CASE REPORT – OPEN ACCESS

International Journal of Surgery Case Reports 26 (2016) 150–153

Contents lists available at ScienceDirect

International Journal of Surgery Case Reports

journal homepage: www.casereports.com

Diagnostic and therapeutic dilemmas in intra-abdominal desmoid tumors: A case report and literature review

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ARTICLE INFO

Article history:
Received 5 July 2016
Accepted 25 July 2016
Available online 28 July 2016

Keywords:
Desmoid tumor
Surgery
Immunohistochemistry
Mesenteric fibromatosis
Case report

ABSTRACT

INTRODUCTION: Intra-abdominal desmoid tumors (DTs) are a rare and anatomically diverse group of locally-aggressive, benign neoplasms. They are often difficult to diagnose, even in patients who possess risk factors for the disease. Even after a diagnosis has been reached, the optimal therapy is often not well-defined.

PRESENTATION OF CASE: The case discussed of a 33-year old male with a giant intra-abdominal desmoid is an example of both the diagnostic and therapeutic dilemmas that arise when confronted with a patient with a DT. Initial confusion over diagnosis led to ineffective therapy, but once the correct diagnosis was made, the patient went on to definitive surgical resection.

DISCUSSION: The differential diagnosis of DTs is broad, and the diagnosis is often delayed due to nonspecific presentations. Immunohistochemistry is crucial in the accurate histological diagnosis, which guides treatment. Chemotherapy and radiation have a role in the management of both primary and recurrent lesions, but surgical resection remains the cornerstone of treatment.

CONCLUSION: DTs present a clinical challenge in their diagnosis and management, and despite providing standard medical and surgical treatment, recurrence rates are high and continued surveillance is crucial.

1. Introduction

Intra-abdominal desmoid tumors (DTs), are benign locally-aggressive mesenchymal neoplasms that lack the potential for metastasis. DTs are rare as they account for only 0.03% of all neoplasms and less than 3% of all soft tissue tumors [1,2]. Approximately 80% of DTs are sporadic and can occur anywhere in the body, but the balance are associated with pregnancy, trauma and genetic syndromes such as familial adenomatous polyposis (FAP) and are more commonly intra-abdominal [3]. In the absence of these risk factors, intra-abdominal DTs present a diagnostic and therapeutic dilemma for the surgeon.

We present the case of a 33-year-old otherwise healthy male with a giant intra-abdominal DT that masqueraded as a gastrointestinal stromal tumor (GIST) until ultimate diagnosis at the time of surgical resection.

2. Case report

A 33-year-old male without medical, surgical or significant family history presented with a complaint of several months of vague, diffuse abdominal pain that was initially attributed to constipation and treated in the outpatient setting. The patient was referred to a surgeon after a computed tomography (CT) scan was obtained due to continued symptoms. A large soft tissue mass in the mid abdomen extending to the upper pelvis was discovered (Fig. 1). The mass, which consisted of hypodense regions and multiple vessels, measured $15.7 \times 28.6 \times 24.2$ cm. The mass was displacing bowel loops, but was not causing obstruction or other mass effect.

In order to inform initial treatment, an ultrasound-guided core needle biopsy was performed. Histologically, neoplastic spindle cells were identified that were moderately positive on immunohistochemical staining for muscle specific actin, S-100 and CD117 and negative for cytokeratin AE1/AE3, desmin and CD34. Cytology revealed no clonal proliferation of lymphocytes.

Given the patient's clinical presentation and histopathology, there was high suspicion that the lesion represented a large GIST. The patient began neoadjuvant treatment with imatinib. After four months, the patient's pain, anorexia and weight loss did not resolve. A repeat CT scan revealed that the mass had increased significantly

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http://dx.doi.org/10.1016/j.ijscr.2016.07.044

