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Uterine Leiomyosarcoma: A Case Report and Review of Relevant Literatures

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Authors' contributions

This work was carried out in collaboration among all authors. All authors read and approved the final manuscript.

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Case Report

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ABSTRACT

Leiomyomas are benign soft tissue neoplasms that arise from smooth muscles. Leiomyosarcoma is a malignant lesion of smooth muscle origin. This is a rare lesion in the uterus with 1-2 % of its benign counterpart, leiomyoma, transforming into the lesion. It is often aggressive and could develop at any site where the smooth muscle is found. We present a case of a 48-year-old farmer with pelvic mass with Ultra-sonography(USS) suggestive of leiomyoma. Hysterectomy was done and histologic diagnosis of leiomyosarcoma (LMS) was made. The patient was discharged 7 days post operation but was lost to follow up. Uterine LMS is an aggressive tumour, therefore, a high index of suspicion is needed especially for huge uterine nodules and such patients must be closely monitored for adequate management.

Keywords: Uterus; leiomyosarcoma; smooth muscle.

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1. BACKGROUND

Leiomyomas are benign soft tissue neoplasms that arise from smooth muscles. Leiomyosarcoma(LMS) is an aggressive malignancy of smooth muscle origin. It accounts for 5-10% of sarcoma derived from soft tissues [1]. The soft tissues commonly affected include the uterus, stomach, intestines, wall of blood vessels, skin, and peritoneum. LMS of the bone has also been reported [2]. LMS in the uterus should be suspected if an apparent leiomyoma is soft, shows areas of necrosis on gross examination, has irregular borders (invasion into neighbouring myometrium), or does not bulge above the surface when cut [3]. Microscopically, typical LMS is hypercellular, with nuclear atypia and pleomorphism, mitotically active (with some of the mitoses being atypical), and with areas of necrosis. Histological variants of LMS are epithelioid (clear cell) LMS (malignant leiomyoblastoma), myxoid LMS, LMS with osteoclast-like giant cells, intravenous leiomyosarcomatosis and LMS with skeletal muscle differentiation.

Ultrastructurally and immunohistochemically, the features of LMS are those of smooth muscle cells [2]. Patients with primary intra-abdominal tumours had better outcomes than those with recurrent intraperitoneal tumours. Whites had a more favourable prognosis. In patients with intraabdominal tumours, only mitotic count >10M/10HPF portended a poorer prognosis. Patients with pulmonary metastasis had improved outcomes with "curative" metastasectomy. Uterine LMS samples exhibited loss of ER and PR expression overexpressed Ki-67, and altered p53, Rb, p16, cytoplasmic β-catenin, EGFR, PDGFR-α, PDGFR-β, and AXL levels. Metastatic tumours had increased VEGF, Ki-67, and survivin expression versus localised disease. Survivin and β-catenin expression were associated with intraperitoneal recurrence; high bcl-2 expression predicted longer DSS [4]. LMS occur in an older age group than leiomyomas (median age, 54 years); patients are usually in their fifth and sixth decade of life; although they can also occur in younger patients [1,2]. The incidence is higher in female than male (2:1). This may be related to the ability of smooth muscle cells to proliferate in response to oestrogen [3]. The incidence is only 1/1000 that of its benign counterpart (leiomyoma). It accounts for 2% of uterine malignancies [5, 6].

The aetiology is unknown. Its pathogenesis is uncertain, but at least some appear to arise from within leiomyomas. Women with LMS are on average more than a decade (age above 50) than those with leiomyomas and the malignant tumours are larger (10-15cm versus 3-5cm). The symptoms of uterine LMS include pelvic mass, menstrual irregularity, symptoms of metastasis in advanced cases. Most leiomyosarcomas are large and are advanced when detected. They thus are usually fatal despite combinations of surgery, radiation therapy, and chemotherapy and response to these agents are generally poor. The surgery is a hysterectomy. Nearly half of recurrences first present in the lung, and 5-year survival is about 20%. Uterine LMS usually spreads within the pelvis and in the form of distant metastases to the lung, bone, and other sites. Lymph node metastases are exceptional. The standard treatment is total abdominal hysterectomy with bilateral salpingo-oophorectomy. Prognostic factors include the following: Tumor stage, Tumor size, Microscopic grade and DNA ploidy.

