

# Leiomyosarcoma of the vagina in pregnancy

B. Bassaw<sup>1</sup>, H. Fletcher<sup>2</sup>, J. Chinnia<sup>1</sup>

<sup>1</sup>Department of Obstetrics and Gynaecology, The University of the West Indies, Mt. Hope Maternity Hospital, Champs Fleurs, Trinidad

<sup>2</sup>Department of Obstetrics and Gynaecology, The University of the West Indies, Mona Campus, Kingston, Jamaica (West Indies)

## Summary

Primary vaginal leiomyosarcoma in pregnancy is an extremely rare disease which continues to have a poor prognosis. This is due to the late diagnosis as well as the treatment is based on limited experience based on case reports and not randomised trials. The authors report the first case of leiomyosarcoma of the vagina in a pregnancy in a 31-year-old Afro-Caribbean multigravida at the Mt. Hope Maternity Hospital. Despite the administration of systemic chemotherapy and irradiation, the patient succumbed to her illness 11 months after the initial diagnosis. If the prognosis is to be improved, it is prudent that healthcare providers must be more aware and knowledgeable of this tumour and be proactive in its management.

*Key words:* Leiomyosarcoma; Vagina; Pregnancy.

## Introduction

Vaginal neoplasms are uncommon accounting for less than 1% of female genital malignancies [1]. The majority of these vaginal tumours are squamous cell cancers. Rarely, one may encounter a vaginal sarcoma in an adult. It is estimated that primary sarcoma represents about only 2% of all vaginal malignancies. In adults, the most likely form of sarcoma is leiomyosarcoma whereas about 90% of rhabdomyosarcoma (sarcoma botyroides) are seen in children less than five-years-old.

Most cases of leiomyosarcoma occur in the uterus either de novo or less commonly as a result of malignant transformation of a leiomyoma. In common with other sarcomas, leiomyosarcomas display variability in size and shape of the cells and varying degrees of anaplasia in the nuclei. The chromatin is coarse and clumped and mitoses are often numerous and distinctly atypical. The site of origin of vaginal leiomyosarcoma is usually the smooth muscle of the vaginal wall but it may arise in smooth muscle cells in adjacent structures. Unlike other gynaecological malignancies in which the prognosis depends on the staging, leiomyosarcoma, which is confined to the vaginal wall, still carries a poor outcome [2].

Vaginal malignancies are even less common in pregnancy. Leiomyosarcoma which is rarely found in pregnancy is usually detected in the uterus or the vulva [3]. From the present authors' literature search, they report the first case of a vaginal leiomyosarcoma in pregnancy in an Afro-Caribbean woman.

## Case Report

A 31-year-old Afro-Caribbean woman of gravidity 6, parity 4+1 presented at her 30-week antenatal appointment with a one-month history of a growth in the vagina which was increasing in size. There was no associated pelvic and abdominal pain nor any abnormal vaginal bleeding, urinary or bowel complaints. She had clinical diabetes mellitus which was well-controlled with a diabetic diet and soluble insulin. There was no past history of any sexually transmitted diseases.

On pelvic examination, two one-two-cm, non-tender wart-like growths were identified on the right posterior aspect of the lower third of the vagina. There was a thick cottage-cheese vaginal discharge consistent with vaginal candidiasis. The patient subsequently had a spontaneous vaginal delivery. She was discharged one day later with a six-week appointment for the postnatal clinic for further evaluation. However, she returned one week later complaining of pelvic discomfort and a burning sensation in the vagina. The growth had increased in size to four to five cm in diameter. The consistency was firm and the base appeared necrotic. Under general anaesthesia, the tumour which was pedunculated was excised at the base of the stalk. Haemostatic sutures were inserted. On gross examination, two irregular nodular pieces of tissue measuring 1.5 x 2.0 x 3.0 cm were submitted for histological analysis which revealed that the neoplasm contained fascicles of spindle-shaped cells with fusiform, hyperchromatic, and pleomorphic nuclei with frequent abnormal mitoses (greater than ten per ten high-power fields). The surgical margins were minimally breached at two sites. The diagnosis was poorly-differentiated leiomyosarcoma of the vagina. The patient refused any further surgical intervention.