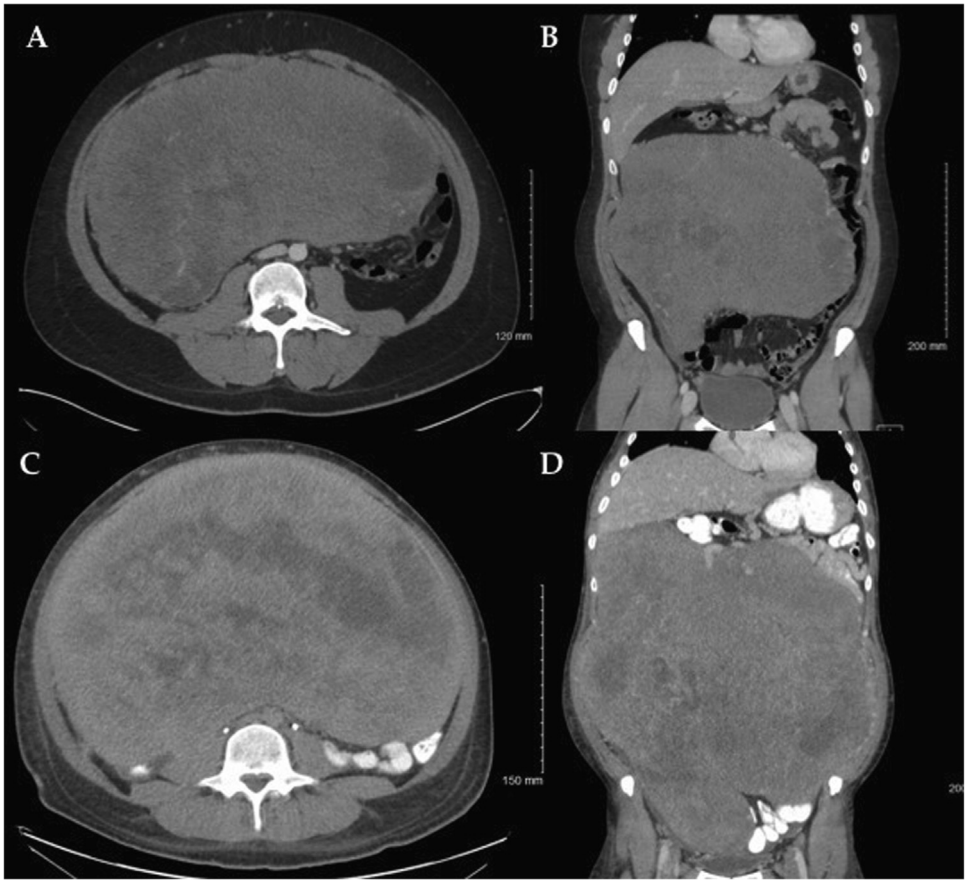

Fig. 1. Computed tomography (CT) scans. At initial presentation (A and B), the soft tissue mass was 15.7 × 28.6 × 24.2 cm. After a trial of four months of imatinib, the mass had grown to a size of 20 × 37 × 32 cm and demonstrated mass effect on surround organs (C and D).

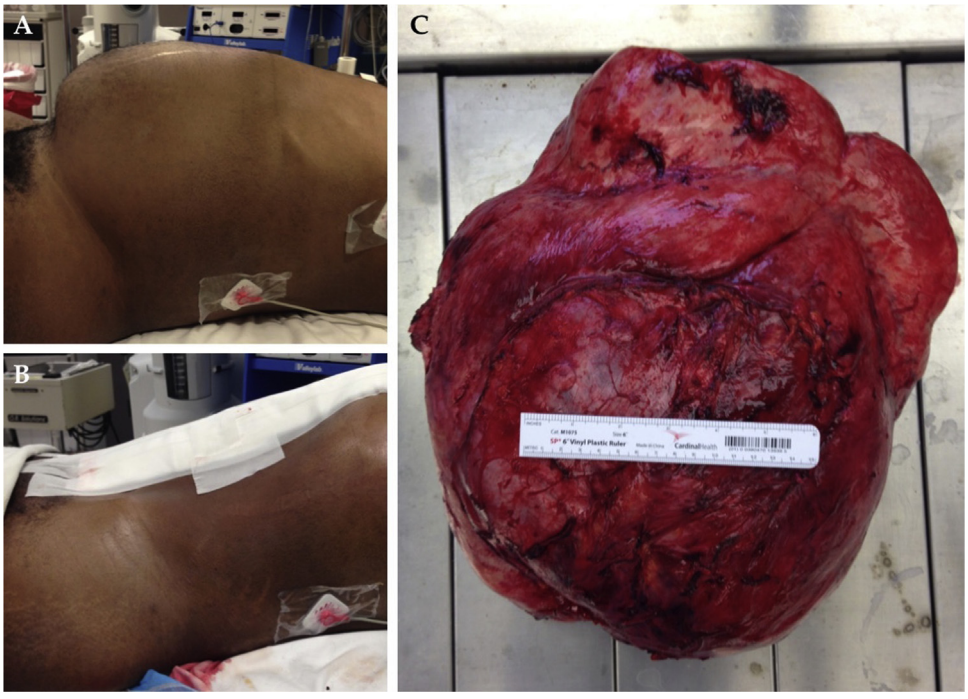

Fig. 2. Gross images. The patient's abdominal distension improved when compared preoperatively (A) and postoperatively (B). The resection specimen (C) was 45 × 33 × 23 cm, and it is pictured here immediately after resection with a 15 cm ruler.