Inflammatory myofibroblastic tumor present as an abdominal wall mass in an adult patient

ABSTRACT

Inflammatory myofibroblastic tumor of the abdominal wall is a rare soft-tissue tumor presentation in adults. A 50-year-old woman was referred with abdominal pain and a palpable mass in the left lower quadrant. Computed tomography scan and magnetic resonance investigation revealed an 8-cm heterogeneous abdominal wall mass. Tumor markers were within normal limits. Fine-needle aspiration cytology and tru-cut biopsies yielded necrotic material. A preoperative diagnosis of a resectable rhabdomyosarcoma was suggested. On exploration a tumor measuring 8 x 8 x 6 cm was resected along with the involved structures. Histopathologic examination of specimen revealed an inflammatory myofibroblastic tumor of the abdominal wall. The patient has been followed up for the last 12 months without clinical or radiographic evidence of recurrence. Inflammatory myofibroblastic tumor arising from the anterior abdominal wall in adults is an unusual manifestation of soft-tissue tumors, which can be managed by a multidisciplinary team of surgeons, oncologists, radiologists and pathologists.

KEY WORDS: Abdominal wall, adult, inflammatory myofibroblastic tumor

INTRODUCTION

Inflammatory myofibroblastic tumor (IMT) is a rare soft-tissue tumor. These tumors have a pathologic differentiation of dominant spindle cell proliferation with a variable inflammatory component. These lesions are diagnosed as masses relating to their anatomic location.1 The most commonly involved site is the lung, although there are a few case reports of extrapulmonary IMTs. Herein we report a case of an adult patient with anterior abdominal IMT presenting as an abdominal wall mass suggesting rhabdomyosarcoma, treated with surgical intervention.

CASE REPORT

A 50-year-old female patient was referred with abdominal pain and a palpable mass in the left lower quadrant without any history of trauma. Physical examination revealed a palpable mass lesion with tenderness, 8 cm in size. An abdominal computed tomography (CT) scan was performed, and an 8-cm heterogeneous abdominal wall mass lesion with suspect bowel involvement was found in the left lower quadrant [Figure 1]. No regional lymphadenopathy was noted. Tumor markers were within normal limits. Chest x-ray was normal. An abdominal magnetic resonance investigation did not show bowel involvement [Figure 2]. Colonoscopy revealed a hyperplastic polyp in the transverse colon without malignancy. Fine-needle aspiration cytology (FNAC) showed chronic inflammatory cells, benign columnar cells and scant stromal cells with no evidence of malignancy. Repeated tru-cut biopsies also yielded only necrotic material. The preoperative clinical and imaging diagnosis of a resectable rhabdomyosarcoma was suggested. The patient underwent operation. On exploration, a tumor on the anterior abdominal wall, 8 x 8 x 6 cm in size, was found with focal omentum involvement. Segmental rectus abdominis and internal abdominal oblique muscles resection with at least 2-cm clear margins macroscopically and partial omentectomy were performed. The abdominal wall defect was repaired by a prosthetic material. Histopathologic examination of specimen revealed that it was predominantly composed of cytologically bland spindle-shaped cells arranged in hyaline or myxoid stroma with an infiltration of lymphoplasmacytic cells. No cellular anaplasia was found and few mitoses were observed [Figure 3]. Immunohistochemistry revealed a strong, diffuse staining of tumor cells with myogenic marker, including smooth muscle actin and vimentin with positivity for CD68 (KP-1) but negativity for desmin and S-100 protein. Hence we confirmed a diagnosis of IMT [Figure 4]. The postoperative course was uneventful. The multidisciplinary onco-surgery team decided not to proceed with...
any adjuvant treatment. The patient has been followed up for the last 12 months without clinical or radiographic evidence of recurrence.

