Primary sarcomas of the corpus uteri: A clinico-pathologic and literature review of 13 cases seen in 5 years in Yaounde, Cameroon

Enow-orock GE¹, Nsagha DS², Assob NJ², Doh AS², Muna WFT²

¹Pathology Service General Hospital Yaounde, Cameroon.
²National Cancer Control Program, Yaounde, Cameroon.

Corresponding author E-mail: enowrock24@yahoo.com

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Cancer is a public health problem in Cameroon. 13 cases of primary sarcomas of the uterine corpus were diagnosed at the General Hospital Yaounde, in this 5 years retrospective study, giving an annual incidence of 2.6. The tumours included 5 leiomyosarcomas (38.5%), 6 endometrial stromal sarcomas (46.1%), 1 undifferentiated endometrial sarcoma (7.7%) and 1 carcinosarcoma of the endometrium (7.7%). There were 46.2% of the tumours classified as low-grade against 15.4% high grade. The age of patients ranged between 27 to 75 years, at an average of 45.25 (±2) years. Patients were predominantly in the 30-49 years age group (61.5%). The average age of patients was 38.8 years for leiomyosarcoma, 63 for endometrial carcinosarcoma, 48 for undifferentiated endometrial sarcoma, and 39.7 for endometrial stromal sarcoma. Majority of the patients (75%) were postmenopausal. All of the tumours were early stage diseases (I and II). The patients presented mainly with classical signs of pain, uterine mass and bleeding. A clinical diagnosis of uterine leiomyoma was made in 81.8%. All the patients were discharged alive from hospital after initial treatment. Though sarcomas of the uterine corpus are rare, this diagnosis should be ruled out in all apparently benign tumours of the uterine corpus.

Key words: Sarcoma, uteri, Cameroon.

INTRODUCTION

Cancer is often neglected in developing countries including Cameroon where it has been reported to be a public health problem (Mbakop et al., 1992). Uterine stromal tumors constitute a spectrum of rare neoplasms that exhibit varying degrees of malignancy and can present a number of challenges with respect to diagnosis and classification (Jason et al., 2001). They are derived from the mesenchyme of the corpus consisting of endometrial stroma, smooth muscle and blood vessels, or an admixture of these.

Although uterine sarcomas are rare, their benign counterparts, especially leiomyoma is very common and unless a high level of suspicion is shown with complete pathological examination of surgical specimens, uterine sarcomas are likely to be misdiagnosed.

MATERIALS AND METHODS

Thirteen histologically confirmed primary sarcomas of the uterine corpus were analyzed in this retrospective study between 2004 and 2008 in the pathology service of the General Hospital Yaoundé. The diagnosis was review-confirmed in each case and the pathological and epidemiological data of the patients were retrieved, reviewed and analyzed. All cases registered out of this period were rejected. Histological grading was done using tumour size, mitotic index, cell nuclear and tumour border characteristics, necrosis and vascular invasion. Staging was by the Tumour Node Metastasis (TNM) standards. Patients were graded socio-economically by their level of education and profession. Chi-squared tests were done for data evaluation.

RESULTS

Clinical parameters

Age

The age of all patients ranged between 27 to 75 years, at
an average of 45.25 (+2) years, and a median of 45 years. Patients were mainly in the 30 to 49 years age group (61.5%) Table 1. The average age for patients with leiomyosarcoma was 38.8 years, 39.7 years for endometrial stromal sarcoma, 48 years for undifferentiated endometrial sarcoma and 63 years for endometrial carcinosarcoma. About 33.3% of endometrial stromal sarcoma (ESS) cases were premenopausal against 50% postmenopausal, while 80% of leiomyosarcoma (LMS) patients were premenopausal against 20% postmenopausal. Both cases of undifferentiated endometrial sarcoma (UDES) and endometrial carcinosarcoma (ECAS) were postmenopausal.

Clinical presentation

Patients presented with abdominal pain (23%), uterine enlargement/mass (100%) and bleeding (15.4%). In 7.7% each, the presentation was a complication of either a pregnancy or an intrauterine contraceptive device (IUCD) respectively.

