

# Primary Ovarian Rhabdomyosarcoma: A Case Report

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**Key words:** *Rhabdomyosarcoma, ovary, child.*

## SUMMARY

**Background:** *Ovarian cancer is the second most common female genital tract cancer in Nigeria. The prognosis is usually poor due to late presentation.*

**Objective:** *The objective of this case report is to present a case of rhabdomyosarcoma of the ovaries a very rare type of malignant ovarian tumour in children.*

**Method:** *This is a case report of a thirteen year old girl who presented with abdominal swelling.*

*Ultrasound examination revealed ascitis and bilateral enlargement of both ovaries. The findings were confirmed at laparotomy. Bilateral ovariectomy was done. Histology report showed rhabdomyosarcoma of the ovaries. Post operatively the patient presented with metastatic deposits to the spine. She could not afford chemotherapy or radiotherapy and died ninety three days after presentation.*

**Conclusion:** *A rare ovarian malignancy has been reported. Treatment was inadequate largely because of poverty and delay in obtaining a histology report. A plea is made for increased training of, pathologists and oncologists and for free treatment of malignancies in the third world*

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## INTRODUCTION

Ovarian cancer is a serious disease which often presents late and is consequently associated with a low five year survival rate. It is more common in the wealthy developed countries although its incidence is rising in the third world<sup>1</sup>. Unlike in Nigeria where cancer of the cervix is the most common genital tract cancer, it is the most common genital tract cancer in the United Kingdom where it is responsible for more deaths than all other genital tract cancers<sup>2</sup>. It is a disease of the elderly with its incidence peaking at about the age of 67 years and is relatively uncommon in young women under the age of 35 years<sup>1</sup>. When it occurs in the young it is usually of germ cell origin rather than the epitheloid type seen in adults<sup>2</sup>. With regards to its aetiology environmental, genetic, hormonal and chemical factors like asbestos have been implicated<sup>3</sup>. Ovarian

cancer may be primary or secondary and the commonest variety seen is the serous-cystadenocarcinoma<sup>2</sup>. The ovary consists of totipotent cells and so has the capacity to produce virtually any type of cell in the body. This accounts for the bewildering variety of tumours seen in the ovary. One of such tumours which is very rare is primary ovarian rhabdomyosarcoma which is presented below.

