare malignancies of endothelial origin. Peritoneal angiosarcomas are rare and behave aggressively, resulting in 100% mortality. Its incidence is approximately 0.01 per 100,000 cases.. A 55 year old female presented with complaints of vomiting for 2 days. USG abdomen showed? Mesothelioma with moderate ascites and bilateral pleural effusion. CT of abdomen and pelvis showed? Ovarian neoplasm /? Mesothelioma . On histopathology, the diagnosis was consistent with angiosarcoma. IHC for CD 34 was positive. Angiosarcomas are often misdiagnosed and the rate of tumour-related death is high with a 5-year survival rate of only 20%.*

Keywords: Angiosarcoma, Omentum

1. Introduction

Angiosarcoma is an aggressive, rapidly proliferating and quickly metastasizing malignant neoplasm, derived from endothelial cells and forming irregular blood-filled spaces. [1] They account for a vanishingly small proportion of all vascular tumours, and they comprise less than 1% of all sarcomas. [5]

Peritoneal angiosarcomas is an extremely rare sarcoma. They are difficult to differentiate from other malignancies, such as mesotheliomas and behave aggressively, resulting in 100% mortality. Its incidence is approximately 0.01 per 100,000 cases, usually in older adults but can develop at any age. [1]

We report here a case of angiosarcoma of omental mass which is rare and often misdiagnosed.

2. Case Report

A 55 year old female presented with complaints of vomiting for two days. On examination, abdomen was distended with a vague mass in right iliac region measuring 10×8 cm, which was soft and non-tender. USG abdomen showed heterogeneous lesion measuring 21×18×12cm in the abdomen? Mesothelioma with moderate ascites and bilateral mild pleural effusion. CT of abdomen and pelvis showed large solid heterogeneous density intraperitoneal lesion with inhomogeneous enhancement in the lower abdomen and pelvis. ? Ovarian neoplasm /? Mesothelioma.

Ascitic fluid analysis showed 500 cells/cumm, with 60% lymphocytes, 10% neutrophils and 30% mesothelial cells. Negative for atypical cells/ malignant cells. CA125 was raised (376mIU/ml). CT of chest showed no obvious abnormality.

She was taken up for surgery under the diagnosis? GIST /? Ovarian malignancy. Intraoperatively was seen a huge haemorrhagic mass covering whole of peritoneal cavity measuring 19 cm in diameter, soft to firm in consistency with well-defined margins. The upper border of the Tumour? Pedicle was attached to anterior wall of stomach and lower border to highly vascular omentum.

Excision of haemorrhagic mass with total abdominal hysterectomy with bilateral salpingo-oophorectomy with infracolic omentectomy was done and sent for HPR.

3. On Gross Examination

Specimen consisted of uterocervix with bilateral adnexa measuring 6 cms. External and cut surface was unremarkable. Right ovary measured 2×1×0.5 cms. Right fallopian tube measured 5cm in length. External and cut surface was unremarkable. Left ovary measured 2×1.5×1cm. Left fallopian tube was 4cms in length. External and cut surface was unremarkable.

Separately sent specimen was grey to grey brown fibro fatty tissue which measured 40× 9×2 cms. External and cut surface was unremarkable.



Figure 1(A): Gross picture showing uterus, cervix with attached bilateral adnexa . (B) Gross picture showing cut surface of utero cervix . (C) Gross picture showing specimen of omentum

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A single globular mass was sent separately which was grey brown to grey black measuring 19 cms in diameter. External surface was congested and nodular in some areas, soft in consistency with some areas being cystic. Cut surface was

predominantly haemorrhagic with grey brown to grey white solid areas with mucoid filled and necrotic areas.



Figure 2 (A): Gross picture of separately sent haemorrhagic mass .(B) Gross picture showing cut surface of haemorrhagic mass

4. On Microscopy

Sections studied from uterocervix with bilateral adnexa and separately sent omentum were unremarkable. Multiple sections studied from mass arising from anterior wall of stomach showed malignant neoplasm. These tumour cells were arranged in sheets predominantly around thin vascular

channels. These cells were round to polygonal with vacuolated cytoplasm, pleomorphic nuclei and coarse chromatin. Few cells showed intra-cytoplasmic lumina formation. Some of the areas showed well- formed multiple, anastomosing vessels and extensive haemorrhage.

