Primary leiomyosarcoma of the fallopian tube: A case report

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Summary

A primary leiomyosarcoma (LMS) arising from the left fallopian tube in a perimenopausal 48-year-old woman is reported. Primary tubal LMS is an uncommon, exceedingly rare neoplasm, accounting for only a few reported cases so far. To our knowledge, the present case is the 17th tubal LMS reported in the English-language literature. The diagnosis is usually made at the time of laparotomy for a pelvic or adnexal mass or other gynaecological indications.

As in ovarian neoplasms, the mainstay of treatment is represented by debulking surgery consisting of total abdominal hysterectomy, bilateral salpingo-oophorectomy, random biopsies, peritoneal washing and excision of all the abdominal tumour masses. Although the approach is radical, the clinical behaviour is very poor. The role of adjuvant radio- or chemotherapy still remains unsolved.

Key words: Leiomyosarcoma; Mixed mesodermal tumour; Ovarian and fallopian tube neoplasm; Treatment.

Introduction

Sarcomas are very uncommon, heterogeneous and aggressive malignant neoplasms of the female genital tract, which usually affect the uterus, accounting for 4% of all uterine tumors [1]. In the ovary they are very rare, representing less than 1% of all ovarian malignancies [2], with an estimated incidence of about one in 40 compared to the epithelial type [3].

In the fallopian tube, overall sarcomas are also very uncommon. Few cases of malignant mixed müllerian tumors [4-7] and carcinosarcomas [8-10] have been reported. Primary tubal leiomyosarcomas (LMS) are even more rare than other sarcomas. This gynaecological neoplasm is usually very aggressive and characterised by a poor clinical behaviour, with a significantly high rate of local recurrences and blood-borne metastases [11].

A case of primary fallopian tube leiomyosarcoma treated with an optimal surgical approach is reported.

Case Report

A 48-year-old female smoker presented in May 1999 to the Department of Gynaecologic Oncology of Regina Elena National Cancer Institute of Rome with a suspected ovarian tumour.

Physical examination showed a palpable abdominal pelvic mass arising from the left adnexa and moderate abdominal pain. Ultrasound examination showed an antiflexed bulky myomatous uterus and an hypoechoic mass of 8 cm in diameter, arising from the left adnexal area. The right ovary and Douglas pouch were unremarkable.

Magnetic resonance imaging (MRI) with a paramagnetic enhancement agent (Gd-DTPA) confirmed the presence of a bulky myomatous uterus (10 x 9 x 8 cm) and a 7 cm solid, oblong structure arising from the left adnexa, likely from the corner of the left fallopian tube.

In the axial and coronal T2-weighted MRI (Figures 1 and 2) the adnexal tumour presented diffuse hyperintensity, with some central fluid areas. The contours were regular with a thin hypointense capsule and good cleavage from the pelvic and vascular structures. The retroperitoneal lymph nodes were normal.

Her past history was unremarkable with the exception of an appendectomy when she was 12 years old and a colecystectomy at age 46. No menstrual disorders had been reported by the patient.

Serum chemistries, blood count, haemoglobin, alpha-fetoprotein, carcinoembrionic antigen and CA125 were all within the normal range. Chest X-ray was also normal.

The patient was submitted to explorative laparotomy which confirmed the presence of a bulky fibromatous uterus with a 7 x 7 x 5 cm left fallopian tube mass. Once excised, the left tubal tumour showed an undifferentiated neoplasm at intraoperative frozen section.

As routine radical staging, a total extrafascial hysterectomy, bilateral salpingo-oophorectomy with excision of the tubal tumour, omentectomy, peritoneal washing and random biopsies were performed. No retroperitoneal lymphadenopaties were detected at intraoperative palpation. Postoperative recovery was normal.

Macroscopically the tubal tumour was pseudoencapsulated, greyish-rose and fleshy. Histologic examination of the mass demonstrated a malignant tumour characterized on one side by spindle-shaped cells arranged in fascicles with blunt-ended nuclei and eosinophil cytoplasm and on the other side by a more epitheliomorphic cell population with extensive cytoplasm and bloated nuclei. Mitotic activity was very high, with an average of 20 mitoses per 10 high-power fields (HPF). There were focal areas of haemorrhage and necrosis (Figure 3).

In both the cell populations the immunohistochemical study showed negative keratin and neuroendocrin markers, and positive muscular markers (desmin and actin).

