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Case Report

A case of unilateral ovarian fibrothecoma and pelvic aggressive angiomyxoma with a concurrent diagnosis

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Abstract

Aggressive angiomyxoma (AA) is a rare mesenchymal tumor that is more common in women and can cause local recurrences, usually originating from the pelvis. Surgical excision is the main treatment method. In patients who cannot be completely excised or who relapse, hormone therapy and radiotherapy can be other options. A rare subtype of ovarian sex-cord stromal tumor is called fibrothecoma (FT). Early diagnosis is common and surgical excision is typically enough for treatment. In this report, we describe the case of a 56-year-old woman who was simultaneously diagnosed with two distinct cancers.

Keywords: Aggressive angiomyxoma, fibrothecoma, pelvic tumors

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INTRODUCTION

Aggressive angiomyxoma (AA) is a rare benign stromal tumor that is more common in women and commonly located in the pelvic area (1). The most prevalent places of its origin are the vulva, perineum, and hips (2). Its rareness and lack of distinctive radiological features could make diagnosis challenging (3). Although surgical excision is the primary treatment, local recurrences are frequent (4).

One of the sex-cord stromal tumor subgroups, fibrothecoma (FT), is a relatively rare tumor characterized by both fibromas and thecomas (5). It makes up approximately 1.2% of all ovarian tumors (6). In contrast to other ovarian tumors, it is typically detected in the early stages and has a favorable prognosis after total surgical excision (7).

In this study, we present a case report of a patient with two rare medical conditions.

CASE REPORT

This study investigated a 56-year-old female patient with complaints of lower quadrant abdominal pain, constipation, and a palpable mass at our education and research hospital for several months. She had no disease except hypertension and there was no family history of malignancy. There was no vaginal or rectal bleeding. During lower gastrointestinal endoscopy, no lesions were detected in the colon. Pelvic computed tomography (CT) revealed a 15x10 cm heterogeneous mass in the right half of the rectum in the rectouterine area. After gynecological and urological examination, this lesion was found to exert external pressure on the relevant organs.

For a more comprehensive assessment of the pelvic magnetic resonance imaging (MRI) data, a mass lesion was observed in the pelvic region. This lesion was approximately 190x113 mm in size and filled the presacral area posterior to the uterus, extending to the perineal region. It was hyperintense on T2, hyperintense on fat-suppressed sequences, and hypointense on T1. The mass lesion pushed the uterus and bladder anteriorly and the rectosigmoid colon to the left lateral and had a significant compression effect (Figure 1).

Surgery was chosen for patients whose blood carcinoembryonic antigen (CEA) and cancer antigen 125 (CA-125) levels were normal and who had no metastases visible in the abdomen or thorax. Left unilateral salpingo-oophorectomy (USO) and total excision were performed. The presence of fibrothecoma was reported as a final pathological investigation of the left ovary (Figure 2, Figure 3). Upon examination of the pelvic mass, round, uniform, slightly spindled cells and numerous thick and thin-walled vessels were observed in the fibromyxoid stroma in hematoxylin-eosin-stained sections. Diffuse staining for estrogen and progesterone, as well as focal staining for desmin and smooth muscle actin (SMA), was observed through immunohistochemical analysis (Figure 4, Figure 5). No staining was observed for CD34, H caldesmon, S100, PAN CK, or beta-catenin. The Ki67 proliferation index was less than 1%. Aggressive angiomyxoma was diagnosed by histopathological and immunohistochemical findings.

The patient was assessed and followed up in the oncology clinic following the operation and an MRI was conducted four months later. In the present series, there was a lesion with modest peripheral capsular contrast enhancement. There was a possibility of residue or recurrence. Patients who had no distant metastases underwent a second operation at a different tertiary center. Right USO, total abdominal hysterectomy (TAH), and pelvic mass excision were carried out. Pathology of the mass indicated that it was chronic fibrin-granulation tissue. The normal histology of both was compatible with TAH + USO. Following surgery, the patient's general health was good. Her follow-ups are ongoing at our hospital, which is a tertiary center.

