

# CASE REPORT

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## SPINDLE CELL SARCOMA OF VAGINA- A CASE REPORT

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**ABSTRACT:** Malignant tumors of vagina are rare accounting for 1 to 4 % of all genital malignancies. Rarest of rare is spindle cell sarcoma of vagina having a very poor prognosis. 47yrs old, regularly menstruating woman, completed family, presented with hard painful ulcerated swelling near the vaginal introitus and lower vaginal wall of 4 months duration. On examination, 2x4cms sized ulcerated growth with excavated base, covered with necrotic material, present in the left antero-lateral lower end of vagina, inner to hymenal ring which was tender, hard, indurated, infiltrating, fixed to base and did not bleed to touch. Another nodule of size 1x1cm tender, hard, fixed and necrotic was present below the external urethral meatus. HPE revealed Amelanotic malignant melanoma of vagina. Immunohistochemistry revealed Spindle cell sarcoma of vagina. Tumor board recommended neo-adjuvant chemotherapy and radiotherapy (CT + EBRT). Of the recommended 50Gy, she completed 46Gy in 23days and 1 course of chemotherapy (VAC). On review after 9 months, the lesion disappeared clinically and she was advised to complete the treatment. In spite of radio and chemotherapy secondaries to lungs and brain could not be prevented and the patient expired 20 months after the final diagnosis.

**KEY WORDS:** spindle cell sarcoma, immunohistochemistry

**INTRODUCTION:** Malignant tumors of vagina are extremely rare and accounts for 1 to 4% of all gynecological malignancies.<sup>3</sup> Rarest of rare is spindle cell sarcoma of vagina having a very poor prognosis. Newer diagnostic techniques like immunohistochemistry are now available to pick up even submicroscopic diagnosis of many pathological lesions.<sup>1,2</sup>

**CASE REPORT:** A 47 years old P<sub>2</sub>L<sub>2</sub>A<sub>0</sub> woman came to gynecology out-patient department with painful swelling near the vaginal introitus of 4 months duration. She was not menstruating for the past 7 months and previously her menstruation pattern was 2 / 30 days regular cycles with normal volume without dysmenorrhea. She had 2 vaginal deliveries and was sterilized. Her personal and family history was nil relevant. She was moderately built and nourished, conscious, well oriented. She was restless, intensely suffering with pain and distressed. She was pale. There was no icterus, no clubbing, no cyanosis, no pedal edema. There was no significant lymphadenopathy. Her blood pressure was 120/80 mm Hg and her pulse rate was 72/min. she was afebrile. R.S, C.V.S and C.N.S were clinically normal. Examination of the abdomen was normal. External genitalia appeared normal. Speculum examination revealed an ulcerated growth of size 2x4cms with excavated base, covered with necrotic material, present in the left

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antero-lateral lower end of vagina, inner to hymenal ring which was tender, firm to hard in consistency, indurated, infiltrating, fixed to base and did not bleed to touch. Another nodule of size 1x1cm tender, hard, fixed and was necrotic present below the external urethral meatus. Per vaginal examination revealed cervix healthy; pointing down; uterus normal in size, mobile, fornices free.

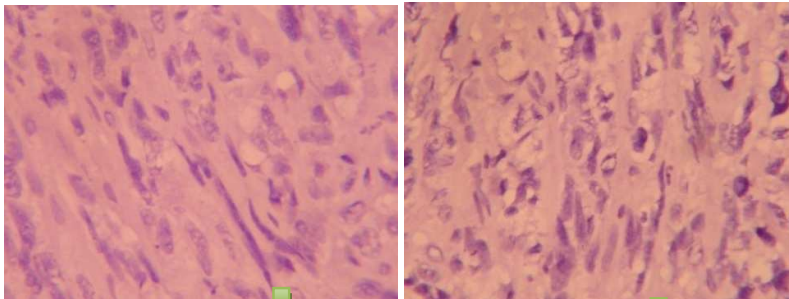


Ulcerated growth with excavated base with necrotic material

FIG.1.

