

PRIMARY OVARIAN MYXOMA

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Abstract: Primary ovarian myxoma is exceedingly rare, with only five cases known to be reported. We present an 18-year-old female with an ovarian myxoma.

Index Terms

ovarian myxoma, histology, histogenesis

INTRODUCTION

Virchow described myxoma in 1863¹⁾ and precisely defined it in 1871²⁾. Stout in 1948³⁾ established the basic histological criteria for diagnosis. However, its histogenesis remains a moot point.

Myxomatous tissue exists postnatally only in the umbilical cord and the dental pulp. Primary ovarian myxoma is exceedingly rare, with only five cases previously reported in the world literature⁴⁾⁵⁾⁶⁾⁷⁾⁸⁾. We recently observed a patient with ovarian myxoma.

CASE HISTORY

A 18-year-old Japanese female, gravida 0, para 0, complained of abdominal distension. She underwent a gynecologic examination in March 1993 and was diagnosed as having a huge pelvic tumor. Since the menarche at age 12, menses had been regular every 35 days.

The patient was 160.5 cm tall and weighed 49.5 kg. A huge, firm, nontender mass was palpable in the abdomen. A trans-abdominal ultrasound examination, computed tomogram (CT) and magnetic resonance image (MRI) confirmed the presence of a predominantly cystic mass, measuring 18 cm in diameter. It consisted of fluid having a high specific gravity and little fatty tissue (Fig. 1). Laparotomy revealed a well circumscribed cystic, elastic, pale mass, 18×15×16 cm, that had replaced the right ovary. The size of the uterus was normal. The left ovary revealed a solid tumor measuring 3×4 cm. The remainder of the abdomen and pelvis appeared normal. We performed a right salpingo-oophorectomy and partially resected the left ovary. There was no evidence of recurrence at 12 months postoperatively.

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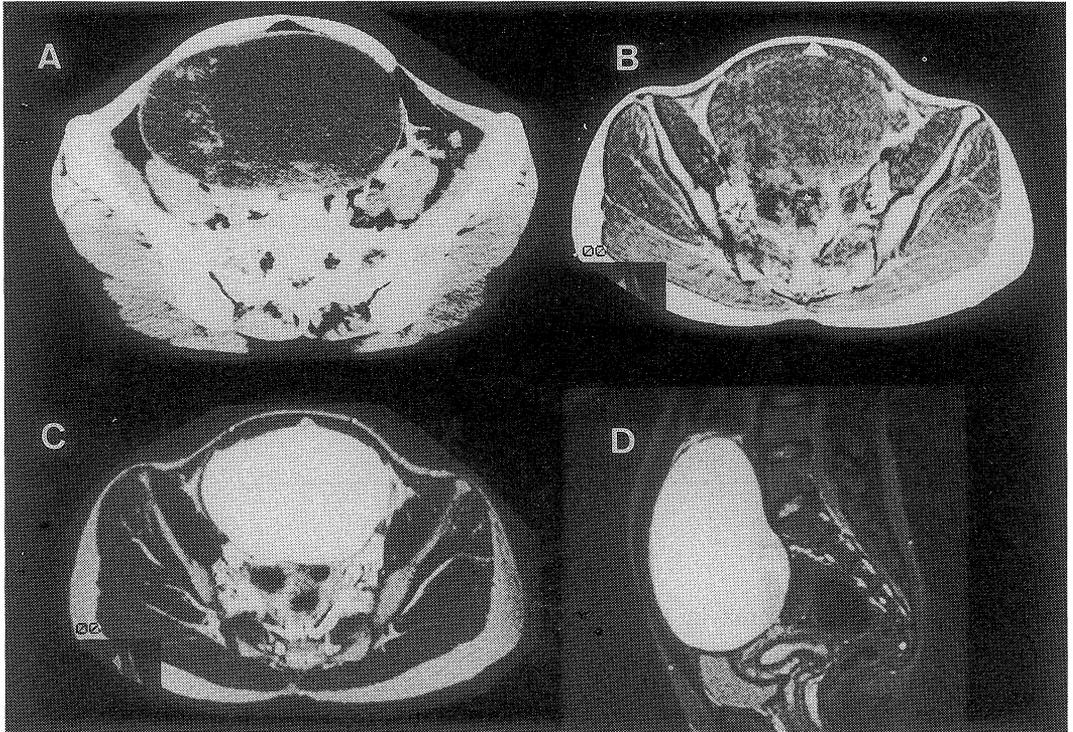


Fig. 1. A : CT scan of the pelvis.

