Endocervical Stromal Sarcoma of the Cervix – a Case Report

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Abstract

A 50-year old perimenopausal woman with clinical symptoms suggestive of invasive carcinoma of the cervix was referred to our Gynaecological Clinic. Chest X-rays, blood film appearance, mid-stream urine and intravenous urography were essentially normal. Histological examination revealed that the lesion was an endometrial stromal sarcoma, an extremely rare form of sarcoma in the uterine cervix. Features included stellate and plump spindle-shaped cells with scanty cytoplasm and abnormal mitoses of 3-4 per high power field. Histopathologists are reminded to keep the lesion in mind when examining cervical biopsies or hysterectomy specimens so as not to miss the diagnosis.

Key Words: Cervix, Endocervix, Stromal Sarcoma, Malignancy [Trop J Obstet Gynaecol, 2003, 20: 167-169]

Introduction

Sarcomas of the uterus are very rare, occurring in approximately 2 in 100,000 women over the age of 20 years [1]. Rarer still is sarcoma of the uterine cervix, which could present in various forms such as leiomyosarcoma, embryonal rhabdomyosarcoma (botryoid type), alveolar soft-part sarcoma, osteosarcoma and endocervical stromal sarcoma [2]. The last decade has witnessed reports of isolated cases of cervical sarcomas such as primary endocervical extra-osseous Ewing's sarcoma [3], malignant cervical schwannoma and mullerian adenosarcoma of the cervix [4]. The clinical presentation of these tumours may mimic that of invasive carcinoma of the cervix.

In a recent review of the histopathological characteristics of tumours of the uterine cervix at Ile-Ife, Nigeria over a ten-year period [5], sarcoma of the cervix accounted for just 1% of the malignant tumours. These were mainly leiomyosarcoma, which is the most common malignant mesenchymal tumour of the cervix. Although leiomyosarcomas of the cervix are rare, rarer still is endocervical stromal sarcoma. Our literature search did not reveal the report of this tumour in Nigeria and indeed in Africa continent and hence the documentation of this case report as it presented to us at Ile-Ife, Nigeria.

Case Report

Mrs F.F. was a 50 year-old postmenopausal woman. She was multiparous and her last confinement was 15 years prior to presentation. She had not engaged in coitus since her last childbirth. Her last menstrual period was 5 years prior to the onset of symptoms. She presented with a one-month history of unprovoked irregular bright red vaginal bleeding, associated with passage of blood clots. There was no history of multiple sexual partners or previous history of post-coital bleeding. She also had a foul-smelling watery vaginal discharge, which started shortly after the onset of vaginal bleeding. Patient also had weight loss, generalised body weakness and dizziness. There was no history of pelvic pain, chronic cough or hospitalisation except those associated with chilbirth.

Physical examination revealed a middle-aged woman with moderate pallor. The cardio-respiratory systems were normal. The significant findings were confined to the genital tract, which revealed a blood-smeared vulva, normal vaginal wall and an irregular fungating soft mass protruding from the external os of the cervix into the vagina. The mass bled on contact but the surrounding ectocervix looked grossly normal. The uterus was bulky and consistent in size with that of a 10-12 week gravid uterus. Rectal examination showed no abnormalities. An initial clinical diagnosis of infected fibroid polyp was made.

The patient had examination under anaesthesia (EUA), avulsion of the polypoid mass and fractional curretage of the uterus. The results of chest X-rays, full blood count, mid stream urine were essentially normal while a high vaginal swab for microscopy, culture and sensitivity revealed a growth of *Staphylococcus aureus* sensitive to ampicillin. She had standard therapeutic doses of ampicillin and metronidazole for ten days after the EUA. Despite these steps, the irregular vaginal bleeding persisted.

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The patient eventually had a total abdominal hysterectomy, bilateral salpingo-oophorectomy and removal of the upper third of the vagina six weeks after the initial EUA. She went home ten days after the operation. She reported a month later with fresh vaginal bleeding of one day's duration and vaginal examination revealed a soft fleshy new growth occupying the vaginal vault. She was counselled to have radiotherapy, which she declined for financial reasons. She died two months later from anaemia and the effects of the malignancy. Autopsy was not done because the patient died at home.