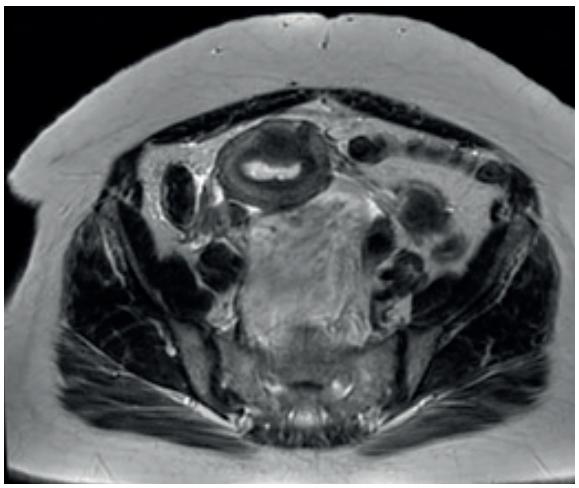

Figure 1. T2A axial image, the lesion is hyperintense as there is no fat suppressionon magnetic resonance imaging

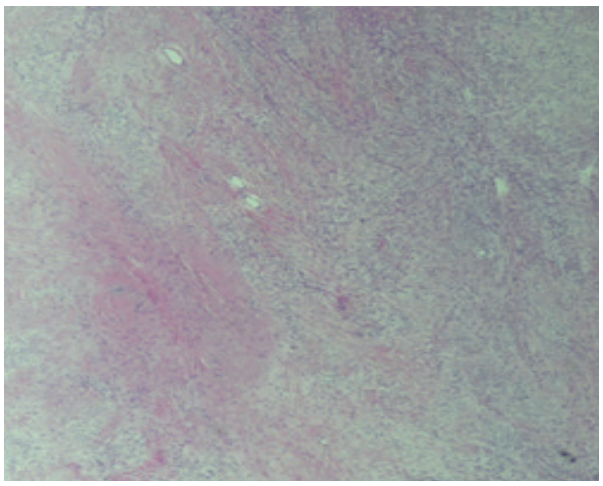

Figure 2. Spindle cells distributed in a fascicular and fusiform pattern within the collagenous stroma in sections (H&E-X20)

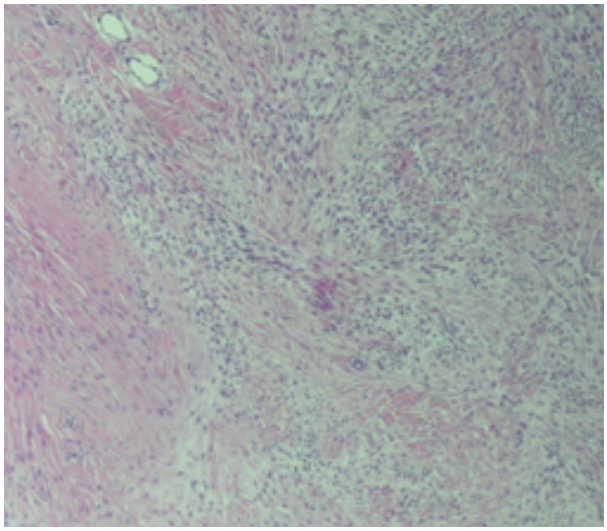

Figure 3. Spindle cells distributed in a fascicular and fusiform pattern within the collagenous stroma in sections (H&E-X40)

