

# Primary Angiosarcoma of Ovary- A case report with review of literature

Naseer D. Chowdhary\*, Tahir M.Malik\*\*, Mansoor A.Laharwal\*, S.Manzoor Kadri\*\*\*

## Abstract

Primary ovarian angiosarcoma is an extremely rare tumour. Only 17 histologically proven cases have been reported till date in the medical literature. A case of angiosarcoma arising from the right ovary in a 41 year old female is reported. Grossly the resected ovary was replaced by a haemorrhagic brownish white tumour mass, with areas of necrosis. Microscopy revealed typical features of angiosarcoma with sinusoidal and solid patterns of anaplastic tumour cells. A total of 18 cases reported as Primary ovarian angiosarcoma, including the present case are clinico-pathologically reviewed.

## Key words

Angiosarcoma, Ovary, Sinusoidal, Solid tumour

## Introduction

Angiosarcoma constitutes only 2 percent of all the soft tissue sarcomas with a five-year survival varying between 10-35%. Angiosarcoma arising in the female reproductive system is exceedingly rare. It has been most frequently reported in the uterus and also in the vagina. It can also arise in the vulva and the cervix (1-4). 17 cases of Primary angiosarcoma of the ovary have appeared in the world literature (5). We present an additional case and describe the clinicopathological characteristics of these 18 cases.

## Clinical Summary

A 41 year old female reported to us with a history of a swelling in the right iliac fossa over the previous two months, associated with pain. There was no history of any menstrual abnormality, bowel or urinary symptoms. 2 years ago she had undergone a lapotomy for a left

sided ovarian mass for which a left sided salpingo oophrectomy was performed. Histopathology suggested a histiocytoma and she received postoperative chemotherapy.

Abdominal examination revealed a solid mass about 9x 8 cm size, fixed with no ascitis. Rest of the physical examination was normal. Vaginal examination confirmed the presence of a solid right adenexal mass, which was fixed to the lateral pelvic wall, the uterus felt normal and separate from the swelling.

Ultrasonography of the pelvis confirmed the presence of a solid mass 10x10 cm size in the right ovary, absence of ascitis and a normal sized anteverted uterus. There were grade I hydronephrotic changes in the right kidney and grade III hydronephrotic changes in the left kidney as well.

From the Departments of \*Pathology, \*\*\*Microbiology, Government Medical College, Srinagar and \*\*Department of Gynaecology and Obstetrics S.K.I.M.S Medical College, Bemina, Srinagar (J&K).

Correspondence to : Dr. Nasser.D. Chowdhary Post Box: 776 GPO, Srinagar-190001 Kashmir, India

Laboratory investigations revealed Hb- 10 Gms%, TLC, DLC-within normal limits. Normal urea, creatinine, sugar and LFT. CA 125 and CEA were within normal limits.

A laparotomy was performed on 30/03/2001 which revealed a boss elated tumour about 12x12 cm in size in the right ovary, which was adherent to the sigmoid colon and the rectum, infiltrating between the leaves of the broad ligament up to the pelvic wall and covering the ureter and the iliac vessels. The tumour was very friable and bled on touch. A subtotal abdominal hysterectomy along with right-sided salpingo oophrectomy was performed, involving extensive bowel dissection. During the process of dissection the tumour bled profusely necessitating blood transfusion. As the bleeding was uncontrollable a few abdominal packs were placed in the pelvic cavity. The patient was haemodynamically stable post operatively. The abdominal packs were removed 48 hours postoperatively. The patient developed a fecal fistula on the seventh postoperative day but there was no distal obstruction. She was managed conservatively for the same and opted to go home on the 20th day of operation. She reported after a fortnight in a much-improved condition. She is still under follow up (2 months) and will be given chemotherapy in consultation with oncologist.

**Pathological findings**

Grossly a fragmented brownish white biopsy specimen, with necrotic and haemorrhagic areas, together 12 cms in diameter was received which was immediately fixed in 10 percent formalin and processed using routine methods. Microscopically the tumour was composed of sinusoidal vascular channels (Fig I) and had a solid appearance with features of slit like structure (Fig II). Reticulin staining revealed tumour like structures surrounded by reticulin fibers that out lined the vascular lumina in each tumour nest. The tumour cells had large hyperchromatic nuclei with marked atypia. Abnormal mitotic activities were frequently observed. Foci of necrosis were seen here and there. The tumour cells infiltrated the capsule.

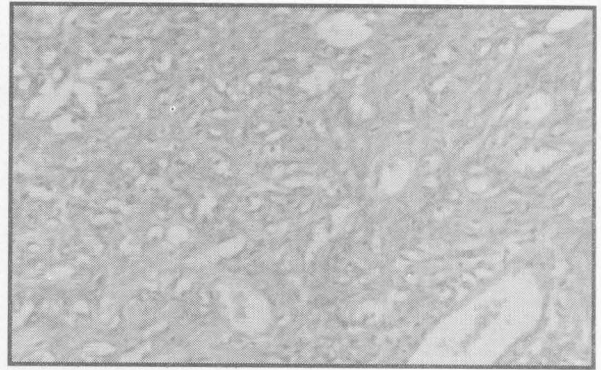


Fig I. Vascular Channels with solid nests of cells. H&E 10x

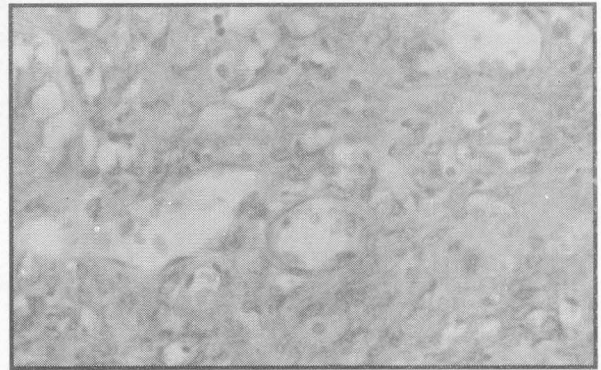


Fig II. Vascular Channels with solid nests of cells and mitotic activities. H&E 40x

