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Retroperitoneal Inflammatory Myofibroblastic Tumor Originating from Round Ligament

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ABSTRACT:

Retroperitoneal inflammatory myofibroblastic tumor originating from round ligament

Objective: Inflammatory myofibroblastic tumor (IMT) is a rare tumor. The etiology and biological behaviour is controversial. It could be seen in many different anatomical sites, however it is commonly seen in the lungs, mesentery, genitourinary tract and retroperitoneum. Uterine round ligament is a very rare location for IMT.

Case: We report the morphological, immunohistochemical and clinical features of an IMT which is located in the retroperitoneum / uterine round ligament of a young woman in this paper.

Conclusion: It is essential to differentiate IMT from benign and malignant mimickers for providing an appropriate therapy and follow-up.

Key words: Inflammatory, myofibroblastic, retroperitoneal

ÖZET:

Round ligaman kaynaklı retroperitoneal inflamatuar myofibroblastik tümör

Amaç: İnflamatuar myofibroblastik tümör (İMT), nadir görülen bir tümördür. Etyolojisi ve biyolojik davranışı tartışmalıdır. Genellikle akciğerde, mezenterde, genitouriner traktta, retroperitonda bildirilmesine rağmen birçok farklı lokalizasyonda bulunabilir. Uterin round ligaman oldukça nadir bir lokalizasyondur.

Olgu: Genç bir hastada, retroperitonda / uterin round ligamanda yerleşmiş bir inflamatuar myofibroblastik tümör vakasını, morfolojik, immunhistokimyasal ve klinik özellikleriyle birlikte değerlendirdik.

Sonuç: Uygun tedavi ve takip için İMT'yi malign ve benign taklitçilerinden ayırmak önemlidir.

Anahtar kelimeler: İnflamatuar, myofibroblastik, retroperitoneal

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GİRİŞ

Inflammatory myofibroblastic tumor (IMT) is a rare tumor. It was previously evaluated as a heterogeneous group of reactive lesions; it is now considered a neoplasm with a specific genetic origin, locally aggressive behavior (1-3). These tumors with a potency of intermediate behavior are known to recur and rarely metastasize (2,4). Despite being reported generally in the lungs, mesentery, genitourinary tract and retroperitoneum, they may be in many different localizations (4). Because of its special histopathological features, the intraoperative pathology consultation may not be helpful for the diagnosis (1,5). The tumor is composed of myofibroblastic and fibroblastic plexiform cells and accompanying lymphocytes, plasma cells and eosinophilic leukocytes. With this histopathological view, it is differentiated by many benign and malignant lesion.

CASE REPORT

A 21-year old female patient was admitted to our obstetrics and gynecology clinic with left lower quadrant pain. Ultrasonography (USG) and magnetic

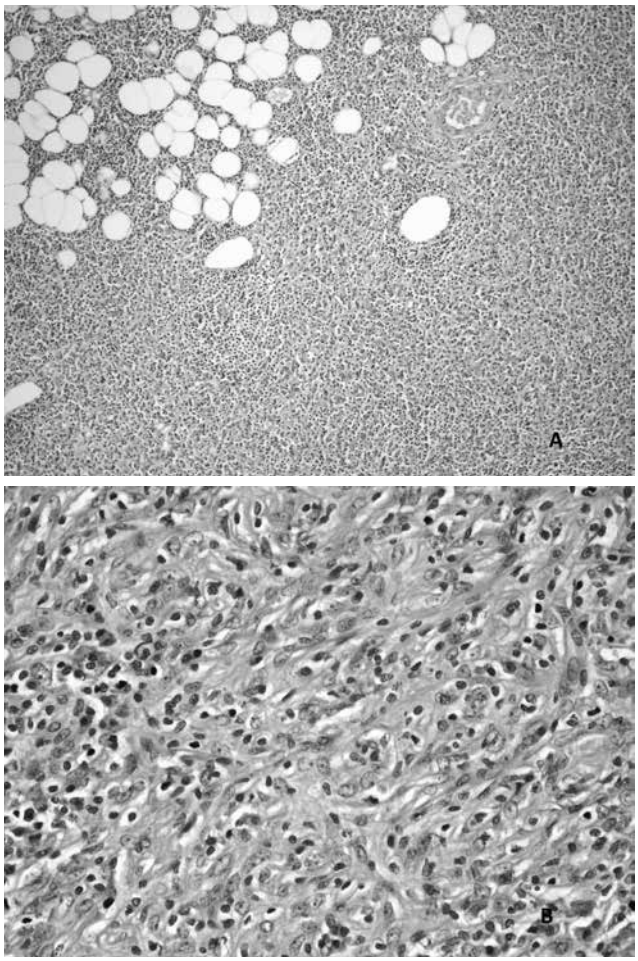

Figure-1A, 1B: Plexiform- shaped myofibroblastic cells in the myxoid stroma, occasionally inflammatory cells and peripheral fatty tissue infiltration (A; HEx100, B; HEx200).