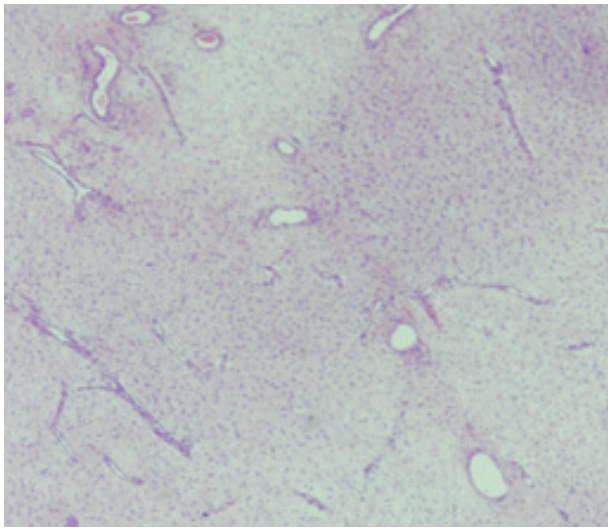

Figure 4. Round, uniform, slightly spindle-shaped cells and numerous thick and thin-walled vessels in the fibromyxoid stroma on sections (H&E-X20)

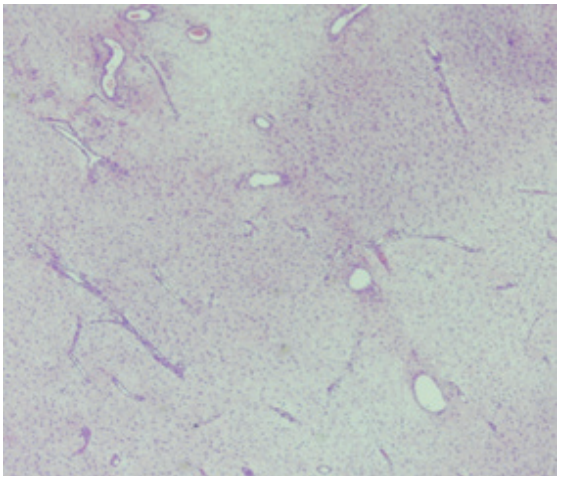

Figure 5. Widespread and strong estrogen staining of cells in the stroma (X100)

DISCUSSION

AA is a slow-growing mesenchymal tumor mostly observed in women. Although the most common location is the pelvis, cases originating from the larynx and maxilla have rarely been reported (8,9). Since they grow slowly, most patients are asymptomatic, but some of them, as in our case, may have genitourinary and gastrointestinal symptoms as a result of compressive symptoms (10). On imaging, there is an attenuated mass on CT and a characteristic's will' pattern on T2-weighted images on MRI (11). The fundamental therapeutic strategy is to perform resection with a negative surgical margin. AA is a benign tumor, but it is not always possible to achieve total excision because it has a local infiltrative aspect (12). Earlier studies have recorded local recurrence rates as 25-83%, even after full resection (13). Radiotherapy may be an option in patients who experience recurrence and are not suitable for surgery. However, a standard approach has not yet been developed for all these patients (3,14). Hormone therapy is a new treatment option for inoperable recurring cases because recurrent ER and PR expression is frequently found in AA patients. Given the inconsistent results of studies investigating the use of gonadotropin-releasing hormone agonists in patients with neoadjuvant, adjuvant, or recurrent diseases, their use as a standard treatment is not recommended (3,14,15). Because of its high local recurrence rate, AA is considered a low-grade sarcoma by some authors (16,17). This viewpoint is supported by the evidence that it can metastasize, albeit extremely infrequently (18).

FT, on the other hand, is a rare variant of sex cord-stromal tumors of the ovary and is mostly observed unilaterally (19). It is often benign, diagnosed in the early stages and rarely shows malignant features (20,21). Clinically, patients often present with pelvic pain or menometrorrhagia, but they can also be asymptomatic (22). In our patient, due to a giant retroperitoneal mass, FT was detected in the ovary during the operation and oophorectomy was performed. The primary treatment for FT is surgery, which is often effectiveas it can be detected at an early stage (20).

CONCLUSION

The neoplasms AA and FT are distinct and separate. There are no cases of these neoplasms documented in the English literature occurring on the same patient simultaneously. Despite having extremely different neoplastic mechanisms, the origin of both tumors is mesenchymal connective tissue, which may have led to their copresentation.

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