Socio-economic status

The patients were of low (38.4%), average (15.4%), or high (15.4%) socio-economic class. There was no significant difference (p> 1.5) in the status amongst the patients.

Clinical diagnosis

81.8% of cases were clinically diagnosed as uterine leiomyoma.

Disease outcome

All patients (100%) survived after the initial treatment that included surgery and chemotherapy. Long term mortality studies were not done as most patients were lost to follow up.

Pathological parameters

Histology

There were 5 LMS (38.5%), 6 ESS (46.1%), 1 UDES (7.7%), and 1 ECAS (7.7%). Grading: 46.2% of the tumours were low-grade against 15.4% high grade.

Tumour size

The average largest diameter for all tumours was 4.5cm. Respectively, the average size was 7.5cm for LMS, 4.4cm for UDES, 3.5cm for ESS and 2.8cm for CAS.

Stage

All tumours were found to be stage I or II at time of diagnosis.

Macroscopic specimens

The diagnosis was made from dilatation and curettage (D&C) products (23%) and/or, myomectomy (23%), hysterectomy (64%) specimens.

DISCUSSION

From previous studies, the most common sarcomas of the uterine corpus are LMS and ESS, both reported to be frequent in black than white women (Harlow et al., 1986; Madison et al., 1998). Sarcomas constitute only about 0.01% of all malignant tumours of the uterus in our community (Abondo et al., 1994). The sarcomas of the uterine corpus found in this study include ESS (46.1%), LMS (38.5%), UDES (7.7%) and ECAS (7.7%) (Table 2). This outcome is different from Liokomovich (1999), who found 10% for ESS in his series.

However, our annual incidence of 2.6 and 75% postmenopausal patients is similar reports in literature (Olivia et al., 1998). The patients presented with a uterine mass (100%), abnormal uterine bleeding (15.4%) and pain (23%). These are classical signs and symptoms of the disease (Madison et al., 1998). 84.5% of our cases were clinically diagnosed as leiomyoma. The diagnosis was made on specimens which were either products of cervical dilatation and curettage (D&C) (23%) and/or myomectomy (23%), hysterectomy (64%).

In Tahereh’s study, ESS was diagnosed on D&C products in 28.5% (Tahereh et al., 1996). Histological grading of the tumours was by use of mitotic index, cell nuclear characteristics, necrosis and vascular invasion as in other studies (Taylor and Norris, 1996; Bell et al., 1994).

Our patients were classified socio-economically using the level of education and profession. The p value for this parameter was not significant amongst them (p>1.5). ESS and LMS have been reported to be very rare tumours of the uterus that can be mistaken for leiomyoma (Richard and Michael, 2000; Leibsohn et al., 1990; Parker et al., 1994).

In our study, ESS comprised about 0.16% of all genital tract malignant neoplasms recorded in our service during the period under study (Abondo et al., 1994). This is a similar finding in the report by Chang et al of 0.2% (Chang et al., 1990). Some studies have demonstrated
the mean age for patients with ESS to be 42 to 58 years, with 10 to 25% of patients being premenopausal (Liokumovich et al., 1999; Tahereh et al., 1996). Our patients were relatively younger with a mean age of 39.7 years and 33.3% were premenopausal. Similar to previous reports (Digani et al., 1989), in ESS the tumour was limited to the uterine cavity at time of diagnosis. This is contrary to some authors (young et al 1986) who found extrauterine extension of the tumor in up to 30% of women with LG-ESS at hysterectomy. One patient with ESS in our series had had an intrauterine contraceptive device (IUCD) for about ten years. It was investigations for this device that lead to discovery of the tumor. This finding has not been reported in literature, to the best of our knowledge.