Extensive evaluation with chest X-ray, ultrasound of the liver, and CT scan with intravenous contrast of the abdomen and pelvis were performed. The findings included a mildly enlarged liver without any focal parenchymal lesions, normal gallbladder, spleen, adrenals and kidneys, no evidence of ascites or para-aortic lymphadenopathy, a mildly bulky uterus, normal adnexae, and bilateral

Revised manuscript accepted for publication May 5, 2012

inguinal lymphadenopathy with cystic degeneration of a right groin node which measured 2.4 cm in diameter. No focal lesions were seen in the lumbar and pelvic spine and the lungs were normal. After review by the gynaecological oncologist, she was referred for further adjunctive treatment.

Initially, she received a combination of vincristine, adriamycin, and cyclophosphamide which was followed by gemcitabine and cisplatin. When a firm immobile irregular mass involving the posterior and right lateral aspects of the vagina with parametrial involvement was noted, a total of 3,000 cGy of radiotherapy, given in increments of 300 cGy per session was administered to the pelvis. This was followed by a similar dose to the brain for metastatic disease. She succumbed to the illness 11 months after the initial diagnosis.

## Discussion

The authors' literature search has revealed that this is the first reported case of vaginal leiomyosarcoma in pregnancy in an Afro-Caribbean woman. Behzatoglu *et al.* reported on the first possible case of leiomyosarcoma in a 21-year-old primigravida at 39 weeks' gestation [3]. Primary vaginal malignancies are rare accounting for only approximately 2% of all female genital neoplasms. The majority of these are squamous cell carcinoma [4]. Primary sarcoma represents only about 2% of all vaginal malignancies, and leiomyosarcoma is the most frequent histological type in adults while rhabdomyosarcoma is more common in children. Ahram *et al.* identified only 138 cases of vaginal leiomyosarcoma in a thorough literature search [2]. Even rarer is this type of sarcoma in pregnancy with only a handful of cases reported thus far. This is the first case identified at the Mt. Hope Maternity Hospital in over 150,000 deliveries.

Our patient presented at 30-weeks' gestation with a small swelling in the vagina which was assumed to be a benign wart and as a result the treatment was scheduled for after the delivery. The aggressiveness of this tumour was depicted by the rapid increase in size, as well as the presence of pain most likely from deep invasion in the sub-vaginal tissue in less than two months. A predilection for this tumour to occupy the posterior and lateral aspects of the vagina was also evident in the present case. Until the aetiology and pathogenesis of this tumour are fully elucidated, it is not possible to give a clear explanation for this occurrence.

Although at the time of the local excision the authors were aware that the swelling was likely to be malignant, they did not suspect a leiomyosarcoma. As a result, their plan was to excise the mass and to await the histological evaluation before making a definitive plan. The diagnosis confirmed by histology was a poorly-differentiated leiomyosarcoma with involvement of the margins. The patient refused any further surgical options instead she opted for adjuvant therapy.

Early diagnosis of leiomyosarcoma with the disease at Stage 1 is critical if simple excision is likely to yield good results [2]. In the present case there was a breach of the margin which may be indicative of the aggressiveness of the tumour. Tavassoli and Norris recommended extensive surgi-

cal excision of the lesions which are likely to recur [5]. Some of the risk factors for early recurrence include a tumour size of three cm or more in diameter, irregular contour, cellular atypia, and five or more mitoses per ten high-power fields. In the present patient all of these features were evident.

The lack of responsiveness of poorly-differentiated advanced disease is demonstrated in the present case who failed to respond to vincristine, adriamycin and cyclophosphamide, and also, a combination of gemcitabine and cisplatin. The authors added both pelvic irradiation targeting the tumour and the involved groin nodes as well as external irradiation to the brain for cerebral metastases. Hensley advocated the need for primary surgery plus adjuvant irradiation to decrease local recurrences, and chemotherapy for persistent and recurrent disease [6]. The present patient experienced a fairly rapid downhill course despite several chemotherapeutic agents and irradiation which concurs with the findings of Ngan *et al.* who found that neither chemotherapy nor radiotherapy was particularly useful in late or recurrent disease [7]. It is unknown whether leiomyosarcoma behaves in a more aggressive manner in pregnancy and if so, what factors may be responsible. The present case highlights the fact that leiomyosarcoma of the vagina remains a difficult diagnosis to be made clinically, and it continues to carry a poor prognosis despite the advent of many newer and more effective chemotherapy. It should be reported due to its rareness in order for us to compile sufficient evidence on the most appropriate treatment. Furthermore, a greater sense of awareness is required by healthcare workers if we are to diagnose this highly malignant tumour early especially in pregnancy.

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Address reprint requests to:  
B. BASSAW M.D.  
12 Realspring Avenue  
Valsayn, Trinidad (West Indies)  
e-mail: obsgynfms@gmail.com