Leiomyosarcoma of the broad ligament arising in a pre-existing pure neurilemmoma-like leiomyoma

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Summary

A 35-year-old woman presented with an adnexal mass which came out as a leiomyosarcoma of the broad ligament arising in a pre-existing pure neurilemmoma-like leiomyoma. This is the first reported case of leiomyosarcoma arising in a pure neurilemmoma-like leiomyoma, the third reported case at an unusually young age showing evidence of a pre-existing leiomyoma and the 12th reported case of leimyosarcoma of the broad ligament. Is the likelihood of malignant transformation, especially in large leiomyomas of the broad ligament, a rare event?

Key words: Leiomyosarcoma; Broad ligament.

Introduction

The most common solid tumor of the broad ligament is leiomyoma [1]. Gardner *et al.* established strict criteria for tumors of the broad ligament, requiring that "they are completely separated from and in no way connected with either the uterus or the ovary". Adhering to these criteria, only 11 cases of typical leiomyosarcoma and two cases of leiomyosarcoma arising within a leiomyoma of the broad ligament have been reported in the modern literature [2-5].

Case Presentation

A 35-year-old woman presented with abdominal distension and pain in the right lower side of a year's duration. On physical examination, her abdomen was slightly distended with tenderness in the right lower quadrant; a 15 cm, firm, solid mass was noted in the right adnexal region. Ultrasonography showed a solid mass measuring 15 x 15 x 10 cm. Her past history was significant for a previous right bilateral salpingo-oophorectomy four years before revealing chronic salpingitis and a follicle cyst in the ovary.

A solid, firm mass, arising from the right broad ligament measuring 17 cm in the greatest diameter was noted during laparotomy. The mass was removed in conjunction with a hysterectomy. Frozen section was reported as benign mesenchymal tumor of the broad ligament.

The tumor measured 17 x 15 x 8 cm and was unencapsulated, lobulated but contained multiple confluent gelatinous nodules ranging from 2-3 cm in diameter. These nodules occupied approximately 10-20% of the tumor volume. The tumor surrounding these nodules had the typical whorled appearance of leiomyoma while the nodules lacked this appearance.

Microscopic examination of the gelatinous nodules showed pleomorphic sarcoma, characterized by high mitotic activity, marked nuclear pleomorphism, giant cells, necrosis and vascular invasion.

Microscopic examination of the surrounding leiomyoma revealed interlacing bundles of syncronously oriented spindle-shaped cells having elongated snake-like nuclei arranged in a palisade fashion, showing the typical Verocay bodies associated with Antoni type A nerve sheath tumors (Figure 1).

At immunohistochemistry the neoplastic cells of both components showed positive staining for antibodies against vimentin, desmin and actin; they were negative for S-100.

The mitotic count in areas of pleomorphic sarcoma was 15-20 mitoses per 10 HPF, however, in areas of neurilemmoma-like leiomyoma, it was 0-2 mitoses per HPF. MIB-1 immunostaining showed positive nuclear staining in approximately 60% of the cells in areas of the pleomorphic sarcoma (Figure 2), whereas in areas of neurilemmoma-like leiomyoma staining was seen in approximately 1% of the cells.

The endometrium was in the proliferative phase. There were seven small leiomyomatas in the myometrium. There were no findings to suggest that the tumor originated anywhere other than the broad ligamant. The whole soft tissue margin of the uterus was examined microscopically presenting no evidence of tumor. With the aid of the prior history of a right salpingo-oophorectomy, morphologic and immunohistochemical findings, the lesion was concluded to be a leiomyosarcoma of the broad ligament arising in a pure neurilemmoma-like leiomyoma.

Repeat laparotomy was done four weeks later performing left salpingo-oophorectomy and pelvic lymphadenectomy. No gross abnormality was noted during the exploration of the whole abdomen. On microscopic examination of the left adnexa and lymph nodes, no evidence of tumor was seen.

Discussion

Primary malignant tumors of the broad ligament are quite rare with leiomyosarcoma being the most common from [4]. Our patient is of interest for a number of reasons. Leiomyosarcoma of the broad ligament occurs mainly in women in their postmenopausal years and carries a poor prognosis. The mean age is 57 years [3]. Two patients in the literature were unusually young to have broad ligament leiomyosarcoma at the ages of 31 and 36 years [6, 7]. Our patient was 35 years old.

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Fig. 1

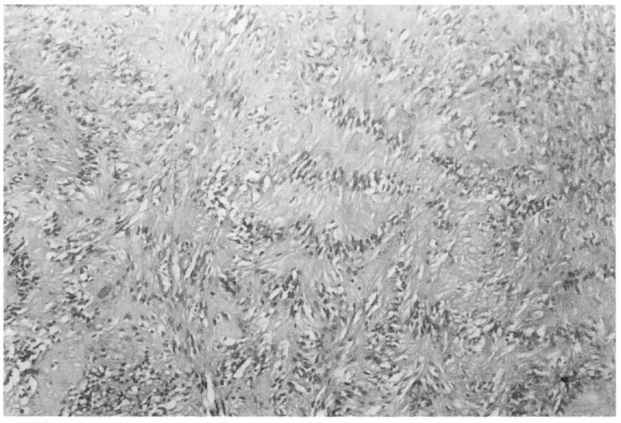

Fig. 2

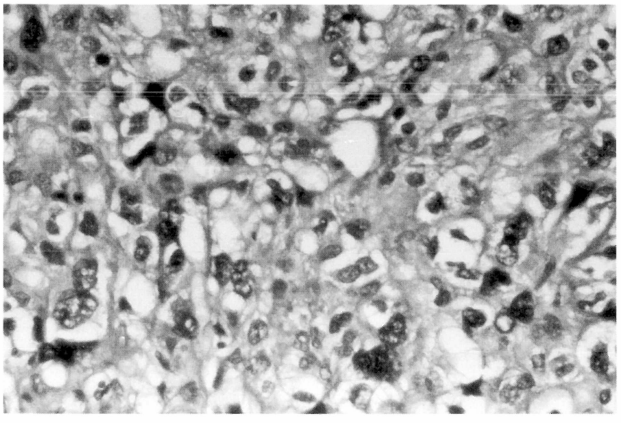

Figure 1. — Interlacing bundles of syncronously oriented spindle-shaped cells having elongated snake-like nuclei arranged in a palisade fashion (H&E x 100).
Figure 2. — Areas of pleomorphic sarcoma (H&E x 400).

In the English literature, two leiomyosarcomas of the broad ligament arising within a pre-existing leiomyoma have been reported [4, 5]. The present case is the third case of leiomyosarcoma of the broad ligament arising within a leiomyoma and it is the first case of leiomyosarcoma of the broad ligament arising within a pre-existing pure neurilemmoma-like leiomyoma which is occasionally a uterine tumor displaying a rhythmic pattern of nuclear arrangement with formation of Verocay-like bodies appearing as those of the Antoni type A neurilemmoma [8, 9]. These numbers, although limited to indicate malignant transformation into leiomyoma of the broad ligament, may not be a rare, isolated event, particularly in larger leiomyomas.

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