## INVESTIGATIONS:

Routine blood and urine examination were within normal limits. Pap smear from cervix showed acute inflammatory cells. There was no evidence of malignancy. Chest X-ray was normal; CT chest was normal; CT pelvis- plain showed mass lesion in the region of cervix with RT markers in situ. On transabdominal sonogram, uterus, ovaries and tubes were normal. No hydronephrosis. No post voidal residual urine. Liver, spleen, kidneys were normal. HPE revealed "RARE AMELANOTIC MALIGNANT MELANOMA" of the vaginal mucosa. IMMUNOHISTOCHEMISTRY revealed Cytokeratin- negative; EMA- negative; HMB 45 - negative; Vimentin - positive; SMA - negative; Desmin - negative. Report was mesenchymal spindle cell malignancy with ulceration - SPINDLE CELL SARCOMA. Patient referred to cancer institute. For staging workup, CT- chest was done in the cancer institute. CT -chest was normal.



CYTOKERATIN- NEGATIVE

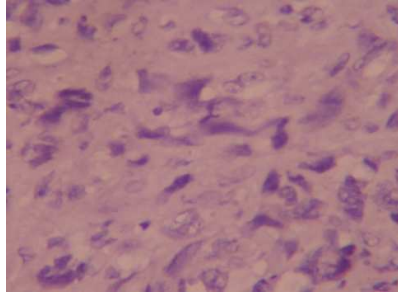
EMA - NEGATIVE



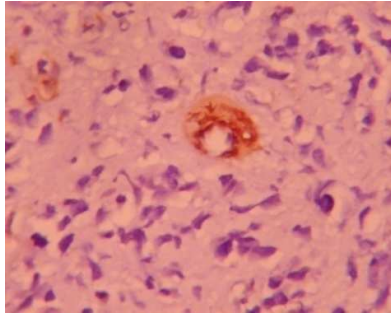
**RULES OUT CARCINOMA**

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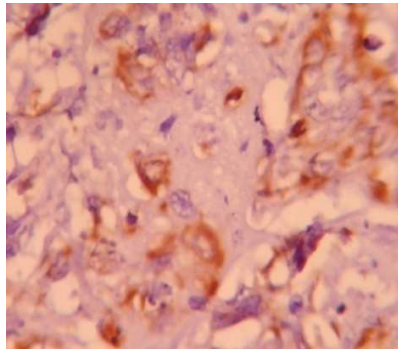
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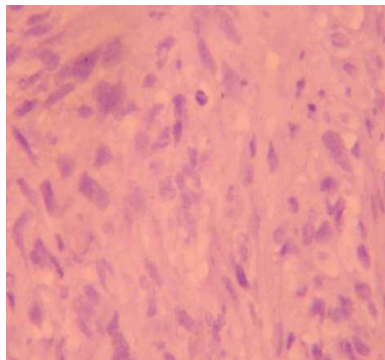
**DESMIN – NEGATIVE –  
RULES OUT  
RHABDOMYOSARCOMA**



**SMA- NEGATIVE –  
RULES OUT  
LEIOMYOSARCOMA**



**HMB 45 – NEGATIVE  
RULES OUT  
MELANOMA**



**VIMENTIN – POSITIVE IN  
ANY MESENCHYMAL  
TUMOUR**

**FINAL DIAGNOSIS: SPINDLE CELL SARCOMA OF VAGINA???**

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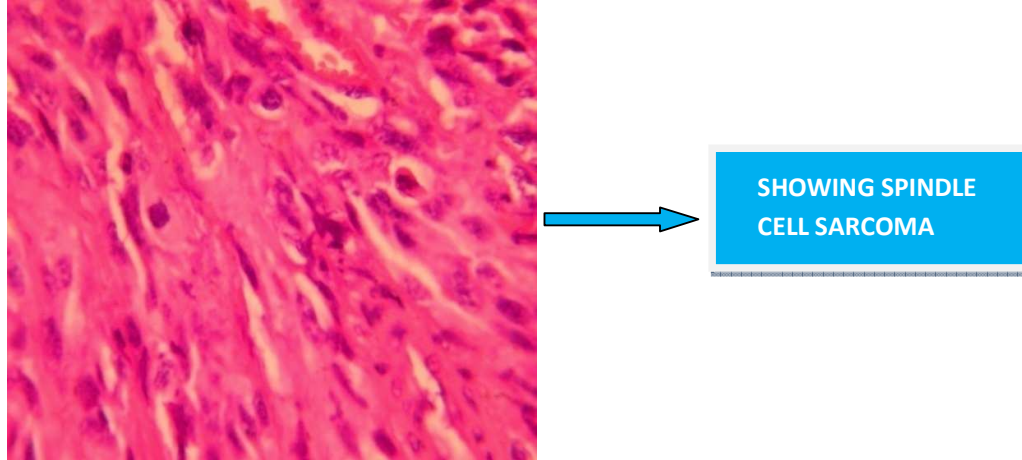