resonance imaging (MRI) revealed a mass in the left adnexial region, and the patient was operated with a prediagnosis of myoma. The exploration revealed an extending solid mass from the the bottom of the round ligament at the left infundibulopelvic line, with a retroperitoneal location of approximately 7x7 cm which is adherent to the surounding fatty tissue. In the intraoperative pathology consultation performed, diffuse plexiform cell proliferation and occasional lymphoplasmacytic cells were observed; mild atypia was present; mitosis and necrosis were absent. With these findings, malignant/benign distinction could not be made and the final conclusion was left after the examination of paraffin blocks. The lesion was totally excised. The mass was dirty-yellow colored, with a lobulated appearance, the cross-section face dirty-white colored, occasionally hard and containing fatty areas in its periphery. In the histopathological examination, plexiform shaped cells with round or elongated nuclei and eosinophilic cytoplasm in the myxoid stroma, as well as plasmocytes, lymphocytes and eosinophil leukocytes were seen. The lesion was hypercellular, with mild to moderate atypia. There were no mitosis and necrosis (Figure-1). Immunohistochemically, vimentin, and smooth muscle actin were positive, CD117 focal positive, kaldesmon, S-100, LCA, desmin, pancytokeratin, CD 34, anaplastic lymphoma kinase (ALK), beta-catenin, DOG1 were negative in tumor cells (Figure-2). The postoperative course was unevetnful and no recurrence was observed at 10 months follow-up.

DISCUSSION

Inflammatory myofibroblastic tumor is defined as a tumor consisting of a mixture of cytologically benign plexiform-shaped myofibroblastic tumor cells and inflammtory cells. Although there have been reported cases from 3 months to 46 years, the majority of the cases are under 30 years old and usually under 14 years old (3,4,6). Our case is a young patient, at 21 years old.

This tumor has also been referred to in the past as inflammatory fibrosarcoma, inflammatory myofibrohistiocytic proliferation, inflammatory pseudotumor, omental-mesenteric myxoid hamartoma, plasma cell granuloma and plasma cell pseudotumor (6).

Histopathologically, it consists of varying quantities of benign plexiform or stellate cells with eosinophilic cytoplasm, central vesicular, oval nuclei and small nucleoli. Hyperkromosis does not exist. Mild nuclear pleomorphism may be present. Mitosis is low, usually 1-2 mitoses is seen at 10x magnification site and atypical mitosis is absent. In some cases, ganglion-like cells with abundant eosinophilic cytoplasm and distinct nucleoli were identified. Inflammatory cell infiltration, mainly consisting of lymphocytes, plasma cells, and less eosinophils and neutrophil leukocytes are observed in the stroma (1-4,6). Three basic

