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Case Report

Inflammatory myofibroblastic tumour

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ABSTRACT

Inflammatory myofibroblastic tumor (IMT) is a rarely described tumor of unknown etiology and pathogenesis. An IMT may occur in almost any part of the body, from the central nervous system to the limbs, and has nonspecific clinical manifestations. Histologically these lesions appear as an inflammatory infiltrate within a variable myofibrotic background. We here present a case of 67-year-old woman who presented with vague abdominal mass and her TAH with BSO was done in view of preoperative diagnosis of ovarian malignancy, which after histopathology came out to be IMT of pelvis.

Keywords: IMT, ALK, Vimentin, CA 125, CEA, Carcinoma ovary

INTRODUCTION

The inflammatory myofibroblastic tumor (IMT) is a rare stromal tumor with intermediate biological potential with affliction of various tissues and organs like head and neck, genitourinary tract, lung, and retroperitoneum. The etiology and pathogenesis of IMT is not much understood. Histo-morphology, ultrastructure, and immune histochemical analyses of IMT are complex and a variety of benign and malignant lesions have to be excluded. According to the WHO classification, IMTs are considered to have intermediate biological potential with a 25% rate of local recurrence and up to 5% rate of distant metastasis.[1] Surgical resection is usually the definitive treatment option, in cases of recurrence chemoradiation may be considered.[2] Herein, we describe the clinicopathologic features of a case of IMT arising in the pelvis and abdomen. The diagnosis was made by histology and immunohistochemistry analysis of the specimen.

CASE REPORT

A 67-year-old post-menopausal woman presented with complaints of abdominal pain, gradual abdominal distension and anorexia for last 4 months. On abdominal examination an abdomino-pelvic mass was felt which was corresponding to 32 weeks uterine size with restricted mobility, firm consistency and was non-tender on palpating. Findings were confirmed on bimanual examination. On per rectal examination rectal mucosa was free. All her hematological and biochemical investigations including tumour markers i.e. CA--125, Inhibin, CA 19-9, LDH and CEA were within normal limits. CECT of the abdomino-pelvic region showed a large, thin walled, multiseptated, cystic lesion of size 25×14×22 cm occupying mid and lower abdominal cavity, two focal punctuate calcification are seen in right side of lesion with multifocal deposits of enhancing soft tissues throughout the lesion. Bilateral ovaries were not seen separately. No enlarged lymph nodes were seen. PET CT revealed same abdominal pelvic complex cystic mass possibly arising from left adnexa. No definite evidence of metabolically active disease was noted anywhere in the regions of whole body surveyed. Patient was taken up for staging laparotomy in view of ovarian mass. Intra operatively, a large left sided para-ovarian cystic mass of size 30x30 cm with solid areas was seen. Mass was completely excised with total abdominal hysterectomy with bilateral salpingoopherectomy and infracolic omentectomy (Figure

1). Bilateral pelvic lymph nodes were palpated and no lymphadenopathy was found.

Final diagnosis was confirmed on histopathology which was consistent with IMT with extensive mucoid changes (Figure 2). IHC was positive for Vimentin, SMA, CD34 (vascular +), Ki-67 (low positivity 5%+) but ALK negative. Medical oncologist opinion was taken and patient was asked to remain under regular follow up. No further treatment was given to the patient. Patient is on regular follow up without any recurrence since last 16 months.

Figure 1: Large mass 30×30 cm seen intraoperatively.

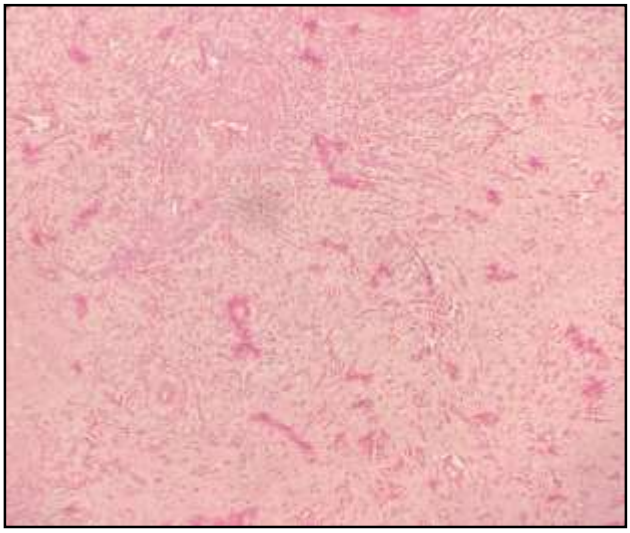

Figure 2: Histopathology of IMT with mucoid changes.