Finally, a diagnosis of primary fallopian tube leiomyosarcoma was made. The tumour infiltrated largely the fallopian

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Fig. 1

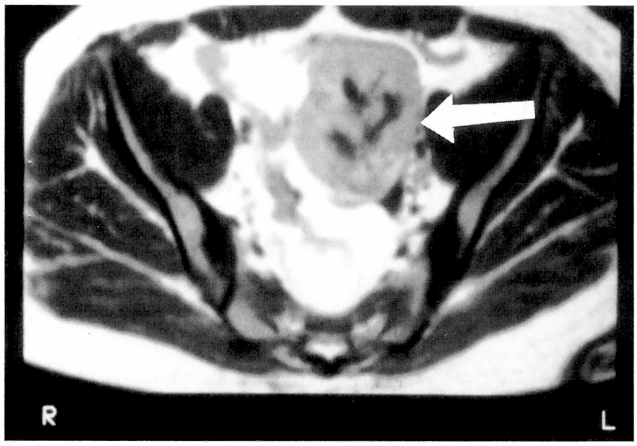

Fig.

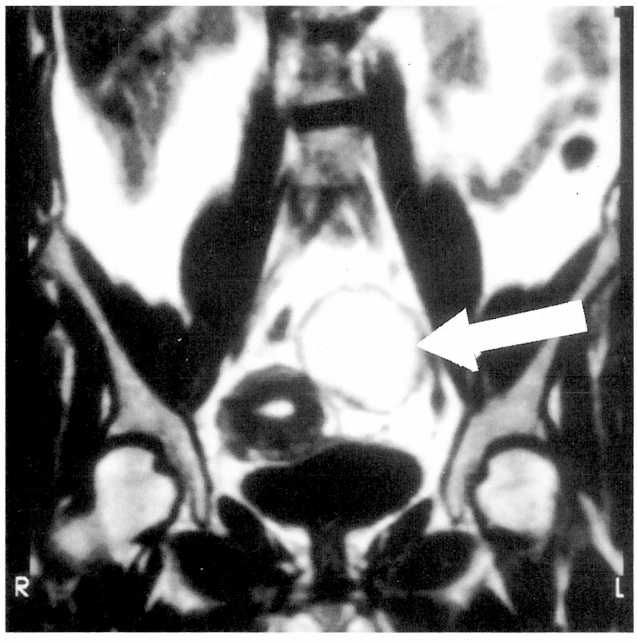

Fig. 3

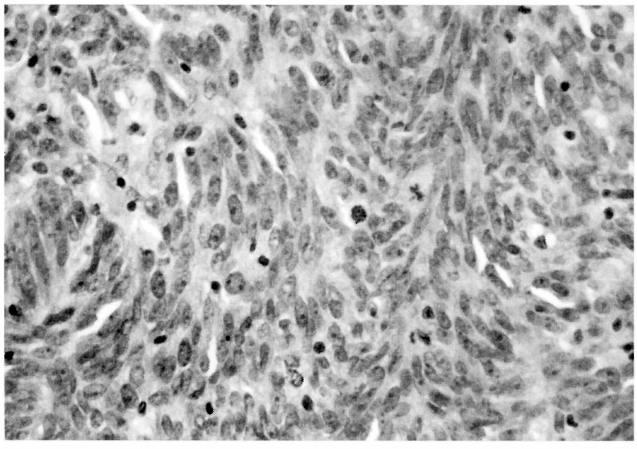

Figure 1. — Axial T1-weighted MRI. After IV infusion of Gd-DTPA the tumour (arrow) shows a high enhancement of signal with central heterogeneity.

Figure 2. — Coronal T2- weighted MRI. The tumour (arrow) arising from the corner of the left fallopian tube presents a diffuse hyperintensity with small central fluid areas. The contours are regular with a thin hypointense capsule and good cleavage from the pelvic and vascular structures.

Figure 3. — Leiomyosarcoma of the fallopian tube is characterised by spindle-shaped cells with blunt-ended nuclei aranged in fascicles (x 400).

tube muscularis, but the serosal surface was not affected at all. Microscopic examination of the homolateral and controlateral ovary, uterus and omentum showed no evidence of any malignancy. The peritoneal washing, using both morphologic and immunocytologic evaluations, was negative.

According to the current surgical staging for tubal carcinoma [12], the tumour was staged as FIGO Ia.

