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A Case Report of Extra Ovarian Giant Fibrothecoma

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1. Abstract

We report the case of extra-ovarian fibrothecoma in a young female patient in this article. She presented with sudden-onset abdominal pain, giant abdomino-pelvic mass, massive peritoneal ascites. Serum analysis were normal except high CA-125 level. The result of USG-guided percutaneous tru-cut biopsy was reported as mesenchymal tumor, but there aren't any tumor-cell in ascite cytology. Total excision of tumor and left salpingo-oophorectomy procedure was performed. Histopathological diagnosis was reported as benign fibrothecoma. No complication were observed in postoperative period. SerumCA-125 levels returned to normal in a few days.

2. Introduction

Fibrothecomas are sex-cord stromal tumors originating from theca cells, fibroblasts and lutein cells. Fibrothecomas, which constitute 0.5-1% of ovarian tumors, and rarely be seen outside the ovary [1-3]. Most fibrothecomas are smaller than 5 cm in diameter, they are generally asymptomatic and are detected incidentally [4].

Fibrothecomas can easily be confused with malignant tumors due to high CA-125 level and ascites, and their solid and heterogeneous structure [5]. Advanced radiological examinations are needed for the differential diagnosis of these tumors, MRI for this purpose is very useful. Characteristics of fibrothecomas such as well-circumscribed borders, lack of deep invasion and any metastatic findings, and mild contrast enhancement on MRI are important in distinguishing from malign tumors. The definitive diagnosis of the disease can only be made as histopathological examination.

Although fibrothecomas are associated by some authors with the syndrome known as the triad of abdominal acid, hydrothorax and ovarian tumoral mass, defined by Meigs and Cassa in 1937. But, ascites or pleural effusion is not observed in every case of fibrothecoma. Only 10-15% of fibrothecomas occur together with peritoneal ascites. The incidence of both ascites and hydrothorax is less than 1% of fibrotechomas [6-8].

There are different opinions about the mechanism of formation of peritoneal or pleural ascites and high CA-125 levels in fibrothecomas. The most effective mechanism in acid secretion is the role of a transudative mechanism along the tumor surface. Other possible mechanisms are blockage of peritoneal lymphatics by the tumor or increased capillary permeability in the neovascular structures. CA-125 level also increases as a result of chronic irritation to the peritoneum caused by the acid formed by these mechanisms [3,4,9].

In macroscopic appearance, the surface of fibrothecomas are gray, white and yellowish in color [10]. Heterogeneous structures, where hyaline plaques and large and small calcified nodules are observed on the cross-sectional surface. In microscopy, although there is no obvious atypia or mitosis, there are widespread spindle cell with lipid-laden and pale cytoplasm. Pathological differential diagnosis should mainly be made with other sex cord-stromal tumors, steroid cell tumors and adult granulosa cell tumors. Apart from these, non-degenerate subserous stalked uterine leiomyomas and other pelvic masses are also important in differential diagnosis [11].

For surgical treatment can be done different procedures depending on the size and location of the tumor and the age of the patient. Since these tumors are benign, total excision of the mass is usually sufficient. However, in the presence of multiple tumor foci or invasion of other organs, extensive organ excisions such as salpingo-oophorectomy or hysterectomy may need to be added to the treatment. The postoperative recurrence rate in fibrothecomas is very low, and mass excision is recommended in recurrent cases [7].

3. Case Report

A 19-year-old female patient applied with complaints such as abdominal pain, bloating and dyspepsia. On physical examination, a large mass was palpated in the lower abdominal region. The patient was hospitalized and further radiological and laboratory tests were performed. In hematological examinations, hemogram, liver and kidney function tests and electrolytes were normal. Abdominal USG showed a giant abdomino-pelvic mass of 17cm in size and widespread ascites. The uterus and tubo-ovarian structures were in normal position and appearance. In laboratory tests, the CA-125 value was 1925 IU, CEA 40 IU. A crescent-shaped pelvic mass measuring 17x12x10 cm in size was observed in the posterior neighborhood of the uterus, almost completely filling the presacral distance in MRI. It was observed that there was heterogeneous contrast enhancement in the central part of the mass, T1A was heterogeneously isointense, T2A was heterogeneous iso-slightly hypointense, and there was widespread ascites in the pelvic region (Figure 1).

USG-guided percutaneous tru-cut biopsy for tissue diagnosis and cytological examination by taking fluid from the ascites in the abdomen by paracentesis were done. Ascites cytology was tumor-negative and the core-biopsy was reported as a mesanchymal tumor. No other tumor focus was detected in the patient during the whole body scan. Preoperative preparations were completed and the decision for surgical treatment was made. At the laparotomy, a white-yellow-looking, smooth-edged, hard-consistent tumoral mass was observed. There was closely adjacent to the left ovary but no direct connection with any gynecological organs, or retroperitoneal invasion (Figure 2). Both ovaries, uterine tubes and uterus were in normal position and appearance. The diameter of the ovaries was less than 3 cm. No metastatic and pathological findings were detected in other abdominal organs during exploration. Approximately 1500 cc of free fluid was aspirated from the pelvic region and the tumor was dissected easily from its retroperitoneal connections and resection was performed. Due to its close proximity to the left ovary, left salphingo-oopherectomy was performed. No significant complications were observed in the patient in the postoperative period. During the follow-up period, tumor markers and hormone levels were decrease to normal levels.

