Intra-abdominal inflammatory myofibroblastic pseudotumor: case report and review of the literature

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Introduction

Inflammatory myofibroblastic pseudotumors (IMP) are very rare tumors characterized by unpredictable clinical behavior. They arise in soft tissues of almost every organ and the most common site is the lung. Over 200 cases of inflammatory myofibroblastic pseudotumor of the lung have been described in literature. Intra-abdominal IMP are very rare.

We describe a case of intra-abdominal IMP in a boy of 15 years old who presented symptoms and signs of acute appendicitis. Exploratory laparotomy revealed a mass in the peritoneal cavity. The mass was removed. The histological examination showed that it was an IMP. Surgical treatment was the only therapy. Six months after the surgical operation the patient has no sign of illness.

Case report

A 15 year old boy was referred to our emergency room with abdominal pain, mainly localized in the right iliac fossa. There was no history of nausea, vomiting, weight loss and fever. One year before the patient experienced generalized abdominal pain which disappeared without therapy. He had no past history of serious illnesses or surgical operations.

Physical examination of the abdomen revealed tenderness in right iliac fossa, without signs of peritonitis. Laboratory tests showed leukocytosis with neutrophilia. Symptoms, physical examination and laboratory tests led us to suspect an acute appendicitis.

The patient underwent an exploratory celiotomy with a para-
rectal laparotomy. Intraoperative assessment revealed a mass measuring 18 x 12 cm, in the pelvis, arising from caecum and appendix, adherent to bladder, sigmoid colon, mesorectum, without infiltration into these organs. There were 300 cc of peritoneal effusion in peritoneal cavity. No regional lymphadenopathy or liver lesions was identified. The abdominal mass was entirely removed.

The patient recovered normally and seven days after the surgical operation underwent a computed tomography of abdomen which demonstrated no residue of the mass.

Gross examination of the specimen showed a 18x12cm circumscribed mass by a weight of 350 g. The cut-surface of the lesion was yellowish with a soft consistency. On histopathologic examination the tumor consisted of spindle cells, lymphocytes, inflammatory infiltrates in a fibrillar matrix. Immunohistochemical staining was positive for vimentin and CD-68. The proliferative activity was low (Ki67 < 5%).

The patient was asymptomatic at follow up evaluation 6 months later.

Discussion

Etiology of inflammatory myofibroblastic pseudotumor (IMP) is still unknown. According to its histological aspect, it could be an inflammatory reaction to microorganisms such as E.Coli, Epstein-Barr virus, Cytomegalovirus, or an autoimmune reaction (2, 11-14). Interleukin-6 seems to be involved in its pathogenesis (4).

IMP appears as a pseudosarcomatous lesion of admixed inflammatory infiltrates with myofibroblastic spindle cells (4), with a positive stain for smooth muscle actin and vimentin (15).

Clinical presentation is variable according to the anatomic location, the size and the extension. Intra-abdominal pseudotumors sometimes present themselves as masses accidentally discovered by imaging investigation; they can cause abdominal pain, malaise, weight loss, fever, anorexia, nausea and vomiting, palpable abdominal mass, gastrointestinal bleeding, acute abdomen due to rupture or bleeding of the mass (4, 6, 10, 13, 16).

Because of the rarity of these lesions and varied clinical pictures, diagnosis is often very difficult. MRI, CT, ultrasonography and angiography show a malignant-like lesion (2, 10). Only biopsy can confirm the diagnosis of IMP. CT or ultrasonography guided biopsy and needle biopsy (1, 2, 3, 10, 12). Diagnosis is conclusive by histological examination the mass after surgical excision (10).

IMP are characterized histologically by spindle cells within a collagenous or myxoid matrix; inflammatory infiltrates mainly consisting of lymphocytes, histiocytes, plasmacells and eosinophils (4, 2, 12).

Despite the malignant-like aspect of IMP, they usually have a benign clinical behaviour. Prognosis is correlated first of all to the size of the tumor and its extension beyond the confines of the organ in which it arises (17). Prognosis is usually excellent after surgical resection for IMP localized in liver and lung (1, 8, 9). Intra-abdominal and retroperitoneal masses often have a more aggressive behaviour characterized by local recurrences and metastasis (4, 10). Combination of atypia, ganglion-like cells, expression of p53 may indicate an aggressive behaviour (4).

Treatment depends on the anatomic location and the size of the tumor. Most extrapulmonary masses are successfully treated by surgical resection and a long term follow up is mandatory because of the significant rate of local recurrences (10). For inoperable tumors and recurrences radiotherapy and chemotherapy with cisplatin, doxorubicin and methotrexate are sometimes indicated (2, 4).

Surgery remains the main treatment of these tumors and it is often the only method for excluding a malignant nature of the lesion.

Conclusion

IMP are very rare lesions and they can represent an unexpected intra-operative finding at emergency surgery. It is important to know this kind of tumor because of the need for a radical excision and a long term follow up.

Generally IMP have a good outcome but sometimes they can present local recurrences and metastasis.

Surgery plays a predominant role in treatment of these tumors.

References

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