DISCUSSION

IMTs are mostly found in children and young adults but they can occur in older patients too.[3] They can be easily misdiagnosed as pseudotumor.[4] They have a high recurrence rate after excision and exhibit low metastatic potential. IMTs, are classified as intermediate-grade mesenchymal tumors which are mainly composed of spindle cells and inflammatory cell infiltration. Many features in the histopathology suggest that spindle cells which are found in these tumors are actually myofibroblasts, which is based on basis of electron microscopic studies and positive IHC markers like ALK, desmin, actin, calponin etc. Sarcomas do not exhibit ALK positivity and other inflammatory lesions do not exhibit abundant cellularity as is seen in IMT.[5] These tumors are difficult to diagnose on biopsy specimen as adequate amount of tissue is required for histopathological analysis. Because it can clinically and radiographically mimic a malignant process, the correct recognition of the lesion is important to avoid unnecessary radical surgery.

The signs and symptoms of IMTs are associated with the tumor mass effect, swelling, and local inflammation, and vary depending on the anatomical location. The tumors in the upper and lower airways can cause pain, dyspnea, obstruction, epistaxis, numbness, and headache while those of the neck may cause hoarseness. IMTs can also result in obstruction and constipation if they occur in the digestive tract, jaundice, and hepatosplenomegaly if they are localized in the liver. Vaginal bleeding may occur in lesions involving uterus. Furthermore, patients may experience general symptoms, such as fever, weight loss, and malaise, or lead to dysuria and hematuria if the urinary tract is affected.

Around 50-60% of these neoplasms express ALK, which has a good prognosis but this is uncommon in older age groups.[6] Uterine IMTs usually display reactivity to progesterone and estrogen receptors. The overall recurrence rate for IMT ranges from less than 2% in the lung, up to 25% in extrapulmonary locations while distant metastases are very uncommon (2%).[7] Imaging features, are variable and stimulate malignant lesions with aggressive features. Even with a thorough diagnostic workup, distinguishing IMT from other abdominal malignancies is challenging. The gold standard for diagnosis of IMT is histology and immunohistochemistry. Complete Surgical excision is considered the treatment of choice and adjuvant chemotherapy and/or radiation is controversial. Treatment with corticosteroids has also been tried in some cases.[8] In our case, the complete excision of mass along with TAH with BSO was done and a decision to follow up was taken. For cases where IMT cannot be completely resected or in the instance of metastatic disease, chemotherapy is used despite the lack of definitive data for efficacy.[9] Chemotherapeutic agents used are vinblastine and methotrexate. In recent years ALK-directed therapy has been shown to have a partial therapeutic effect on IMT.[10]

Prognostic factors are still undefined. Tumor size and morphologic features are not reliable prognostic factors. The presence of aneuploidy may indicate a more aggressive course. ALK positive IMTs may have higher recurrence rates, while ALK-negative IMTs seem to occur in older patients and have higher metastatic rate.[10] In our

case, the tumor had one poor predictive factor i.e., ALK-negative.

CONCLUSION

In conclusion, IMTs are a rare group of neoplasms, which have not been investigated in detail due to the difficulty of collecting a large number of cases. Thus, our knowledge about this disease remains limited. IMT has a propensity for recurrence so long-term follow-up is a crucial part of management. The tumor needs to be resected completely, as the efficacy of other treatment modalities is unclear. To determine the etiology of the disease as well as decide the optimal treatment and prognostic factors future research including a larger group of patients is needed.

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