FIG.2. IMMUNOHISTOCHEMISTRY SLIDES

Rhabdomyosarcoma was ruled out as Desmin was negative. There were no vascular/ neural elements in the HPE, so the markers for angiosarcoma/ Schwann cell tumor namely CD 34, CD 31 and S100 respectively was not done. CD 68 is the marker for malignant fibro histiocyte. Our definitive diagnosis is more in favour of "**HIGH GRADE UNDIFFERENTIATED SARCOMA OF VAGINA**" whose incidence is extremely rare. Since there was not much of discrimination in the plan of treatment modalities among different types of sarcoma except for rhabdomyosarcoma, the treatment was planned. Plan at the start of the treatment was curative by the tumour board and the policy was, if operable – surgery + RT and CT and if inoperable – neoadjuvant therapy ie CT and RT followed by surgery. Surgical oncologist advised neoadjuvant CT and EBRT. 50 Gy was planned to be given in 25 days. But she received only 40 Gy over 23 days and 1 cycle of chemotherapy with VAC after which she was on irregular treatment. After 9 months, she came for review and there was no lesion clinically. She was advised to continue and complete the course of the treatment. On follow up of the patient inspite of radio and chemotherapy secondaries to lungs and brain could not be prevented and the patient expired 20 months after the final diagnosis. If not for the precise guidance and predictions given by our pathology team, the possibility of arriving at this diagnosis would have not been possible.

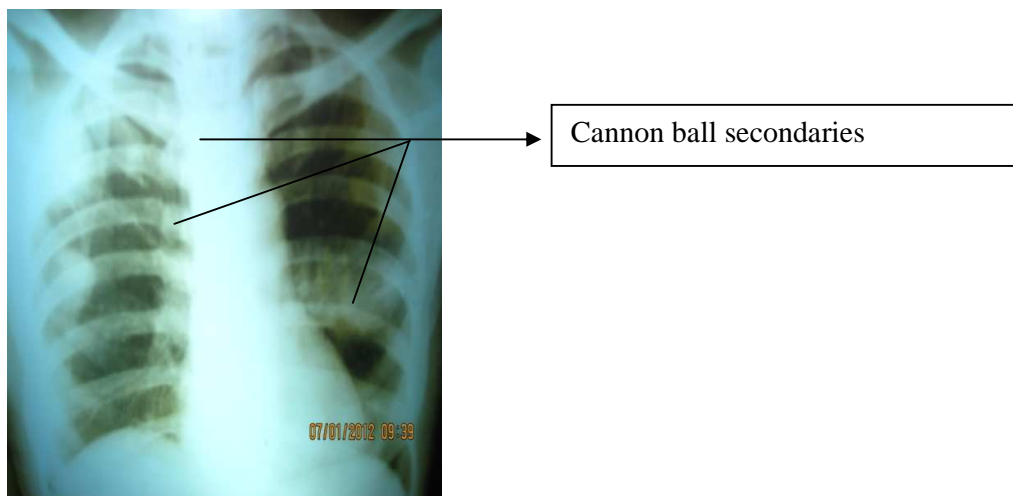


Fig.3.x-ray



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Fig.4a Genitalia-before treatment.