An 18-cm predominantly cystic mass is evident.

B : Transaxial T1-weighted magnetic resonance image (MRI).

A huge low-signal-intensity mass is observed in the pelvis.

C : Transaxial T2-weighted magnetic resonance image (MRI).

The pelvic mass show an increased bright signal.

D : Sagittal T2-weighted magnetic resonance image (MRI).

There is hyperintensity of the tumor which suggests mucinous or myxomatous change.

PATHOLOGY

The right tumor was enclosed by a gray capsule without evidence of capsular invasion. The cut surface of the tumor exhibited multiloculated cystic spaces, which were filled with a glassy, viscid, gelatinous material with focal hemorrhage (Fig. 2). The left tumor showed the appearance of normal ovarian tissue. Microscopic examination revealed the tumor to be composed of uniform stellate and spindle shaped cells that were evenly dispersed in an alcian blue positive, loose abundant myxoid material. The nuclei were fine, uniform and hyperchromatic, but no mitotic figures were seen. The vascularity of the tumor was inconspicuous, and consisted of a few capillary sized vessels without plexiform pattern (Fig. 3).

PTAH staining of the tumor showed no cross-striations. Numerous sections of the tumor failed to reveal components such as fat, muscle, or cartilage. Special staining by antibodies to desmin, myoglobin, S 100-protein, alpha-1-antitrypsin and factor VIII was negative. Silver stain showed fine collagen fibers scattered in the tumor matrix (Fig. 4).

The excised left ovarian tissue consisted of a normal stroma and a mildly myxomatous

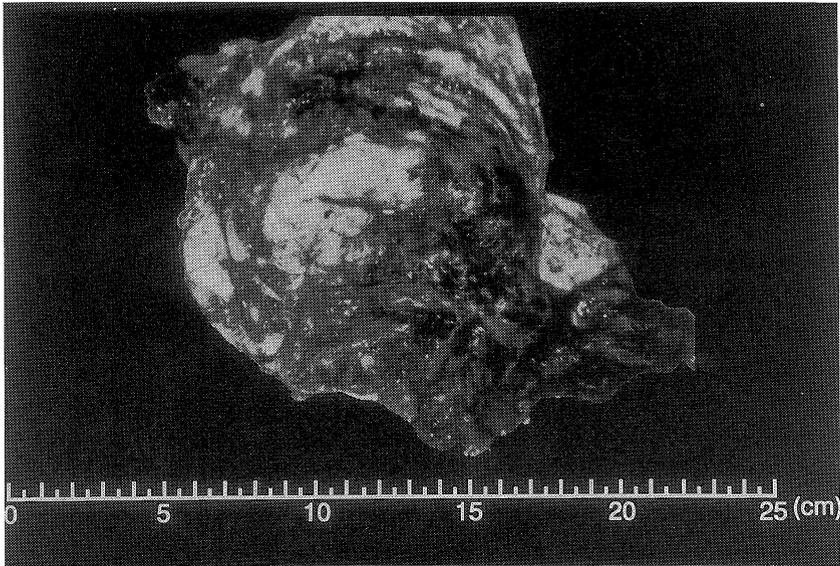


Fig. 2. Gross view of cut surface of the tumor.
Multiloculated cystic spaces are filled with a glassy, viscid, gelatinous material with focal hemorrhage.

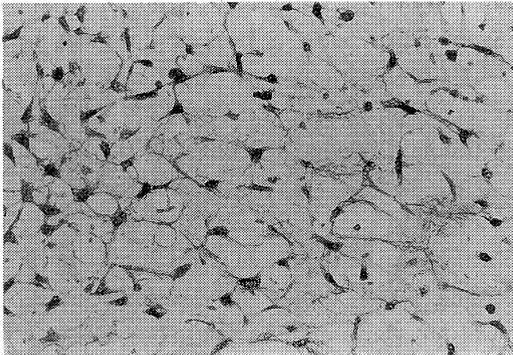


Fig. 3. Microscopic section. (Hematoxylin and Eosin stain)
The tumor is composed of uniform stellate and spindle-shaped cells that are evenly dispersed in an alcian blue-positive, loose, abundant myxoid material. ($\times 400$)

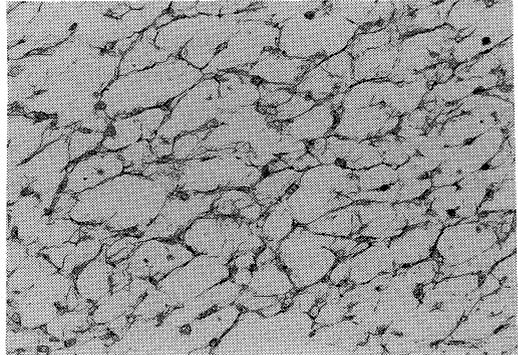


Fig. 4. Microscopic section. (Silver stain)
Silver stain shows fine collagen fibers scattered in the tumor matrix. ($\times 400$)

content, which apparently differs from that of the tumor in the right ovary.