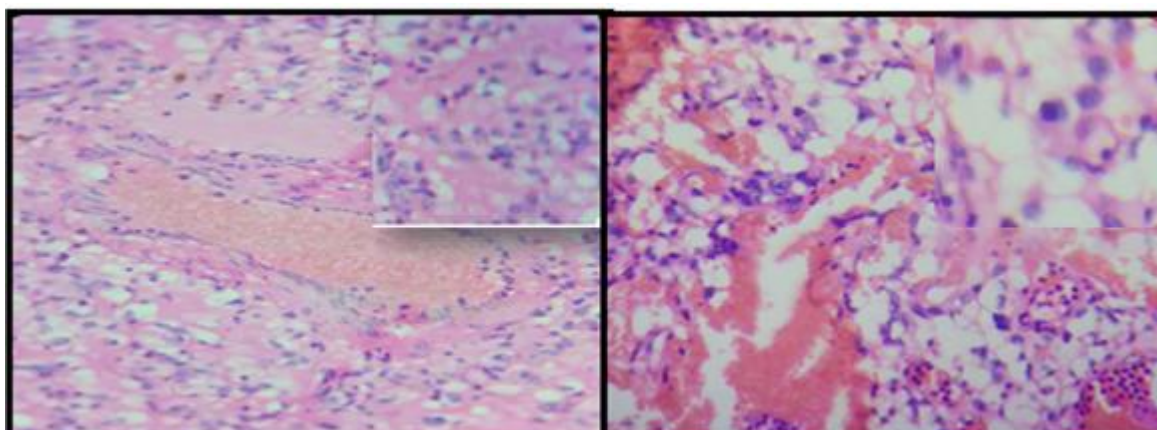


Figure 3: Showing tumour cells which are arranged in sheets predominantly around thin vascular channels (H&E, X40)

5. On Immunohistochemistry

Tumour cells were positive for CD34 and negative for S100.

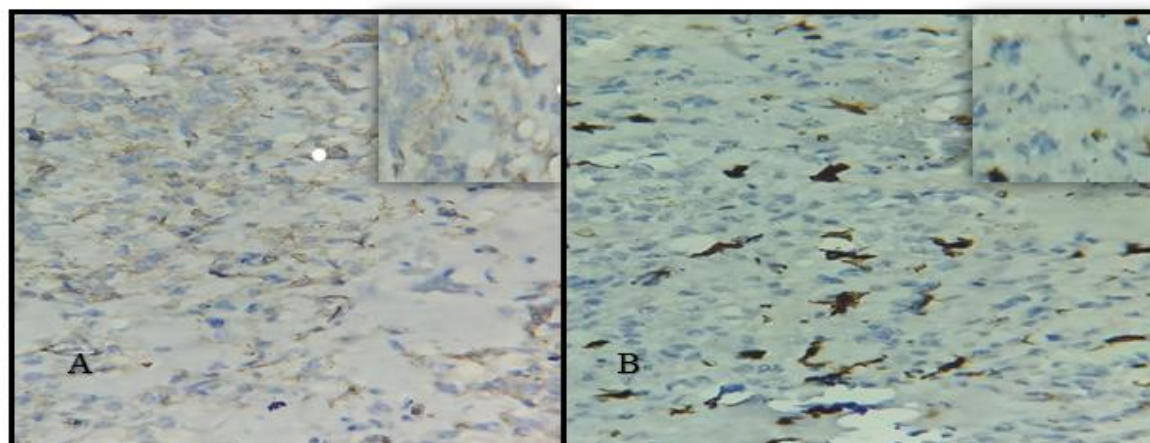


Figure 4 (A): Tumour cells were positive for CD34(x40). (B)Tumour cells were negative for s100(x40).

Based on gross, microscopic and immunohistochemistry features diagnosis of angiosarcoma was made.

6. Discussion

Angiosarcomas are a wide range of malignant endothelial vascular neoplasms that can arise from vascular elements of a variety of sites [1] but is more common in soft tissue than in bone.

The peak age of incidence appears to be the 7th decade, and men are affected more than women (Our case is of a 55 year old female).

They can be linked to various predisposing factors and risk factors such as genetic predisposition (Li Fraumeni syndrome, neurofibromatosis type 1), exposure to chemicals (vinyl chloride and thorium dioxide), radiation therapy (increases the relative risk of developing secondary angiosarcomas by 10-fold or 26.7-fold when combined with chemotherapy), chronic inflammation and lymphedema. [2] Angiosarcomas are observed after lymphedema from any cause, be it surgical, filarial or congenital, and defined as Stewart-Treves syndrome. [4] The head and neck area is probably the most common site of diagnosis, and the most common site of radiation induced angiosarcoma development is the breast. [4] Our case did not have any history of radiation.

CA125 was raised (376 mIU/ml) for which patient was taken up for surgery under the diagnosis of? Ovarian neoplasm. CA125 is raised in conditions like Ovarian cancer, Uterine cancer, Fallopian tube cancer, other intra-abdominal cancers (stomach, colon) and metastases from other sites (e.g. breast, lung). It is also raised in other non-malignant conditions like Endometriosis, PID, Leiomyoma, Pancreatitis etc.