In pathological examination, heterogeneous tissues containing calcifications and hyaline plaques were observed on the outer surface of the macroscopically mixed giant tumoral mass. In microscopic sections, widespread cell proliferation with pale cytoplasm was observed between structures with round and oval nuclei. Mitotic activity and atypical cell proliferation were not observed, and the ovary and tube were observed to have normal structure. The pathological diagnosis was reported as benign fibrofibrothecoma. Immunohistochemical analysis was performed in another institution and positive staining was observed with Vimentin, Actin and PanCK, while negative staining was observed with Desmin, S100 and CD34. The proliferation index with Ki-67 was evaluated as 1%.

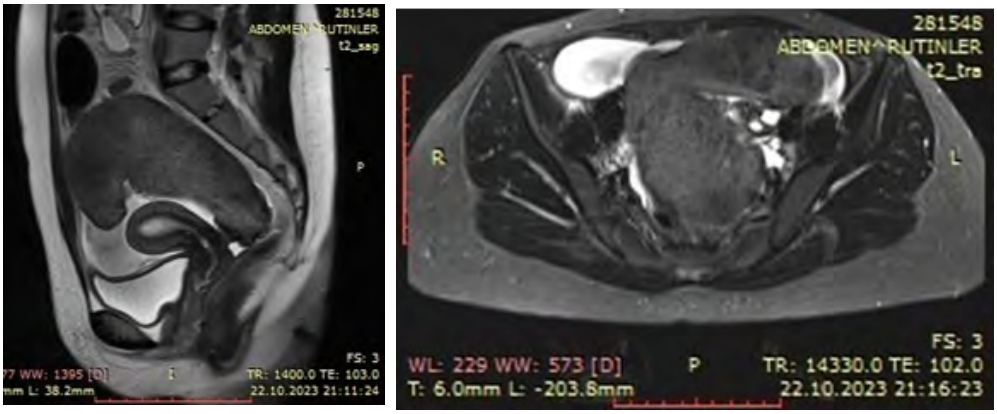

Figure 1: Abdominal MRI. A slightly contrasting, T1A heterogeneous isointense, T2A heterogeneous iso - slightly hypointense mass, measuring 17x12x10 cm, is observed in the posterior neighborhood of the uterus, completely filling the presacral distance.

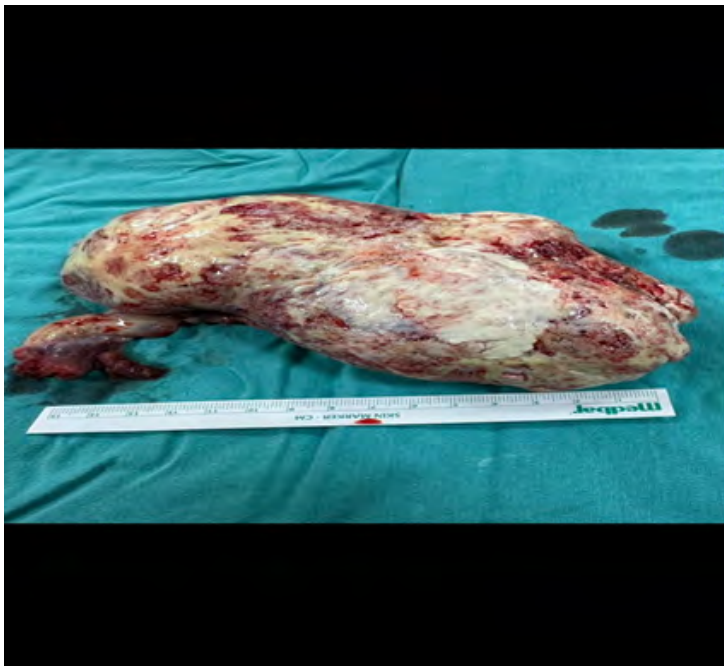

Figure 2: During the surgery, total mass resection was performed along with left salphingo-oopherectomy. Heterogeneous tissues containing calcifications and hyaline plaques are observed on the outer surface, which has regular borders, white-yellow appearance, and hard consistency in macsoscopic view.

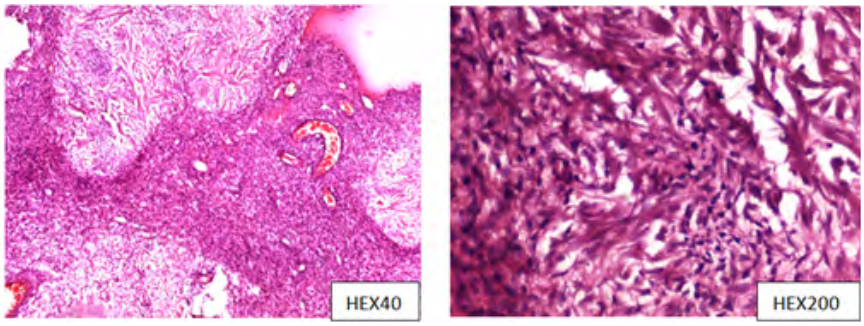

Figure 3: Microscopic appearance: Widespread proliferation of lipid-laden cells with oval and round nuclei, pale cytoplasm, and spindle or round cells with occasionally crossed fibers are observed, with no obvious cellular atypia, mitotic activity, or signs of necrosis, and the ovary and tuba are in normal structure.