Pathological Examination

The initial specimen received from the patient was that of the avulsed cervical polyp and multiple fragments of dark-brown soft tissue aggregating to 1 cm in span. Microscopic examination of the polypoid mass showed tissue composed of endometrial-like stromal cells with round-to-oval hyperchromatic nuclei and amphophilic cytoplasms. The cells had indistinct cell borders and mitoses were infrequent. A diagnosis of low-grade stromal sarcoma was made. The endometrial currettings were normal, showing no evidence of malignancy.

The hysterectomy specimen showed a patulous cervix with a haemorrhagic nodule measuring 2 cm in diameter at the left lateral edge of the external os. Also protruding from the external os and attached to the endocervix by a stalk (Figure 1) was a dark-brown irregular tumour mass which measured 2.5 x 2 cm. The endometrial cavity was not remarkable. The myometrium showed a small greyish-white nodule about 1 cm in diameter. There was no attached palpably enlarged lymph node.

Microscopic examination of the endocervical mass showed diffuse sheets of proliferating stellate and plump-shaped cells which are arranged haphazardly (Figure 2). Other areas showed endometrial-like round to oval-shaped cells with hyperchromatic nuclei and moderate amphophilic cytoplasms. Abnormal mitoses 3-4 per high power field were seen (Figure 3). The tumour was well vascularised. Based on the above findings, a diagnosis of endocervical stromal sarcoma was made. The greyish-white nodule found in the myometrium showed features of leiomyoma.

Discussion

Endocervical stromal sarcoma is a very rare tumour. Clinical experience with sarcoma of the cervix is very limited. This is supported by the paucity of literature on the subject matter. The earliest available report on endocervical stromal sarcoma was by Abel and Ramirez [6] who, in 1973, reported twelve cases of endocervical stromal sarcoma. All patients except one were postmenopausal women who had vaginal bleeding and a polypoid mass occupying the cervix [6].

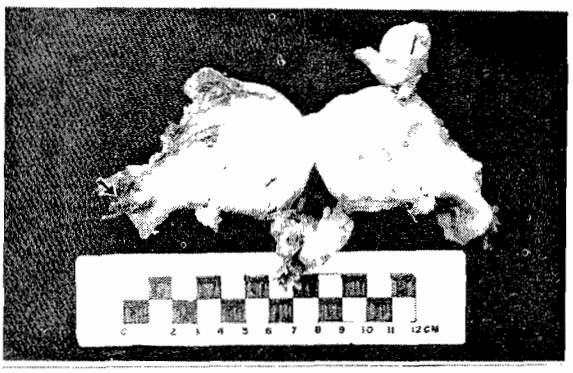

Figure 1
Transverse section of the uterus. The cervix is patulous and shows a dark-brown irregular mass protruding from the endocervix into the cervical canal (arrow)

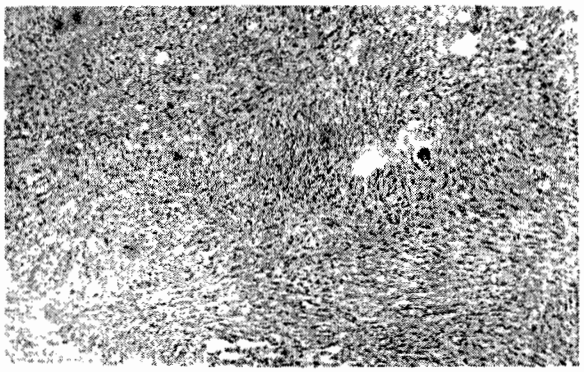

Figure 2
Photomicrograph of the cervical mass showing diffuse sheets of proliferating stellate and spindle-shaped cells arranged haphazardly [H&E, x 64]