**Discussion**

Though we did not have facilities for ultra structural study and immuno- histochemistry available with us, the characteristic histological features including the sinusoidal and solid pattern with necrosis, haemorrhage, numerous mitotic figures and infiltration of the capsule and surrounding structures, justified the diagnosis of angiosarcoma. Use of antibodies specific for endothelial cells, such as factor VIII- related antigens, CD31 and CD34 lead to a definite diagnosis of angiosarcoma. Less than 25% of angiosarcomas however contain sufficient amount of factor VIII- related antigen to be detected by routine immunohistochemical study (5,6).

About 40 cases of benign vascular tumours of ovary have been reported till date (7). These lesions do not exhibit the histological characteristics of malignancy, with no necrosis, mitotic figures and tumour infiltration of surrounding structures.

As the primary ovarian angiosarcoma is very rare, so the possibility of metastatic angiosarcoma of ovary needs to be looked into. In our case no other primary site was detected. 18 cases (including this case) have affected the age group 19 to 77 years (Refer Table). Involvement of right ovary was twice more common. In one case bilateral ovarian involvement is reported. The predilection for right ovary is to be looked into. The lack of alarming early symptoms such as vaginal bleeding might be responsible

for the far advanced clinical course of the reported cases. The most frequent symptoms included discomfort and pain in the lower abdomen with a mass on ultrasonography and generalized fatigue. In general, angiosarcoma arising from any part of the body is an aggressive tumour with high propensity for local recurrence as well as distant metastasis, and has poor prognosis (1). Early detection of the disease may lead to more appropriate management and improvement in the survival.

#### Summary of Clinico-Pathological Findings of Reviewed Cases.

S. No.	Age in years/ Nationality	Symptoms	Side and Size	Histological Diagnosis	Therapy	Outcome after Therapy	Ref.
1	35/ Coloured	Abd. pain	Rt.	Hemangioendothelioma	Salpingo-oophorectomy	21 Months	2
2	77/ White	Generalized weakness	Rt. 8.8 x 2.5 cms.	Mucinous cyst adenoma and Angiosarcoma	Exploratory laparotomy	2-5 Months Autopsy	3
3	42/ Nepalese	Tiredness	Rt. 4.5 cm	Angiosarcoma	Salpingo-oophorectomy	2 Months Autopsy	5
4	19/White	Abd. discomfort, pain, fatigue	Lt. 12x10 cm	Angiosarcoma	Salpingo-oophorectomy	7 Months	5
5	33/Japanese	Persistent cough	Rt. 6x4 cm Lt. 3	Angiosarcoma	Salpingo-oophorectomy	—	—
6-12	20-32 Mean 26/ No data	Pain abd. and distention	Rt. 3 B/L 1 6-13 cm	Angiosarcoma	Not Mentioned	—	7
13-16	25-42 Mean 26 No data	Pain abd. with weight loss, Hemoperitoneum	Unilateral	Angiosarcoma	Salpingo-oophorectomy	—	5
17	46/Japanese	Abd. mass	Rt. 21x16x1 cms 2750 gms	Angiosarcoma	Salpingo-oophorectomy	2 Months	5
18	41/Indian Kashmiri	Swelling rt. iliac fossa	Rt. 12x12 cms	Angiosarcoma	Salpingo-oophorectomy	Alive till date.	Present case

Rt = Right, Lt = Left, Abd = Abdomen, B/L = Bilateral

**References**

1. Mark RJ, Poen JC, Tran LM, Fu YS, Juillard GF. Angiosarcoma: A report of 67 patients and review of the literature. *Cancer* 1996 ; 77 : 2400-06.
2. Sovak FW, Carabba V. Hemangio-endothelioma intravasculare of the ovary. *Am J Obstet Gynecol* 1931 ; 21 : 544-49.
3. Ongkasuwan C, Tylor JE, Tag CK, Prempre T. Angiosarcoma of the uterus and ovary: Clinicopathological report. *Cancer* 1972 ; 49 : 1469-75.
4. Miltne DS, Hinshaw K, Malcolm AJ, Hilton P. Primary Angiosarcoma of the uterus: A case report. *Histopathology* 1990 ; 16 : 203-05.
5. Matsuo F, Tamtsu T, Jun I, Hiroshi S, Yuji O, Akihiko W, Noguyki M, Yusuke S. Primary ovarian Angiosarcoma : A case report and review of literature. *Pathol International* 1998 ; 48 : 967-73
6. Mukai K, Rosai J, Burgdorf WHC. Localization of factor VIII-related antigen in vascular endothelial cells using an immunoperoxidase method. *Am J Surg Pathol* 1980 ; 4 : 273-76.
7. Nielsen GP, Young RH, Prat J, Scully RE. Primary Angiosarcoma of ovary: A report of 7 cases and review of the literature. *Int J Gynecol Pathol* 1997 ; 16 : 378-82.



**Products :**

- Syp. **FAROCID**
- Cap. & Syp. **LYRONE**
- Cap. **XEROCID (20)**
- Tab. **KALICET**
- Tab. **FAMONAC-MR**
- Tab. **CIFROX (500)**

*For further details please contact :*

**M/S MEGHA-POOJA MEDICAL AGENCIES (C&F)**

with F/17, Shiv Nagar behind A. G. Office, Jammu, J&K.

*Stockist :*

**Bee Kay & Co.**

Bhagunath Bazar, Jammu, J&K.