Pelvic inflammatory myofibroblastic tumor mimicking a rectal cancer

Lídia Roque-Ramos1, António P. Matos2, Pedro Pinto-Marques1, Gabriela Machado3, Joana Nogueira1 and Miguel Ramalho2

1Gastroenterology, 2Radiology, 3Surgery, and 4Pathology Departments. Hospital Garcia de Orta, Almada, Portugal

ABSTRACT

We report a case of a 50-year-old woman who presented to the emergency department with large bowel obstruction and anemia. The initial imaging study suggested an inoperable rectal tumor with involvement of surrounding structures. In this paper, we discuss the diagnostic work-up of this patient with a diagnosis of pelvic/perirectal inflammatory myofibroblastic tumor (IMT). IMT is a rare tumor with intermediate malignant potential that frequently mimics clinical and imaging features of malignancy. Additionally, to the best of our knowledge, this is the first case of a pelvic IMT that regressed without surgical excision.

Key words: Inflammatory myofibroblastic tumor. Rectal cancer. Endoscopic ultrasound. Magnetic resonance imaging.

INTRODUCTION

We report a case of a pelvic/perirectal inflammatory myofibroblastic (IMT) in which the initial imaging study suggested an inoperable rectal cancer and illustrate the challenges of pre-operative diagnosis. In addition, we present the endoscopic ultrasound and fine needle aspiration findings of this tumor.

IMTs are rare mesenchymal tumors that can involve virtually any organ, with the lung being the most frequent location (1). Due to its paucity and misuse of the term “inflammatory pseudotumor” as synonym for IMTs, its true prevalence and incidence are unknown (2). According to the World Health Organization classification IMTs are considered to have intermediate biological potential due to the 25% rate of local recurrence and up to 5% of distant metastasis (3). Surgical resection is usually the definite treatment option, in rare cases complemented with chemotherapy and/or radiation (4).

CASE REPORT

A 50-year-old woman presented to the emergency department with lower abdominal pain, fever, anorexia, progressive constipation and loss of 11 kg for one month. She had a history of depression and iron deficient chronic anemia and denied smoking and drinking habits. The patient was medicated with estazolam and oral iron. There was no family history of cancer.

On physical examination, lower abdomen tenderness and abdominal distension were noted. Gynecological examination revealed extrinsic bulging of both anterior and posterior vaginal sacs, which appeared hard and painful, with no evidence of mucosal lesions.

The laboratory panel was remarkable for microcytic anemia (hemoglobin 9.4 g/dl, reference 11.5-18); low mean corpuscular volume (69.4 fl, reference 76-96), elevated white blood cell (19,800/mm3, reference 4-11,000, 84% neutrophils) and platelet (924,000/mm3, reference 140-400,000) counts and a C-reactive protein of 23.3 mg/dl (reference < 0.2). The renal function was within the normal limits. There was no elevation of tumor markers, including carcinoembryonic antigen and carbohydrate antigen 19.9.

Contrast-enhanced abdominopelvic computed tomography revealed an obstructive transmural rectal tumor, centered at the right rectum wall, extending anteriorly to the mesorectum and with ill-defined boundaries with the uterus and the vaginal dome (Fig. 1).

Fibrosigmoidoscopy revealed a rectal stenosis with normal underlying mucosa and passable with moderate pressure. A pelvic magnetic resonance imaging was ordered to further characterize the local extent of the tumor and showed a transmural rectal tumor with 9.5 cm length.
The tumor extensively contacted the posterior wall of the vagina and uterus, ovaries and sacrum, suggesting direct invasion of the surrounding structures. Lymphadenopathies were observed in the mesorectum and at the right internal iliac chain (Fig. 2). Subsequent chest computed tomography was unremarkable.

Considering the presumed diagnosis of a locally advanced rectal tumor and the symptomatic bowel obstruction, an emergent derivative colostomy was performed. Intra-operatively, a rectal tumor adhering to the uterus and pre-sacral area was observed.

To further characterize this rectal stenosis, an endoscopic ultrasound (Olympus UCT140 AL5) was performed. The normal five-layered appearance of the rectal wall was preserved while an ill-defined extrinsic hypoechic mass infiltrated the perirectal fat. These features were suspicious for extrinsic tumor infiltration and fine needle aspiration with a 19 G procore needle was performed (Fig. 3).
cellblock obtained muscle cells and stroma with an exuberant inflammatory cell infiltrate composed of B and T lymphocytes and histiocytes, consistent with an inflammatory process.

During the follow-up, the patient maintained abdominal pain, low-grade fever and elevated inflammatory markers, despite being medicated with intravenous cefuroxime and ciprofloxacin. Since a definitive diagnosis was lacking a second-look surgery was performed, which revealed a whitish infiltrating mass extending from the pre-sacral region to the uterus, involving the ovary, rectum and ileo-cecal appendix. Hysterectomy, bilateral salpingo-oophorectomy and appendectomy were performed. Surgical macrobiopsies revealed a spindle cell proliferation with numerous inflammatory cells, predominantly lymphocytes and plasma cells, in an eosinophilic collagenized stroma. No atypia, cellular mitoses or necrosis was observed. Immunohistochemical staining for smooth muscle actin antibody was positive in the spindle cells. There was no expression of anaplastic lymphoma kinase (ALK), CD34, CD117 or S100 (Fig. 4). These findings were consistent with inflammatory myofibroblastic tumor in the cellular phase. Additionally, a chronic right-sided salpingitis with acute inflammatory infiltrate was identified and the sample was negative for fungi and acid fast bacilli.