Fig.4a.externalTreatment



Fig.5a;6a. lesion - before treatment



Fig.5b;6b. lesion - after treatment



Fig .6 .a



Fig .6 .b

**DISCUSSION:** Tumors of vagina is generally said to have a very poor prognosis. TAUSSIG (1935) quoted 'primary cancer of vagina is rare and almost always universally fatal.<sup>3</sup> Most cancers occurring in the vagina arise by direct spread/metastases from cervix, body of the uterus, vulva, bladder, rectum or sigmoid colon. Primary cancer of vagina is rare and represents 1-4% of all

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genital malignancies. 93% are squamous cell type, 4 to 5% are adenocarcinoma<sup>3</sup> and 2% are sarcomas.<sup>4</sup>

Sarcomas are rare mesenchymal neoplasms that arise in soft tissues and bone. These tumors are usually of mesodermal origin, a few are derived from neuroectoderm. Sarcomas may be divided into two groups: those derived from soft tissues and those derived from bone. Soft tissues include muscles, tendons, fat, fibrous tissue, synovial tissue, vessels, and nerves. Approximately 20 different groups of sarcomas are recognized on the basis of the pattern of differentiation towards normal tissue.<sup>8</sup>

A variety of sarcomas have arisen in the vagina or para vaginally. Primary sarcomas of the vagina are Rhabdomyosarcoma, Leiomyosarcoma, Malignant fibrous histiocytoma, Haemangiopericytoma, Malignant schwannoma, endometrial stromal sarcoma, Fibrosarcoma, Alveolar soft part sarcoma, and Angiosarcoma<sup>5</sup>. Upto 1985 there were 68 cases of primary sarcomas reported.<sup>2</sup> Vaginal sarcomas should be distinguished among themselves as to their precise pathogenetic origin by using special stains, electron microscopy, and immunohistochemistry. Various immunohistochemical markers are used for confirmation of a given phenotype or differentiation or histogenesis.<sup>1, 2</sup> Sarcomas of the elderly are leiomyosarcoma, angiosarcoma, spindle cell sarcoma, alveolar soft part sarcoma, fibrosarcoma, neurofibrosarcoma, and mixed mesodermal tumors of the vagina. Sarcomas of the young are embryonal rhabdomyosarcoma [sarcoma botryoids] and endodermal sinus tumors that develops in young girls between 6 months and 16 years of age.<sup>4</sup> Neoplasms containing >5 mitosis/10HPF are locally aggressive and occasionally give rise to distant metastasis. Consequently they are malignant.<sup>2</sup>

**CLINICAL PRESENTATION:** Mostly asymptomatic. Others presenting as a lump near introitus which may cause problems with micturition, defecation or intercourse. Bleeding or discharges are late features.<sup>4</sup> Complete history and physical examination should be performed including speculum and per vaginal examination, cervical cytologic examination, endometrial biopsy when indicated, colposcopy and biopsy of the vaginal tumor. Pretreatment evaluation may include the following studies: chest x-ray, intravenous pyelogram, cystoscopy, proctosigmoidoscopy and CT, MRI scan of the abdomen and pelvis.<sup>10</sup>

**DIFFERENTIAL DIAGNOSIS:** When a patient presents with vaginal swelling the following possibilities are to be considered:

- vaginal cysts (mullerian cyst, Gartner's cyst, cyst from skene's tubules, implantation dermoids, epidermoid cyst)
- Benign vaginal neoplasms (papilloma, angioma, adenoma, fibroma, lipoma, myxoid soft tissue tumours, granuloma)
- Malignant neoplasms-may be primary or secondary
  - o Carcinomas-squamous cell carcinoma, adenocarcinoma, melanoma, embryonal carcinoma.
  - o Sarcomas- leiomyosarcoma, embryonal rhabdomyosarcoma.<sup>10</sup>

**TREATMENT:** The treatment plan depends on (1) the patient's age and general health state, (2) the tumor location and size, (3) the need to maintain the function of the vagina (4) and stage of the disease. Prognosis is related to the stage of the disease [American Joint Commission on Cancer staging system for sarcomas]<sup>8</sup>

The treatment options available are the following.

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## **SURGERY:**

- Wide local excision with reconstructive surgery
- Vaginectomy with lymphadenectomy
- Radical (total) hysterectomy
- Pelvic exenteration ( removal of vagina, rectum, colon, bladder, part of bowel)
- Laser surgery.<sup>9</sup>

## **RADIATION THERAPY:** types are

1. External beam radiation therapy
2. Internal radiation therapy or brachytherapy:
  - a. Intracavitary radiation
  - b. Interstitial radiation<sup>9</sup>

**PRIMARY CHEMOTHERAPY** with vincristine, actinomycin D, and cyclophosphamide plus radiation leads to excellent results in treating patients with this disease.<sup>10</sup>

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