DISCUSSION

Inflammatory myofibroblastic tumor is a rare spindle-cell lesion of intermediate malignant potential, occurring in both pulmonary and extrapulmonary tissues.\(^1\) IMTs in the abdomen are rare entities, and most of the cases arise in the small bowel, colon mesentery, liver, spleen, retroperitoneum and other gastrointestinal sites.\(^2\) Only one pediatric case of IMT that originated from the abdominal wall has been reported.\(^3\) To our knowledge, this is the first adult case of IMT of the abdominal wall. The etiology of IMT is unknown, but likely explanations include prior inflammatory or infectious etiologies, and possibly foreign body reactions.\(^4,5\) No causative agent is evident in most cases, as in our case. Clinical symptoms of abdominal IMT are not specific to the disease and depend on localization and growth pattern of the tumor. In general, the most common initial symptoms and signs are a palpable mass, abdominal pain, weight loss or fever.\(^6\) The radiological examination findings of the tumor are not specific and are seldom diagnosed preoperatively.\(^3\) As in the current case, CT images of IMT mimic those of an invasive malignant tumor. In most cases, a definitive diagnosis is made based on the histopathological findings from either a resected tumor or a needle biopsy. IMT is histopathologically composed of myofibroblastic spindle cells, with an inflammatory cell infiltrate of plasma cells, lymphocytes and eosinophil leucocytes, and different patterns can be found within the same tumor.\(^7\) Immunohistochemistry is a valuable adjunct to light microscopic diagnosis. Vimentin is almost invariably positive in the spindle cells. Smooth muscle actin, muscle-specific actin and desmin are present in majority of the cases. CD68 (KP-1), CD30 (Ki-1), cytokeratin and p53 are positive in some cases.\(^8\) IMTs have a variable biologic

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**Figure 1:** Computed tomography scan demonstrates the tumor originating from the abdominal transversal and internal oblique muscle fascia

**Figure 2:** Magnetic resonance investigation demonstrates the tumor’s location — entrenched in the nearby structures

**Figure 3:** (A) Hypocellular lesion is composed of spindle-shaped cells and intermingling inflammatory cells (H & E, ×50); (B) the tumor cells show slight atypia without any mitotic figures. The intermingling inflammatory cells are predominantly composed of lymphocytes, histiocytes and eosinophil leucocytes (H & E, ×100)

**Figure 4:** (A) Strong and diffuse cytoplasmic reaction for alpha SMA (smooth muscle actin) in the tumor cells (alpha SMA, ×100); (B) strong and diffuse cytoplasmic reaction for vimentin in the tumor cells (vimentin, ×100); (C) numerous histiocytes intermingling with the spindle-shaped cells of the tumor and lymphocytes (CD68, ×100)
behavior that ranges from the frequently benign lesions to more aggressive variants. Predictive of aggressive behavior are cellular atypia; ganglion-like cells; necrosis; nucleolar prominence; and mitotic activity, including atypical mitotic figures expression of p53 and aneuploidy.\(^7\) An extrapulmonary location of the tumor is more frequent in childhood. Coffin \textit{et al.}\(^6\) reported 84 patients with extrapulmonary IMT, who were all children and adolescents. Adult cases are even rare in this study. Of the 53 patients under follow-up, 13 (25\%) had one or more recurrences at 1 to 24 months after initial excision, and distant metastases were not documented. According to them, an inflammatory myofibroblastic tumor is a benign, nonmetastasizing proliferation of myofibroblasts with a potential for recurrence and persistent local growth, similar in some respects to fibromatosis. Meis and Enzinger\(^8\) reported 38 cases in which 10 (37\%) of 27 patients on follow-up were found to develop at least one local recurrence, with a median of 5 months after initial excision. Three (11\%) patients had histologically proven lung and brain metastases. Spontaneous regression has also been reported in some cases of IMT.\(^8\) In general, the primary therapeutic approach is surgery for complete resection if the anatomic location is amenable. Inadequate resection has been shown to be a risk factor for recurrence. For incomplete resection, adjuvant approaches using corticosteroids, COX-II inhibitors or chemotherapy and radiation have been attempted with limited success.\(^6\)

In conclusion, inflammatory myofibroblastic tumors of the abdominal wall are rare entities in adults. Although their proper preoperative diagnosis is troublesome and they are difficult to differentiate clinically and radiologically from other soft-tissue tumors of the abdominal wall, accurate multidisciplinary investigation including clinical examination, histopathology and radiology may guide exact diagnosis. Successful management can be achieved by adequate resection of the tumor along with the involved structures.

REFERENCES


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