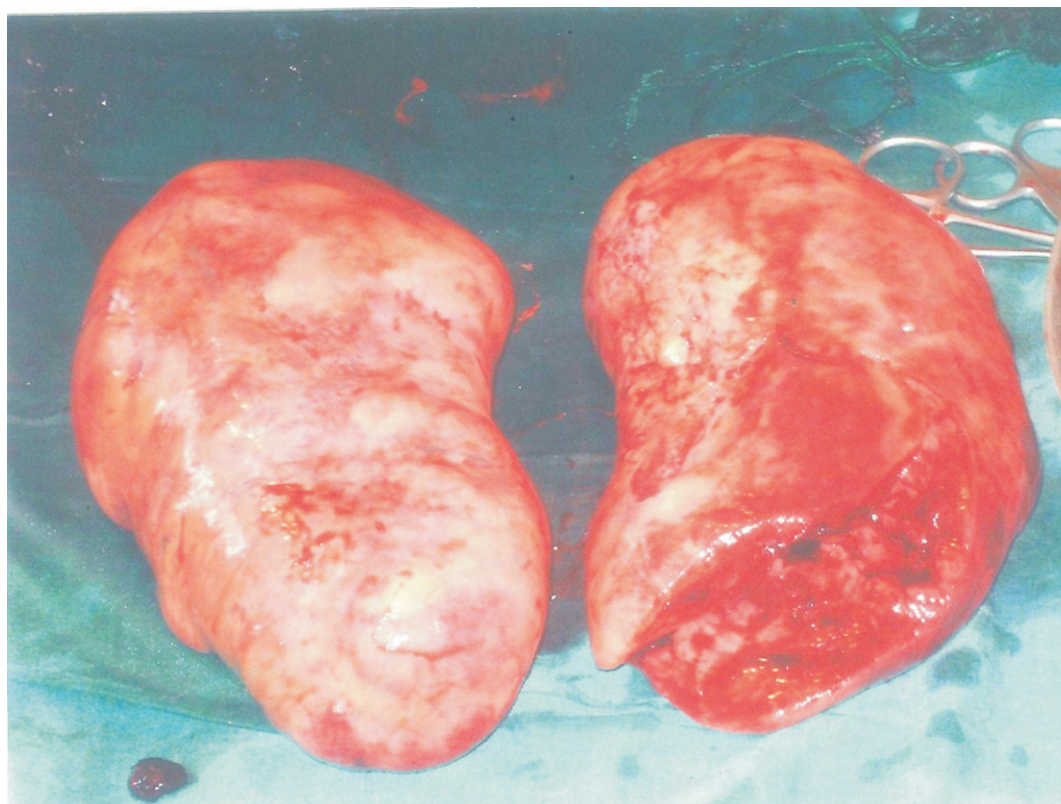
## CASE REPORT

A twelve year old girl presented with a three month history of progressive abdominal swelling fever and pain on the lower limbs associated with weight loss. On examination she weighed 39 kg, was febrile (temp 38.3°C), anicteric and had no peripheral lymphadenopathy. Her blood pressure was 130/90mmHg, pulse rate 74 beats per minute and her heart sounds were normal. The abdomen was distended and tense with an abdominal circumference of 65cm around the umbilicus. Vague masses one on either side were palpable in the right and left lumbar regions of the abdomen, and massive ascitis was demonstrable. The liver, spleen and kidneys were not palpable. A suspicion of ovarian carcinoma was made. Results of investigations were haemoglobin 11.6 gm/dl, packed cell volume 34%, malaria parasite +, urinalysis normal, urea and electrolytes normal. Chest X-Ray was also normal. Abdomino-pelvic ultrasound scan showed bilateral solid abdominal masses, ascitis and right hydronephrosis. The fever responded to antimalarial treatment. At laparotomy, the following findings were made: (i) About three litres of non haemorrhagic ascitis (ii) Enlargement of both ovaries which were friable and bean shaped (Fig 1). The right ovary measured 21x10cm while the left measured 22x10cm. (iii) Normal uterus and fallopian tubes. (iv) No deposits were seen in the peritoneum liver or other abdominal organs. Bilateral ovariectomy was done. Estimated blood loss was 500ml. Post operatively, she was transfused with one unit of blood. She was stable and was discharged on the seventh post operative day, to await the histology report. Four weeks later, she returned to the clinic complaining of fever, difficulty in locomotion and pain in the waist. Her haemoglobin had fallen to 5.8 gm per dl, white blood cell count 4,800 per deciliter and X-ray showed metastatic deposits to the lumbar spine. The patient could not afford chemotherapy or radiotherapy. She subsequently deteriorated and died ninety three days after presentation. The histology report (Fig. 2) which was received post mortem read as follows: the tumour is composed of medium sized cells with large vesicular nuclei and scanty pink cytoplasm; these cells are arranged in loose clusters separated by thin fibrovascular septa; giant tumour cells are seen scattered within the tumour. Overall features are those of a malignant small round cell tumour consistent with alveolar rhabdomyosarcoma.

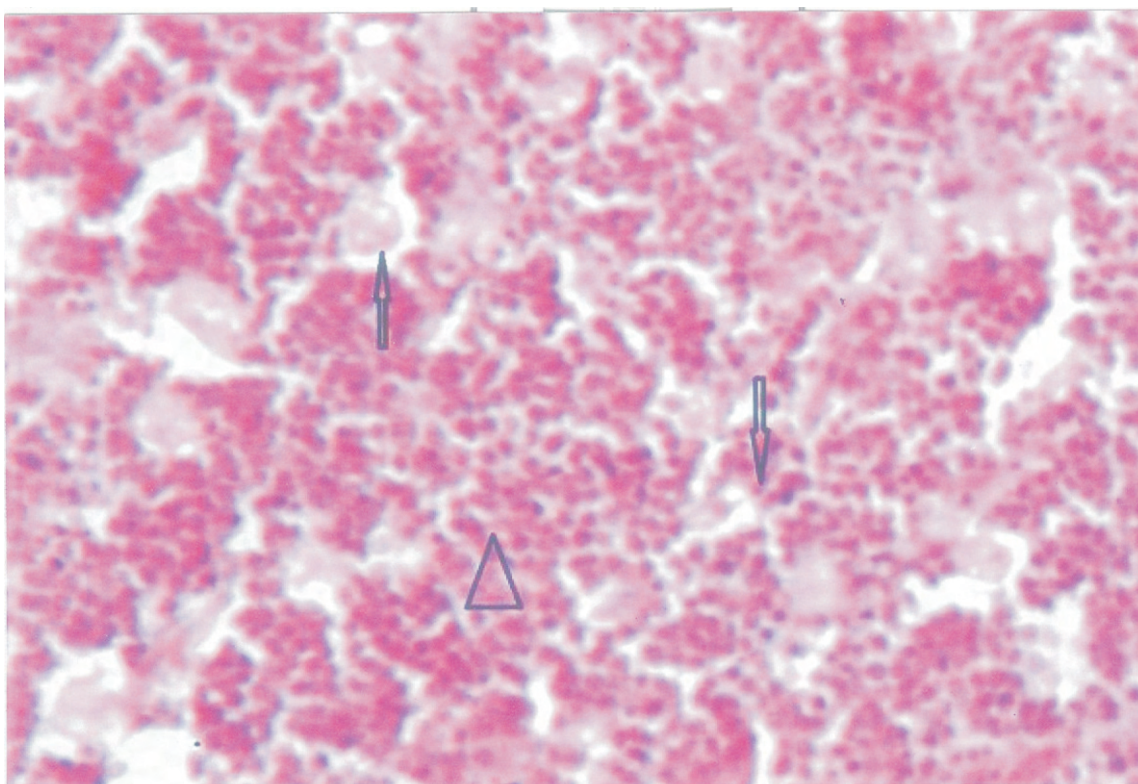
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**Figure 1 Enlarged Right and Left Ovaries 20 x 18 5.5 cm**