2. CASE REPORT

We present a 48-year-old Para 5 $^{+2}$ who was referred from a General Hospital in the outskirt of the state on account of lower abdominal mass and pain of over 6months duration. On presentation in our institution, further examination and investigation revealed a 26th-week palpable pelvic mass. Ultra-sonography (USS) suggested of huge uterine nodule with degenerative changes. Blood parameter showed anaemia while electrolytes revealed metabolic acidosis. The metabolic derangement and the anaemia were corrected and the patient subsequently prepared for surgery. A uterine nodule weighing 5.2kg and measuring (28x24x18) cm was seen. The mass was poorly encapsulated and the cut surface showed a greyish white soft tissue with areas of gelatinous material, cystic changes, haemorrhage and necrosis. The histology is that of proliferating mesenchymal tumour with marked pleomorphism and hyperchromasia. There is nuclear atypia with abnormal mitotic figures. Also seen are areas of haemorrhage and necrosis. The histology report came out after the patient was discharged from the hospital; about a week after the operation. The patient was to be seen in the clinic after 4 weeks but was, however, lost to follow up.

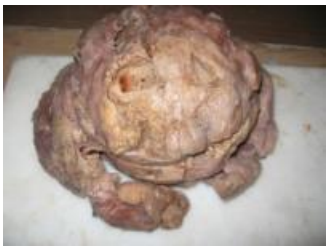

Fig. 1. Figure showing the external surface of a meaty brownish non encapsulated soft tissue mass

Fig. 2. Figure showing the cut surface of the tissue

The gross appearance of the lesion showing irregularly shaped meaty tissue with lack of capsule and the cut surface shows areas of haemorrhage and necrosis.

Fig. 3. Histologic section of the lesion shows a highly cellular tissue with proliferating spindle-shaped cells (H&E x 40)

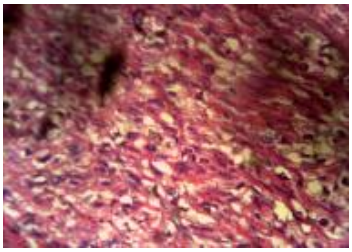

Fig. 4. Histologic section of the lesion showing pleomorphism, hyperchromatic and coarse chromatin pattern with numerous abnormal mitotic figures (H&E x100)

3. DISCUSSION

Studies done by Adesiyan in Kano; Ogunbiyi in Ibadan and Fubara in Port Harcourt, Nigeria and other parts of Africa [7,8,9] show that LMS is a rare tumour; this agrees with this study. Uterine LMS is often seen between the fourth and the fifth decade of life; however, a case of 14-year-old, thin, African female, gravida 0, with abdominal discomfort and increased abdominal circumference and ruptured uterus secondary to LMS was once seen in Istanbul, Turkey [10]. Our index case is that of a 48-year-old female with abdominal swelling and pain, which are the usual presentations of the lesion as seen in most studies. However, there was a case of a 63-year Para 10 woman with vaginal bleeding reported in Lagos, Nigeria [11].

The USS done for our case was suggestive of huge leiomyoma with degenerative changes; this is the usual presentation seen. However, according to Seki et al. increased serum level of lactate dehydrogenase levels in a patient with degenerative changes in leiomyoma is diagnostic of leiomyosarcoma [13].LMS is often in corpus uterine, however, they could be found rarely in the uterine cervix as presented by Varsha in South Africa while a vulva LMS was also seen in Nigeria [14,15]. Adesiyan in Zaria, Nigeria further stated that LMS could present as for the cause of subfertility in women [7], therefore a high index of suspicion is key in early diagnosis and proper management of the disease condition.

4. CONCLUSION

Uterine leiomyosarcoma is a rare disease condition with high morbidity and mortality rate. An increased index of suspicion is necessary for any huge uterine nodule that is un-encapsulated and shows areas of haemorrhage and necrosis. Further ancillary investigation for histogenesis of the disease condition is necessary.

CONSENT AND ETHICAL APPROVAL

As per university standard guideline participant consent and ethical approval has been collected and preserved by the authors.

COMPETING INTERESTS

Authors have declared that no competing interests exist.

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