RETROPERITONEAL INFLAMMATORY MYOFIBROBLASTIC TUMOR: A CASE REPORT, A DIFFICULT APPROACH

Canhoto C, University of Coimbra Hospitals, Portugal
Azevedo F, University of Coimbra Hospitals, Portugal
Baptista H, University of Coimbra Hospitals, Portugal
Pinho A, University of Coimbra Hospitals, Portugal
Oliveira P, University of Coimbra Hospitals, Portugal
Carvalho H, University of Coimbra Hospitals, Portugal

Corresponding author: Canhoto C, Surgery Department, University of Coimbra Hospitals, Portugal. E-mail: carolinacanhoto@gmail.com


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ABSTRACT

Inflammatory myofibroblastic tumor (IMT) is a rare neoplasm and with different etiologies and behavior. It occurs more often in children and the prognosis varies a lot accordingly to the histological characteristics. We present a clinical report of a 19-year-old female with a retroperitoneal mass compatible with an IMT that was resected surgically.

Keywords: Retroperitoneal, Children, Tumor.

INTRODUCTION

Inflammatory myofibroblastic tumor (IMT) is a rare neoplasm and can pose a challenge to diagnose and treat correctly (Pungpapong et al. 2004). In recent years the outlook on these tumors changed: from benign neoplasm to undefined behavior leaning to malignant tumors (Coffin et al., 1998). There are three major histological subtypes:
Desmoid/scar tissue like, fibrous histiocytoma-like and nodular fasciitis-like type (Freeman et al. 2004). Although morphologically similar they present a different spectrum of local recurrence, metastatic potential, and different etiologies (Pungpapong et al., 2004; Coffin et al., 1998; Freeman et al., 2004). The most common site of appearance is the lung, and rarely do they appear in the retroperitoneal space.

**CASE PRESENTATION**

A 19-year-old female presented to the emergency department with abdominal pain, localized in the right iliac fossa for several days and recent aggravation. She denied any accompanying symptoms like fever, nausea, anorexia. No previously relevant medical history. On physical examination, she had a palpable mass in the right iliac fossa, tender to the touch and an audible fremitus but no signs of peritoneal reaction. Laboratory studies were inconclusive, with no altered inflammatory markers. An abdominal ultrasound revealed a heterogeneous mass mostly isoecoid with 10 x 4 x 3 cm and showed to be highly vascularized on Doppler ultrasound.

An abdominal Computed Tomography (CT) was performed that revealed a heterogeneous retroperitoneal mass, probably in relation to central tumor necrosis, that displaced the ureter, right colon, and inferior vena cava compression. At this point, our diagnostic hypothesis was between a sarcoma and a low-grade lymphoma or a neurogenic tumor (such as paraganglioma). We performed a Magnetic Resonance Image (MRI) that revealed the same necrotic center and the displacement of the normal anatomy that had been seen on the CT scan (Figure 1). Complete staging, including PET-CT, of the lesion was performed and there were no apparent metastatic lesions anywhere else.

![RMI Image](image.png)

**FIGURE 1**

RMI IMAGE. THE ARROW POINTS TO THE LESION THAT COMPRESSES THE INFERIOR VENA CAVA AND THE X SHOWS CENTRAL NECROSIS

The patient was submitted to a surgical biopsy of the lesion, there was no window to perform it in a non-invasive way and also it carried a high risk of spreading in case of a sarcoma diagnose. The biopsy was inconclusive and only revealed lymphatic tissue and reactive alterations. On a multidisciplinary meeting was decided to surgical explore the mass
and evaluate the resectability. The patient was submitted to a median laparotomy, total mobilization of the right colon in a Cattel-Braasch maneuver fashion and we had access to the retroperitoneal space and the large vessels iliac vessels, inferior vena cava, and the aorta. Immediately we figured that the ureter was crossing over the mass but with no apparent invasion. The ureter was completely dissected and isolated from the mass. Afterward, we approach the inferior vena cava that looked like it had a small fraction of invasion (Figure 2).

![FIGURE 2](image1)

**FIGURE 2**
THE LESION IN PLACE COMPRESSING THE INFERIOR VENA CAVA

The inferior vena cava was then partially clamped and resection was completed with a vascular suture of a 5mm orifice of the inferior vena cava (Figure 3).

![FIGURE 3](image2)

**FIGURE 3**
FINAL LOOK AFTER THE RESECTION

In blue loops we have the inferior vena cava and the right renal vein. In red, more medially, we have the aorta (superior mesenteric artery visible) and the common iliac artery and at the top the renal artery. In a white loop we have the ureter. There were no postoperative complications and the patient was discharged on day 10 due to paralytic ileus after the resection. Histopathologic revealed an inflammatory myofibroblastic tumor with a capsule, desmoid-like cells (spindle-like) and without invasion of the surgical margin and no cellular atypia. Immunohistochemistry showed a high positivity for actine. In conclusion an IMT with no cellular atypia. Since the surgery, the patient has maintained surveillance and no local recurrence were documented, through MRI, since the resection (5-year follow-up).
DISCUSSION

This case illustrates the struggle to diagnose IMT as the course of action was not linear. Radiologically it can mimic several kinds of tumors with different therapies. The therapeutic approach for IMT is not standardized and there is no strong consensus but the surgical approach is usually the preferred treatment. There are some cases described of a partial response to chemotherapy mainly in unresectable cases (Imperato et al., 1986; Dishop et al., 2003). In this particular case, surgical approach was not easy due to the ureter placement and the displacement of inferior vena cava which required a vascular control proximal and distal to the lesion. Being a resectable lesion and with no apparent local invasion or distant spread of the disease surgical resection is the best therapeutic course of action (Tambo et al., 2003).

CONCLUSION

IMT are typically childhood tumors and is a rare condition in adult life. Although it's important to keep in mind that they do happen and in several different locations and should be a part of a differential diagnose and when feasible surgical resection should be offered to these patients. In this case we have a long disease-free survival, 5-years of follow-up, accordingly with the expected good prognosis to these tumors.

REFERENCES