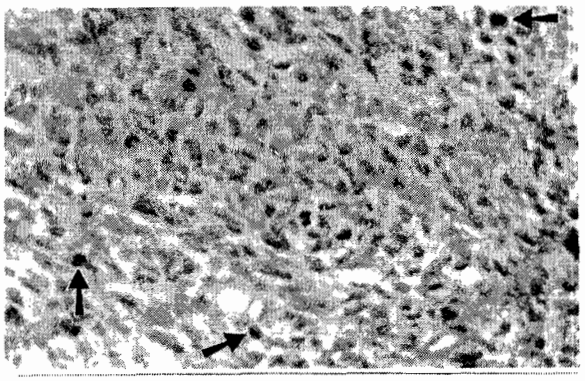

Figure 3
(Higher Magnification) Numerous proliferating oval and spindle-shaped cells are present with numerous abnormal mitoses (arrows) [H&E, x 576]

The mode of presentation of our patient is similar to the cases reported by Abel and Ramirez. Our patient was a postmenopausal woman with spontaneous vaginal bleeding and a polypoid mass in the cervix, mimicking carcinoma of the cervix, which is the most common genital malignancy in females worldwide [8]. Other tumours with similar clinical presentation include embryonal rhabdomyosarcoma, which is commonly seen in infants and young children [7], granulocytic sarcoma of the cervix, primary malignant cervical schwannoma [4], as well as primary endocervical extra-osseous Ewings sarcoma [3].

Embryonal rhabdomyosarcoma is a childhood sarcoma which usually presents as a bulky, grape-like masses protruding through the introitus. The microscopic features is classic and composed of small round- to- oval mesenchymal cells crowding beneath the epithelial surface of the vaginal or cervix (cambium layer) [9]. Granulocytic sarcoma is an unusual variant of myeloid malignancy in which there is an extramedullary tumour mass composed of myeloblasts, immature eosinophils and mature neutrophils [9]. Ewing's sarcoma is composed of sheets of small round cells with clear cytoplasm, which are Periodic-Acid-Schiff (PAS) positive [10]. Immunohistochemistry is also of value in clearly identifying these lesions, especially for the purpose of differentiating extra-osseuos Ewing's sarcoma from endometrial stromal sarcoma. Endocervical stromal sarcoma is certainly a diagnosis to be kept in mind by the histopathologist when examining surgical biopsies from the cervix or hysterectomy specimens.

References

1. Quinn MA, Anderson MC, Coulter CAE. Malignant diseases of the uterus. In: Shaw RW, Soutter WP, Stanton SL (eds); *Gynaecology.* Edinburgh, Churchill Livingstone, 1992; 533-546.
2. Wright TC, Ferenczy A, Kurman RJ. Carcinoma and other tumours of the cervix. In: *Blausteins Pathology of the Female Genital Tract 4th Edition.* New York, Springer-Verlaag, 1994; 315.
3. Cenacchi G, Pasquinelli G, Montanoro L, Cerasoh S. Primary endocervical extraosseous Ewing's sarcoma. *Int J Pathol,* 1998; 17: 83-88.
4. Lallas TA, Mehaffey PC, Lager DJ. Malignant cervical schwannoma: an unusual pelvic tumour. *Gynaecol Oncol,* 1999; 72: 238-242.
5. Somotun GO. *Histopathological review of diseases of uterine cervix in Ile-Ife, Nigeria.* A dissertation submitted to the National Postgraduate Medical College of Nigeria for the award of the Fellowship in Pathology. November 2000.
6. Abell MR, Ramirez JAG. Sarcoma and carcino-sarcomas of the uterine cervix. *Cancer* 1973, 31: 1176.
7. Zeisier H, Mayerhofer K, Fonra BA, Bancher-Todescao T. Embryonal rhabdomyosarcoma of the uterine cervix: case report and review. *Gynecol Oncol,* 1998, 69: 78-83.
8. Crum CP. Female genital tract: In: Cotran RS Kumar V. Collins T. (eds) *Robbins Pathologic Basis of Disease 6th Edition.* Philadephia, WB Saunders, 1999; 1049-1050.
9. Rosai J. *Ackermans Surgical Pathology 8th Edition.* New York, Mosby, 1996; 2080-2081.
10. Rosenberg A. Soft tissue tumours. In: Cotran RS Kumar V. Collins T. (eds) *Robbins Pathologic Basis of Disease 6th Edition.* Philadephia, WB Saunders, 1999; 1244.