Histologically, 50% of cases of ESS were each of low-grade (LG-ESS) and high-grade (HG-ESS) respectively. Low grade ESS is a rare tumor with an indolent growth and late recurrences. This affects younger women than other uterine malignancies with mean ages ranging from 42 to 58 years, and about 10 to 25% of patients being premenopausal (de Fusco et al., 1989). UDES is a high grade and aggressive endometrial sarcoma with no histological resemblance to endometrial stroma and characterized histologically by the presence of both malignant glandular and sarcomatous components (Bell et al., 1994). These tumours are common in postmenopausal women (de Fusco et al., 1989), with poor prognosis even in stage I disease (Kempson and Bari, 1970). We documented one case (7.7%) of UDES aged 48 years. ECAS is a rare, aggressive disease, accounting for approximately 3% of all uterine neoplasms (Kempson and Evans, 1970). ECAS is a rare, aggressive disease, accounting for approximately 3% of all uterine neoplasms.

Ferrandina et al., 2007), that often occur in postmenopausal women, with as many as one third of the patients having clinical evidence of extra uterine spread at presentation (Richard and Michael, 2000). It may be diagnosed as a recurrence in patients with anaplastic endometrial cancer (Ferrandina et al., 2007), and it is found commonly in conjunction with a choriocarcinoma (Hanh et al., 2000). ECAS in our series occurred in a woman aged 63 years, older than the average age of 57 years by Ferrandina (2007).

Uterine LMS is a highly malignant neoplasm Major et al., 1993), with similar symptoms to leiomyoma (Leibsohn et al., 1990). In our study, LMS constituted 38.5% of uterine sarcomas, second to ESS (table 2). Our patients with LMS were relatively younger with a mean age of 38.8 years and a median of 40 years, and 50% of the patients were premenopausal. This is unlike reports invarious studies which found the median age of patients with LMSC to be 50 to 55years (Gadducci et al 1986) and 15% of these patients being younger than 40 years.

In our study, all patients with LMS were above 30 years (Deligdisch et al., 1997) and had a tumour average diameter of 7.5cm, slightly larger than the 5cm reported by Evans (1988). Using known grading criteria, 40% of our LMS were well differentiated low-grade tumours (table 2). All patients in this study were alive and well after initial treatment by surgery and chemotherapy.

Table 1. Distribution of patients by age.

<table>
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<tr>
<th>Age range (years)</th>
<th>Number of cases</th>
<th>%</th>
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<td>50-59</td>
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<td>7.7</td>
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<tr>
<td>70-79+</td>
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<td>7.7</td>
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<tr>
<td>Age unknown</td>
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<td>7.7</td>
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<td>All ages</td>
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<td>100</td>
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</table>

Table 2. Distribution of cases by histological type and grade.

<table>
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<th>Histological type</th>
<th>Grade</th>
<th>%</th>
<th>Total</th>
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<td>Endometrial stromal sarcoma</td>
<td>Low</td>
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<td></td>
<td>Unknown</td>
<td>2</td>
<td>6</td>
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<tr>
<td>Leiomyosarcoma</td>
<td>Low</td>
<td>2</td>
<td>38.5</td>
</tr>
<tr>
<td></td>
<td>High</td>
<td>1</td>
<td>5</td>
</tr>
<tr>
<td></td>
<td>Unknown</td>
<td>2</td>
<td>5</td>
</tr>
<tr>
<td>Undifferentiated endometrial sarcoma</td>
<td>Low</td>
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<td>7.7</td>
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<tr>
<td></td>
<td>High</td>
<td>1</td>
<td>1</td>
</tr>
<tr>
<td></td>
<td>Unknown</td>
<td>0</td>
<td>1</td>
</tr>
<tr>
<td>Endometrial carcinosarcoma</td>
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<td>1</td>
<td>7.7</td>
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<tr>
<td></td>
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</tr>
<tr>
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<tr>
<td>Total</td>
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<td>100</td>
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growing tumours and have a good prognosis. They present clinically with signs suggestive of benign lesions of this organ, like leiomyoma, which delays diagnosis and management. In an environment like ours where most patients see a physician as a secondary option with majority likely to defect, clinicians should have a high level of suspicion for sarcoma on any seemingly benign uterine lesion. Furthermore, all surgical specimens of the uterine corpus should be submitted for pathology analysis to rule out these malignancies. We recommend further in-depth studies to document the trends in incidence and disease outcome of uterine sarcomas in our community.

REFERENCES