Shortly after the procedure the patient became asymptomatic with gradual normalization of laboratory parameters, including hemoglobin (14 g/dl), leucocytes (5,700/mm³), platelets (291,000/mm³) and C-reactive protein (0.1 mg/dl). Due to the favorable course of the disease and histopathological findings, the clinical decision was to follow up without further therapy. Bowel reconstruction was successfully performed. The magnetic resonance imaging exam repeated after one year showed complete regression of the pelvic mass (Fig. 5).

DISCUSSION

Inflammatory pseudotumor (IPT) is a term first used in 1985 and describes the histological appearance of a bland spindle cell proliferation with prominent inflammatory infiltrate. Over the last decades it has become clear that this morphological terminology was broad and included several clinic-pathological entities with different cell lineage and biological behavior. For this reason some authors advocate dividing IPT in secondary, when these lesions are believed to represent a reparative reaction to infection, inflammation or trauma, and primary or idiopathic when the etiology is unknown (5). Idiopathic IPT probably represent true IMT, a neoplastic entity that emerged from the heterogeneous group of IPT for its distinct clinical, pathological and molecular features (2). IMT have an intermediate biological behavior and should be distinguished from benign reactive secondary IPT and malignant tumors with a prominent inflammatory component such as leiomyosarcomas. Although IMT are more frequent in children and adolescents they can occur in older patients such as seen in our patient. Virtually any organ can be involved, with the most common locations being the lung, abdominopelvic region and retroperitoneum (2).

Patients may be asymptomatic, particularly when the tumor is a small solitary mass; present with local mass effects such as abdominal pain, vomiting, constipation, chest pain or cough; and/or have systemic manifestations, including fever, weight loss and malaise, reported in 15-30% of cases. Laboratory work-up reflects an inflammatory process and includes anemia, thrombo-
cytosis, leucocytosis, increased C-reactive protein and sedimentation rate, hypoalbuminemia and polyclonal hypergammaglobulinemia (1,2,5). Imaging features, are variable and nonspecific and may mimic malignant processes with aggressive features such as mural infiltration and extravisceral extension (1,6). Indeed, the reported patient presented with constitutional symptoms and bowel obstruction, as well as increased serum inflammation markers, anemia and an apparent rectal tumor invading surrounding organs on both computed tomography and magnetic resonance imaging.

As in most malignancies the definitive diagnosis of IMT relies on histology (6). There are only 3 cases of abdominal IMT reported where endoscopic ultrasound was performed (7-9). In one case, the IMT was located in the spleen (10), and fine needle aspiration allowed definite histological diagnosis. In the remaining two cases, gastric hypoechoic subepithelial lesions were evaluated with endoscopic ultrasound without fine needle aspiration and the final diagnosis was obtained after gastrectomy (7) and endoscopic submucosal resection (8). In our case, the endoscopic ultrasound evaluation demonstrated a preserved five-layer rectal wall structure, suggesting an extramural process. The fine needle aspiration pointed towards an entity characterized by the presence of abundant inflammatory cells and stroma. However, since soft tissue sarcomas can be accompanied by an inflammatory reaction and the patient had an exuberant clinical presentation, malignancy couldn’t be confidently ruled out and the final diagnosis was obtained after intra-operative macrobiopsies.

Surgical excision is considered the treatment of choice (9) and adjuvant therapy with chemotherapy and/or radiation is controversial (4). In our case the infiltrating and diffuse nature of the pelvic mass precluded a complete excision and a decision to follow-up was taken. Zhao et al. recently published a literature review with 38 patients with abdominal IMT that regressed without surgery, 31% of whom were treated with NSAIDs, steroids or antibiotics. The liver was the most common location (11). Interestingly, shortly after surgery, our patient became asymptomatic and the serum inflammatory markers normalized. The magnetic resonance imaging performed one year later showed complete resolution of the pelvic mass. Regression without surgical tumor resection is uncommon (1). To the best of our knowledge this is the first case of a pelvic IMT that regressed without surgery, with short-term intravenous antibiotics being the only therapies given considering the suspicious of infection.

Recurrence rates range from less than 2% in the lung up to 25% in extrapulmonary locations. Distant metastases are rare (< 5%) and the most common sites are lung, brain, liver and bone (2). Prognostic factors are still undefined. Tumor size and morphologic features are not reliable prognostic factors, however aneuploidy may indicate a more aggressive course (3). Rearrangements involving the ALK locus on chromosome 2p23 have been demonstrated in approximately 50% of IMTs, based on immunohistochemical analysis. ALK-positive IMTs may have higher recurrence rates, while ALK-negative IMTs seem to occur in older patients and have higher metastatic rate (2,12). In our case, the tumor had two poor predictive factors, including the infiltrating characteristics at presentation and negativity for ALK; nevertheless, regression without surgical resection was observed. At 2 years follow-up the patient remains clinically well and with normal inflammatory parameters.

Fig. 5. Follow-up pelvic magnetic resonance imaging 2 years after presentation. A. Axial T2-weighted image shows a resolution of the previously described tumor, with minimal thickening of the rectal wall. B. Sagittal T2-weighted image confirms the macroscopic resolution of the tumor.
REFERENCES