No complementary postoperative radio- or chemotherapy was performed.

The patient was invited to follow-up at 2-month intervals in the first six months, and up-to-date, after almost five years from treatment, she is still alive and free of disease.

Discussion

The female genital tract is rarely affected by sarcomas [1] and even more exceedingly rare is a primary leiomyosarcoma arising from the fallopian tube [11].

Indeed, Senger described the first case of fallopian tube sarcoma in 1886 [13] and, excluding malignant mixed müllerian tumours, only a few of such cases have been reported in the literature. In 1993 Jacoby *et al.* [11] reviewed the historical data on tubal sarcomas, identifying 15 LMS or spindle-cell sarcomas and added one case of their own [13-18]. To our knowledge, from then to 1999 no further cases of such tubal sarcomas have been reported.

Although most of non-uterine LMSs usually show a gradually progressive increase with age [19], fallopian tube sarcoma seems to be predominantly a perimenopausal disease with a median age of 47 years [11].

As for ovarian malignancies, symptoms are not at all specific (pelvic pain, enlargement of the abdomen) and the diagnosis is usually incidental during explorative laparotomy for a pelvic mass, or other gynaecological indications or presumed ovarian masses.

Although the CA125 level may be elevated at the time of diagnosis, such as in ovarian carcinoma, in the present case it was within the normal range (< 35 U/ml).

As shown in Figure 2, MRI findings with a paramagnetic agent (Gd-DTPA) resulted highly enhanced. This data, if further confirmed, could suggest that MRI with Gd-DTPA may be clinically useful for preoperative detection of sarcomas, as already recently suggested for uterine carcinosarcoma by Takemori *et al.* [20].

Like at the ovarian site, tubal sarcomas are characterised by a considerable high tumour-size and are almost always unilateral. Indeed, the present case, as well as other previous cases [11, 15, 16], were staged as early disease confined to the tube.

As also stated for sarcomas arising from the uterus [21], the extent of disease, amount of residual tumour after primary surgery and number of mitoses appear to be

the most important prognostic variables, as well as tumour size. In 37 uterine leiomyosarcomas treated at the M. D. Anderson Cancer Center most patients with tumours 5 cm or less survived compared to patients with bulky masses over 5 cm [22].

Despite of aggressive therapy, uterine sarcomas have been considered as the most lethal of all the gynaecological malignancies, with high metastatic potential, frequent recurrences and cancer-related death [23].

Moreover fallopian tube sarcomas, and particularly LMS, behave very aggressively, with a poor overall survival.

Primary treatment overlaps the standard surgical approach to epithelial fallopian tube or ovarian carcinomas, consisting of total abdominal hysterectomy, bilateral salpingo-oophorectomy, omentectomy, peritoneal washing, cytoreduction of all resectable tumour masses and accurate inspection of the peritoneal surfaces and visceral organs [10-19].

Also after complete surgical debulking a high rate of local and distant relapses, particularly liver and lung metastases [11], have been reported. All recurrences usually occur within two years of diagnosis, although distant metastases may also appear decades later, as in the case reported by Blarkley [18] 19 years after the treatment.

The overall survival rate of tubal sarcomas seems to be worse than for tubal adenocarcinoma which is considered, based on the Roswell Park experience [24], as virulent as ovarian cancer. In a GOG study of ovarian sarcomas, which may resemble the behaviour of tubal LMS, Morrow *et al.* reported cancer-related deaths of 23 out of 30 patients within 16 months from treatment [25].

As already suggested [26, 30], the benefit of complementary postoperative treatment for tubal or ovarian LMS has yet not been confirmed due to the conflicting international data. In Monk *et al's.* series [27] the role of adjuvant chemotherapy in early stage ovarian LMS does not emerge at all.

On the contrary, a survival benefit in ovarian mixed mesodermal sarcomas with a combination of postoperative concomitant radio-chemotherapy with a VAC regimen (vincristive, actinomycin-D, cyclophosphamide) has been reported [28].

Moreover, the MAID regimen (mesna, doxorubicin, ifosfamide, dacarbazine), tested for sarcomas of the trunk and extremities and which has proved to be effective in advanced ovarian sarcomas [29], may also be useful as adjuvant treatment for tubal sarcoma. The extreme rarity of such tumour does not permit sufficient data to be collected to draw any definitive conclusion about the therapeutic strategy.

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