Peritoneal angiosarcomas are rare tumours of the peritoneum which may be histologically difficult to differentiate from other malignancies, such as mesotheliomas and they usually behave aggressively, resulting in 100% mortality. It may arise at the site of previous radiation treatment to the serous membranes. [1] In our case both CT and USG abdomen showed? Mesothelioma.

Most diffuse mesotheliomas are of the epithelial type and can be identified by their characteristic tubulopapillary pattern. This pattern consists of papillary structures, branching tubules, and gland-like acinar and cystic spaces lined by rather uniform cuboidal or flattened epithelial-like cells with vesicular nuclei, one or two nucleoli, and abundant eosinophilic cytoplasm with distinct cytoplasmic borders [5] so diagnosis of mesothelioma was ruled out.

Microscopically liposarcoma was considered as differential diagnosis. Pure or predominantly myxoid liposarcomas are multinodular, gelatinous masses usually devoid of necrosis, yellow to white although occasionally haemorrhage is encountered which was not seen in the gross specimen. Microscopically, the tumour is composed of cells with lipoma like features except for the presence of scattered hyperchromatic, often multinucleated and vacuolated cells

with features of atypical lipoblasts. On IHC S100 was negative which ruled out liposarcoma. [5]

Soft tissue angiosarcomas are multinodular haemorrhagic masses often with secondary cystic degeneration and necrosis. They display a wide spectrum of morphological appearances, ranging from areas of well-formed, anastomosing vessels to solid sheets of high-grade epithelioid or spindled cells without clear vasoformation. [4] Solid areas lacking vasoformation are composed of high-grade spindled and epithelioid cells with abundant amphophilic to lightly eosinophilic cytoplasm, large vesicular nuclei and prominent nucleoli [4]. So diagnosis of angiosarcoma was considered.

The vast majority of angiosarcomas of soft tissue are high-grade neoplasms with brisk mitotic activity, coagulative necrosis and significant nuclear atypia. Careful sampling may be necessary to document malignant cells.

By gene expression profiling angiosarcomas show distinct up regulation of vascular-specific receptor tyrosine kinases, including TIE1, KDR, TEK and FLT1, compared with other sarcoma types. Furthermore, high level MYC amplification on 8q24 is a consistent hallmark of radiation-induced and lymphedema-associated angiosarcoma. [4] FLT4 (encoding for VEGFR3) co-amplification on 5q35 is detected in 25% of secondary angiosarcoma. [4]

MYC has a crucial role in growth control, differentiation and apoptosis, and its aberrant expression is associated with several cancers. More recently, MYC was also implicated as having a major contribution in tumour angiogenesis. [4]

In immunohistochemistry, anti-CD31 antibodies are one of the most specific endothelial cell markers since CD31 (platelet endothelial cell adhesion molecule) is a highly sensitive, specific antigen for endothelial differentiation.

In addition, factor VIII is also a related antigen which identifies tumours of endothelial origin. CD34 tends to stain areas with readily apparent vessel formation and its utility in poorly differentiated lesions is limited.

Flt-1 is the reliable nuclear marker for endothelial differentiation and its immune reactivity in vascular tumours exceeds that of CD31. [6]

Staining for Ki-67 with MIB-1 is usually 10% or greater in 72% of angiosarcomas evaluated, confirming the higher proliferative nature of these neoplasms. [6]

Features that are associated with poor outcome include older age, Retroperitoneal location, large size and high Ki-67 values. [4]

The primary treatment of angiosarcoma is radical surgery with complete resection and adjuvant radiotherapy for the local disease. This is not always possible taking into account the site of origin and tumour extension. Novel strategies to treat angiosarcoma including inhibition of the tyrosine kinase activity of VEGFRs, which can also decrease the

permeability of vessels and the formation of malignant ascites.[2]

The recommended chemotherapy for metastatic angiosarcoma includes anthracyclines, taxanes and ifosfamide. In addition low-dose liposomal doxorubicin or doxorubicin, alone or in combination, have shown positive results.[2]

7. Conclusion

Angiosarcomas are often misdiagnosed and the rate of tumour-related death is high with a 5-year survival rate of only 20%. However this case could not be followed up.

The rarity of peritoneal angiosarcoma makes it difficult to obtain insight into the pathobiology of this tumour.

Therefore, every case in which angiosarcoma is eventually diagnosed

should be thoroughly analysed with the potential aim of identifying signs, symptoms and biomarkers that differentiate this cancer from other closely-related cancers.

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