Ovarian fibrosarcoma with five-year survival: a case report

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Summary

A 45-year-old postmenopausal woman, $Gravida_6$, $Para_4$, $Abortus_0$, Dilatation x $Curhetage_2$, came to the gynaecology department with pelvic pain. The tumor had arisen in the right ovary and measured 15 x 12 x 7 cm. Its cut surface varied from grey-white with a whorled appearance and showed areas of haemorrhage. Histologically the tumor was densely cellular, composed of spindle cells, diffusely involved the entire ovarian stroma with no normal ovarian structures remaining. Tumor cells had hyperchromatic nuclei with prominent nucleoli. There was moderate pleomorphism and the number of mitotic figures was an average of 6 per 10 high power fields. In the immunohistochemical study, the tumor was negative for desmin, muscle-specific actin, estrogen, progesterone receptors and CD31, but was positive for vimentin. A low proliferation index with Ki-67 was determined. The patient has shown no evidence of recurrent disease for five years.

Key words: Sarcoma; Fibrosarcoma; Ovary.

Introduction

Primary fibrosarcoma of the ovary is uncommon [1]. Although fibrosarcoma is usually seen in menopausal and postmenopausal women [2], occasional cases have been observed in children [1]. It has been reported that fibrosarcoma was a new component in nevoid basal-cell carcinoma syndrome in the case of an 8-year-old girl [3]. The prognosis is generally poor [1]. Occasionally, the course of the disease is more protracted, with patients surviving many years from time of diagnosis [1, 4]. We report an unusual case of ovarian fibrosarcoma that has shown no evidence of recurrent disease for five years.

Case Report

The patient, a 45-year-old postmenopausal woman, $Gravida_6$, $Para_4$, $Abortus_0$, Dilatation x $Curhetage_2$ (G_6, P_4, A_0, D x C_2), was admitted to our hospital complaining of pelvic pain. Physical examination revealed a dense and painful mass at the right lower quadrant of her abdomen. The remainder of the physical examination was unremarkable. Abnormal laboratory studies included CA 125: 71.9 (N: 1.7-32). Pelvic ultrasound examination showed a right ovarian mass, measuring 140 x 83 cm that multicystic with solid areas. The clinical diagnosis of a torsioned ovarian mass was made. At laparotomy, a right enlarged solid-haemorrhagic ovarian mass was found which was completely excised. In addition, total abdominal hysterectomy, left salphingo-oophorectomy, omentectomy and appendectomy were performed. Residual right ovarian tissue and tuba were not identified. The tumor mass measured 15 x 12 x 7 cm. The cut surface varied from grey-white with a whorled appearance and showed areas of haemorrhage. The uterus, left tube, ovary, appendix, and omentum were free of tumor. Histologically the tumor was densely cellular and composed of spindle cells with no normal ovarian structures remaining. Tumor cells had hyperchromatic nuclei with prominent nucleoli (Figure 1).

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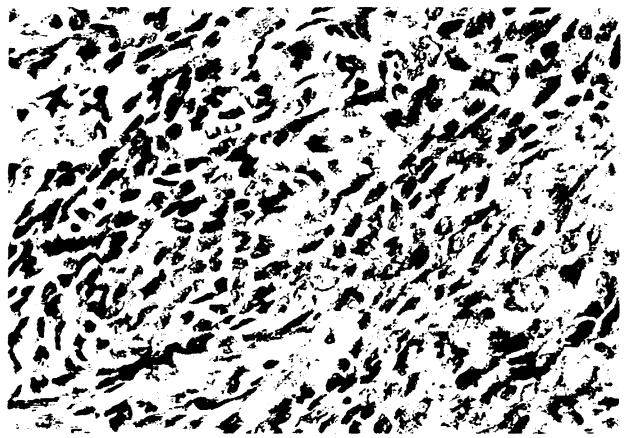

Figure 1. — Fibrosarcoma consisting of spindle-shaped cells with a hyperchromatic nucleus (H-E x 20).

The number of mitotic figures was an average of 6 per 10 high power fields. It was observed with histochemical staining that reticulum fibres were wrapped around tumor cells one by one. A standardized streptavidin-biotin-peroxidase method on formalin-fixed paraffin-embedded tissue for immunohistochemistry was performed. Tumor cells were negative for desmin (NeoMarkers 1:40), smooth muscle actin (NeoMarkers 1:100), S-100 (Neo Markers 1:50), estrogen receptors (NeoMarkers 1:50), progesterone (NeoMarkers 1:50) and CD 31 (Dako 1:20), but were positive for vimentin (NeoMarkers 1:100). A proliferation marker, Ki-67 (Dako 1:50), was determined by counting at least 1,000 cells in more than ten different areas of sections and was observed in 2.5% of tumor cells. One year later, multiple biopsies were performed on abdominal re-exploration and tumor was not determined. Four years following the second surgical procedure, there was no evidence of recurrent tumor or distant metastasis.

Discussion

Fibrosarcoma may arise de novo from ovarian stroma or may originate as a result of malignant change in pre-existent fibromas [1]. Although cellular fibroma is a benign tumor, it mimics fibrosarcoma histologically. Prat and Scully [5] reported that the classification of benign cellular fibroma and malignant fibrosarcoma should be based on mitotic activity. The presence of more than 4 mitotes/10 high power fields places the tumor in the fibrosarcoma category. Immunohistochemical staining for Ki-67 antigen was employed to assess the proliferative activity. The Ki-67 labeling index has been related to prognostic factors for soft tissue sarcomas [2]. Tsuji *et al.* reported MIB-1 and proliferative indices in fibrosarcomas were higher than those in cellular fibromas [2]. Their findings of proliferative activity correlate with the criteria presented by Prat and Scully [5]. In addition, macroscopically, the tumor usually forms a round and oval solid mass like a fibroma, but fibrosarcoma is larger and is mostly associated with haemorrhage and necrosis. Adult granulosa cell tumors may be confused with ovarian fibrosarcomas. Reticulum staining may show abundant intercellular fibrils in fibrosarcoma, unlike the scant reticulum of adult granulosa cell tumors.

Clinically, there is usually an unremarkable gynaecologic history. Our patient's only complaint was pelvic pain. In most reports, although surgical resections for this type of tumor range from simple adnexectomy to total abdominal hysterectomy and bilateral salphingo-oophorectomy and omentectomy, tumor usually recurs within eight years and most patients die within two years [6].

However, the extent of the surgical resection does not offer protection against recurrence [5]. Tumors showing less marked mitotic activity tend to be less aggressive [3]. In the same way, the number of mitotic figures was an average of 6 per 10 high power fields and the Ki-67 labelling index was 2.5% in our case, and there was no evidence of recurrence. We agree that these criteria are very important for the prognosis of ovarian fibrosarcoma.

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