4. Discussion

Fibrothecomas are rare benign sex-cord stromal tumors of the ovaries, and a small number of extra-ovarian fibrothecomas have been reported in the literature [12]. It is thought that these tumors may arise from ectopic ovarian tissue, but their exact etiology is unknown. There are two known types of ectopic ovarian tissue: (I) orthotopic ovarian tissue or accessory ovary originating from the same primordium; (II) heterotopic ovarian tissue arising away from the ovarian region. Other possibilities have been claimed to include fibrothecomas derived from Müllerian remnants or mesothelium [13]. Regardless of their origin, extraovarian fibrothecomas are difficult to distinguish histologically from their ovarian counterparts. There is no difference between them and ovarian fibrothecomas in terms of clinical findings, treatment method and prognosis.

In this study, a case of extra-ovarian giant fibrothecoma in a 19-year-old female patient with abdominal ascites and high CA-125 level is presented. The main complaint in our case was abdominal pain. An abdominal mass and peritoneal ascites were detected on USG.

Fibrothecoma masses are generally seen on USG as a hypoechoic adnexal mass that is well-circumscribed and gives a weak acoustic shadow [14]. Since the disease is usually accompanied by mild symptoms or is asymptomatic in most cases, it is diagnosed incidentally during examinations performed for other reasons. In symptomatic cases, clinical signs are like other gynecological organ tumors and the most common findings are abdominal pain, distension and a mass palpation.

Ovarian or extra-ovarian fibrothecomas may also be associated with peritoneal ascites or pleural effusion. Because of this feature,

it can often be confused with malignant tumors or associated with Meigs syndrome [15-17].

The sizes of fibrothecomas are generally reported to be between 1-5 cm, and our case is one of the largest extra-ovarian fibrothecomas in the literature [12]. Although the factors affecting the size of the tumor in fibrothecomas are not fully known, there is a parallelism between the CA-125 level and the amount of ascites and the tumor diameter [4].

The mechanism and pathogenesis of ascites in fibrothecoma are not fully known. There are different opinions on this subject. It is suggested that vaso-endothelial and fibroblast growth factor and cytokines may be effective in the transudative mechanism due to neovascularization and fibroblast activity in the tumor. And that the pressure of the tumor on the lymph vessels may increase capillary leakage and lead to acid formation [3].

One of the important findings in fibrothecomas is the observation of high CA-125 values, as in the presented patient. It is known that CA-125 level increases in liver pathologies, pelvic inflammatory diseases, gynecological malignancies and peritoneal ascites [3]. CA-125 antigen is a high molecular weight glycoprotein and is expressed by mesothelial cells of the serosal membrane in the pleura, pericardium and peritoneum. It is known that CA-125 level increases in parallel with tumor diameter and amount of ascites [9,12].

It is seen that CA-125 levels vary between 36-1848 IU/ml in cases of fibrotechomas presented in the literature. Our case has the highest CA-125 value have seen in the literature, with a CA-125 level of 1925 IU/ml [8].

It is thought that the high serum CA-125 levels seen in fibrothecoma arise from non-tumor cells and are due to chronic pleural or peritoneal irritation. Biochemical factors caused by the chronic effect of high pressure created by acid and tumor in serosal structures are held responsible [8,18].

The most important radiological examination method in the diagnosis of fibrothecomas is USG. This is very useful in the diagnosis of all abdominal mass and peritoneal fluid considering it's a non-invasive method and is widely used everywhere [19]. In cases where USG is not sufficient for differential diagnosis, CT and/or MRI should be performed. Some authors recommend that diffusion-weighted MRI be performed as a most useful radiological examination in differential diagnosis [14,17].

Fibrothecomas can often be confused with other gynecological tumors due to their rarity, high tumor markers and the presence of ascites. And therefore should be evaluated very carefully [20]. The distinction between ovarian fibrothecomas and extra-ovarian fibrothecomas is only possible by showing that the ovaries are in normal position and histological structure. The treatment of fibrotechomas is surgery. Total resection of the mass is considered sufficient due to the benign nature of the disease.

5. Conclusion

In this study, an extremely rare case of extra-ovarian fibrothecoma is presented. Cases in which unnecessary radical surgical treatments were applied have been reported because these tumors can be confused with ovarian and other gynecological malignancies [12]. To avoid unnecessary organ loss and surgical risks, detailed clinical and laboratory evaluation should be performed, and appropriate patients should be treated with fertility-preserving surgical methods [21]. Since they are benign neoplasms, lymph dissection or adjuvant chemoradiotherapy procedures are unnecessary. As a surgical treatment, resection should be performed by preserving the gynecological organs if possible, considering the location, character of the tumor, the age and social status of the patient.

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