**Figure 2 Magnification x 100 (H and E) Arrow Head shows clusters of small round blue cells Upward arrow shows a tumour giant cell. Downward arrow shows a fine septum on which a row of tumour cells are attached.**

## DISCUSSION

Generally, malignant neoplasms of the ovary account for a small percentage of all malignancies. Figures ranging from 2%, 4.3%, and 12.5% have been reported from Ghana, Pakistan and Ibadan respectively<sup>4-6</sup>. In children however rhabdomyosarcoma with a prevalence of 16.4% was the second most common paediatric tumour after lymphoma<sup>7</sup>. Although it is the most common soft tissue sarcoma found in children with common sites of origin being the head and neck, genito urinary system and the extremities. Its occurrence in the ovaries is very rare<sup>8</sup> and by 2008 only ten paediatric cases had been documented in the literature<sup>9</sup>. In the two series of paediatric rhabdomyosarcoma from Nigeria,<sup>10,11</sup> over a period of nineteen and seven years respectively no case of ovarian rhabdomyosarcoma was reported. Four major types, the alveolar, embryonal, butyroid and pleomorphic have been described. This case was alveolar and is consistent with the finding from Jos<sup>11</sup> but different from the finding in Ibadan<sup>10</sup> where the embryonal type was most prevalent. Because of its rarity the commonest age of presentation remains undetermined. The age of occurrence ranged between 13 months and 86 years in one study<sup>12</sup> with 60% of the women aged 40 or more years while another study<sup>13</sup> reported an age range of 7-79 with a mean age of 37 years. When they arise in the ovary they are thought to arise from uncommitted stromal fibroblasts present in the ovary. The presentation depends on the site of origin but when as in this case it is ovarian it presents classically as any other ovarian malignancy. The presentation of this patient was late. This is a common experience with ovarian malignancies in our institution.

Classically, the cells are small with granular eosinophilic cytoplasm (Fig. 2) and rich in thick and thin filaments. Definitive diagnosis was based on the demonstration of skeletal muscle differentiation, the demonstration of z bands under electron microscopy, or the use of immuno-histochemistry to demonstrate muscle specific antigens such as desmin or actin. The absence of these facilities was a limitation in this case as diagnosis was by light microscopy only. The tumour may be confused with leukaemia small cell sarcoma, malignant lymphoma and neuroblastoma<sup>14,15</sup>. Although formerly invariably fatal, a correct diagnosis of this tumour is important as there have been recent successes in its treatment with the use of a multidisciplinary approach, complete removal where possible, multiagent chemotherapy and radiotherapy where appropriate<sup>8-12</sup>. An overall survival rate of around 20% has been quoted<sup>8,11</sup>. The survival of our patient for

only ninety three days is consistent with findings from other studies. Survival is said to be worse in those older than ten years and who present late<sup>11</sup>. This was the case here. In our environment, many factors militate against favourable outcome in the treatment of this and other cancers. These factors range from late presentation to inability to afford the cost of chemotherapy and radiotherapy<sup>11</sup> as was the case here. Even when the funds are available the radiotherapy centres are few and patients have to travel very long distances to access them. There is also a dearth of oncologists and pathologists and often no facilities for frozen section. Prioritization of requests for histology by pathology laboratories to give preference to cases of suspected malignancy is not done. This results in long delays in getting histological diagnoses in such critical cases. In this patient for instance, the histology report was received post mortem, about two and a half months after it was sent for histology. Fortunately the tumour is rare, but for those that occur in the third world, prognosis is likely to remain high for a long time.

In conclusion a plea is made for the training of more oncologists and pathologists and for free chemotherapy and radiotherapy services in developing countries. More timely histology reports of suspected cancer cases by pathologists would enable doctors start treatment earlier.

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