Electron microscopic examination revealed that tumor cells were widely separated from each other. Lipid inclusions were not observed. As the specimen was obtained after paraffin fixation, we could not receive more detailed findings in the cytoplasm. The myxoid matrix contained only scattered collagen fibers.

DISCUSSION

According to Stout³⁹⁾, myxoma shows stellate and spindle shaped cells in a myxoid stroma that contains mucopolysaccharide with the presence of very delicate reticulum fibers. The tumor is poorly vascularized, and capillaries lack a plexiform arrangement.

Myxomas can arise in differing regions of the body. Pack and Ariel in 1958¹⁰⁾ reported 150 cases of myxoma, none of which had originated in the ovary. Only five cases of primary ovarian myxoma have been previously reported in the world literature⁴⁾⁵⁾⁶⁾⁷⁾⁸⁾. Dutz and Stout in 1961⁴⁾ were the first to report a primary ovarian myxoma (3.5 cm) in a 14-year-old female, whose outcome was unknown. Masubuchi *et al* in 1970⁵⁾ reported a 43-year-old female with right ovarian myxoma (9 cm). This patient showed no evidence of recurrence 12 months postoperatively. In 1978, Majmudar *et al*⁶⁾ described a right ovarian myxoma (8 cm) in a 25-year-old woman; this case also showed no postoperative recurrence. In 1987, Brady *et al*⁷⁾ reported the fourth case, a 32-year old woman with a right ovarian myxoma (9×6.3×4.4 cm) ; she also had no recurrence 18 months postoperatively. In 1987, a 28-year-old pregnant woman with a left ovarian myxoma (9.5 cm) was reported by Wakabayashi *et al*⁸⁾. She had no evidence of recurrence 14 months postoperatively. Our patient appears to be the sixth case reported.

Controversy exists as to the histogenesis of myxoma, especially of the ovary, as myxomatous tissue is not present in ovarian tissue. Willis¹¹⁾ indicated that myxoma is a fibroblastic tumor that exhibits substantial amounts of intercellular mucin. His hypothesis, presented in 1969, now seems unacceptable as a myxoma shows virtually no fibroblastic component. Also, ovarian myxomas are rare, whereas ovarian fibromas are common. Enzinger in 1965¹²⁾ indicated intramuscular myxomas are altered fibroblasts that produce mucopolysaccharides but that are incapable of assembling mature collagen because of the lack of aggressive growth and poor vascularization.

Electron microscopic examination has recently clarified the histogenesis of myxoma. In 1979 Feldman¹³⁾ noted the fibroblastic nature of the intramuscular myxoma cells. However, in 1987, Brady *et al*⁷⁾ suggested a myofibroblastic differentiation, while in 1989, Wakabayashi *et al*⁸⁾ stated that ovarian myxoma arises from primitive mesenchymal cells having the ultrastructural features of myofibroblasts.

Clinically, because of its cystic nature and myxomatous content a myxoma can be mistaken for a mucinous cystadenoma or a pseudomyxoma peritonei. The differential diagnosis based on histologic findings includes primary ovarian myxoma, myxoid liposarcoma, immature teratoma, embryonal rhabdomyosarcoma, fibrosarcoma, aggressive angiomyxoma, and myxoid subtype of malignant fibrous histiocytoma. A focal myxomatous change is also sometimes observed in liposarcoma, rhabdomyosarcoma, and chondrosarcoma. Therefore, patients with sarcomas with myxoid change must be carefully separated from those with pure myxomas.

Enzinger¹²⁾ reported that 34 patients with myxomas who were treated only by simple excision had a benign course. Our patient showed no evidence of recurrence 12 months after a simple resection. Once the myxoma has been differentiated with certainty from a sarcoma, the treatment involves surgical excision, including the uninvolved margins of the myxoma.

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