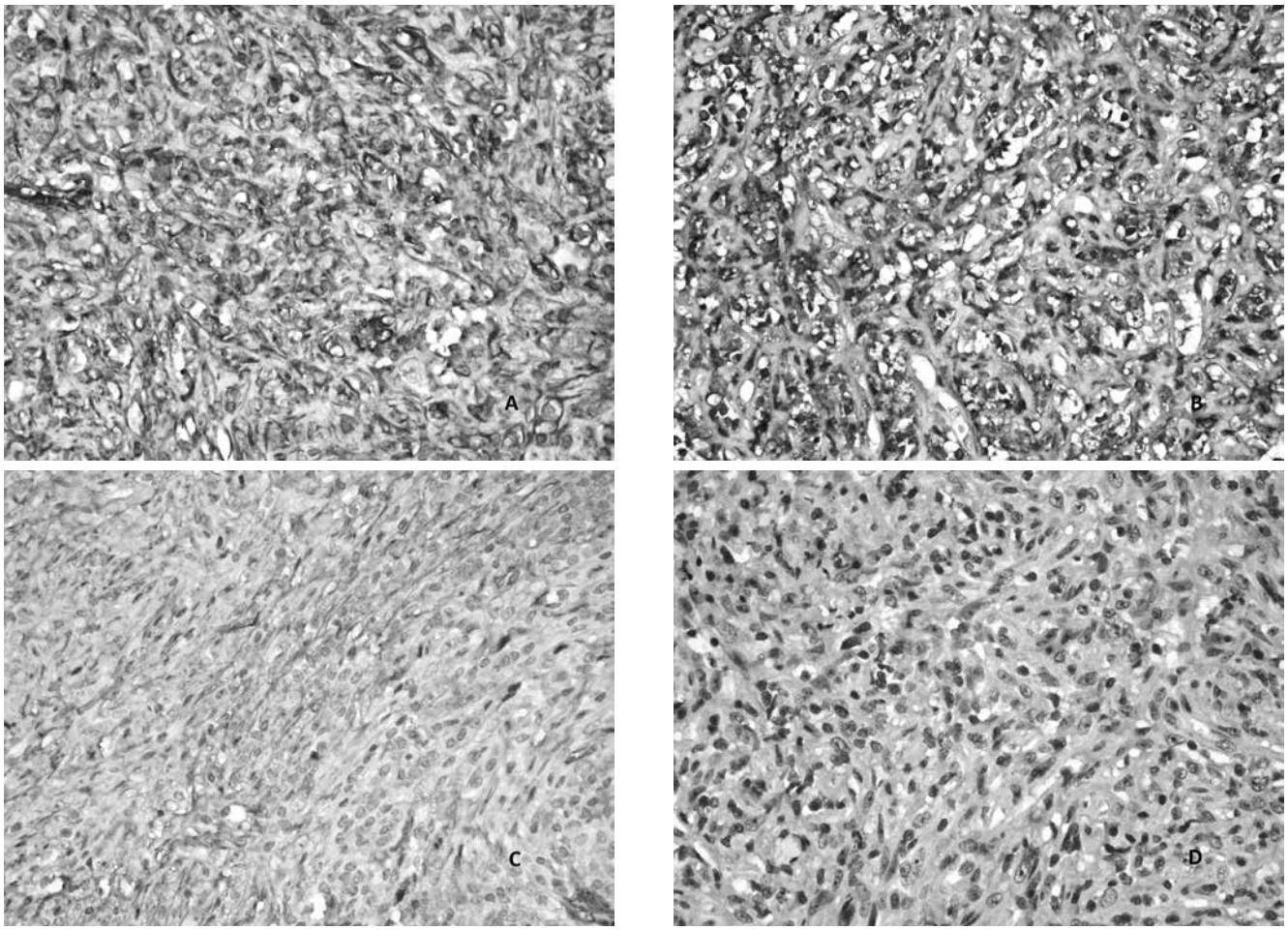

Figure-2: Vimentin (A), CD117 (B), smooth muscle actin (C) positivity, ALK (D) negativity (vimentin, CD117, smooth muscle actin, ALKx200).

histological patterns were described in inflammatory myofibroblastic tumors; a) type formed by myxoid, vascular and inflammatory areas reminiscent of nodular fasciitis, b) type consisting of a mixture of plexiform and inflammatory cells reminiscent of fibrous histiocytoma, c) type consisting of dense collagenous areas reminiscent of scar (3,6). In our case, a benign-looking tumor composed of a mixture of plexiform cells and inflammatory cells, mostly composed of lymphocytes and plasma cells. It was in the form of reminiscent of fibrous histiocytoma of the three histological patterns. A mild atypia was present. There was no hyperkromosis. Mitosis and necrosis were not observed. In the literature, especially the intra- and retroperitoneal tumors are mentioned to invade the neighboring tissues (1). In our case, peripheral fatty invaded areas were also observed.

In the literature, there are cases that were correctly diagnosed via intraoperative pathology consultation; some other cases were diagnosed as benign inflammatory processes or sarcomas and in some other cases a definitive diagnosis could not be reached (5,7). Although the case we present resembled a malignant lesion with the diffuse proliferation of plexiform cells, invasion to the surrounding fatty tissue and mild atypia, we couldn't exclude a benign lesion due to the presence of accompanying intense lymphoplasmacytic cell infiltration and the absence of mitosis and necrosis. The definite diagnosis was postponed after the examiantion of paraffin blocks.

The retroperitoneal region is one of the most

common sites for IMTs to settle, whereas they are known to be relatively rare in the female genital system (4,8). In a study, 10 IMT cases of uterine-placed were presented and a leiomyoma-like growth pattern was mentioned (8). In the literature, we couldn't find a case where the round ligament is mentioned as the settlement place.

Inflammatory myofibroblastic tumors should be seperated from many benign and malignant lesions. Although immunohistochemistry helps differential diagnosis, it is usually a distinction based on the histological pattern (4). Anaplastic lymphoma kinase positivity is important because translocation in the ALK gene and ALK expression immunohistochemically are reported. However, its negativity does not exclude IMT diagnosis, especially in adults. ALK is positive in approximately 50% of the cases and generally in young patients (2,6,8,9). Anaplastic lymphoma kinase was found to be negative, even though our case was young.

Vimentin, smooth muscle actin and calponin are generally strongly positive. Keratin and desmin positivity are present in 30% of cases. S-100 and CD117 are generally negative (1,4,6,9). VaIn our case, in comparison with these results, vimentin and smooth muscle actin were strongly positive, S-100 as well as keratin and desmin were negative. CD117 focal positivity, as an unexpected finding, resembled gastrointestinal stromal tumors and mesenteric fibromatosis (10). DOG1 used for differentiation from gastrointestinal stromal tumors and beta-catenin used for differentiation from mesenteric fibromatosis were found negative in our case and differentiated from these lesions (4,11,12). Due to its localization, it was differentiated from the myomatous lesions with its histological features and caldesmon negativity. It was differentiated from nodular fasciitis with clinicopathologic features, from solitary fibrous tumors with CD34 negativity, from plexifrom sarcomas, plexiform melanomas and sarcomatoid carcinomas with histological and immuno histochemical features.

IMT should be considered in peri-uterine and retroperitoneally-placed tumors. Differential and definitive diagnosis are crucial because of the difference in behavior and treatment.

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