Abdominal inflammatory myofibroblastic tumour

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Inflammatory myofibroblastic tumours (IMT) are inflammatory pseudotumours. They are rare lesions characterised histologically by fibroblast proliferation associated with reactive inflammatory cells, mimicking a malignant process. Described by Brunn for the first time in 1939 in the lung, one of their preferred sites, they can affect any organ.

Observation

A 20-year-old female patient consulted for an impaired general condition, nocturnal sweating, and exertional dyspnea. Clinical examination revealed a hypogastric mass. Laboratory tests revealed iron deficiency anaemia. An abdominopelvic CT scan (Fig. 1) showed a rounded well-circumscribed pelvic lesion, which was spontaneously hypodense relative to the muscles. There was intense peripheral contrast enhancement, due to the presence of highly vascularised tissue. The central area remained hypodense in the venous phase (120 s) suggesting a necrotic or fibrous area. There were no locoregional lymphadenopathies. A single large, well-circumscribed lesion in a young patient, with no lymphadenopathy or organomegaly, suggests a benign tumour. Complete surgical ablation confirmed the presence of a lesion, developing from the greater omentum, measuring 7 cm, with many congested surface vessels and a central fibro-necrotic area (Fig. 2). Histological examination found myofibroblastic spindle cells associated with inflammatory cell infiltrates (Fig. 3). ALK immunohistochemical staining was positive. The diagnosis was thus of an IMT.

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Discussion

IMTs are rare benign lesions, with ubiquitous development and unknown aetiology [1]. These lesions have been reported with various names: inflammatory pseudotumour, plasma cell granuloma, fibroxanthoma, pseudo-lymphoma, histiocytoma, xanthomatous pseudotumour, pseudosarcomatous fibromyxoid tumour, and even inflammatory fibrosarcoma [1,2]. The most prevalent term in current use in the literature is inflammatory myofibroblastic tumour.

Pseudotumours can be divided into two particular types [3]:
• IMTs, which may occur at any age, but are mainly seen in children and young adults, with clear female predominance. The clinical symptoms of abdominal IMT are non-specific: alteration in general condition, fever,

Figure 1. Abdominopelvic axial spontaneous contrast CT (a): well-circumscribed rounded pelvic lesion spontaneously hypodense relative to the muscles, associated with a small quantity of peritoneal effusion. Abdominopelvic axial CT after injection of contrast agent in the venous phase (120 s) (b): there is peripheral contrast enhancement with a central hypodense area.

Figure 2. Case no. 2. Macroscopic appearance after surgical ablation (a): the lesion has developed at the expense of the greater omentum and has many congested vessels on the surface. After sectioning (b), the lesion is polymorphic with a central fibronecrotic area (star).

Figure 3. Case no. 2. Histological examination showed the presence of myofibroblastic spindle cells associated with inflammatory lymphocytes and plasma cells (HES, × 40) (a). Immunohistochemical expression with anti-ALK antibody (brown precipitate) (b).
- weight loss, malaise, and gastrointestinal disorders. A mass may be felt by palpation. Laboratory results sometimes show iron deficiency anaemia, thrombocytosis, or polyclonal hypergammaglobulinaemia. The common immunohistological characteristic of them is the expression of the ALK gene, found in 50% of cases;

- other inflammatory pseudotumours, which are poorly defined. They are reactive fibroblastic or myofibroblastic proliferations of infectious or inflammatory origin, and authentic tumours, which are benign or with low-grade malignancy. The following clinical and histological features should point towards these differential diagnoses: an elderly patient, cutaneous or subcutaneous, lymph node, spleen or bladder involvement, an infiltrate prominently consisting of lymphocytes, moderate to severe nuclear atypia, atypical mitoses, necrosis.

The macroscopic appearance of IMT is of a slow-growing, non-encapsulated, frequently single, multilobar mass, which is firm with a yellow to brownish surface, destroying the tissue that it invades. It may show necrotic-haemorrhagic changes and calcifications. It ranges in size between 1 and 20 cm with a mean of 7 cm [3]. Under the microscope it contains a myofibroblastic slightly atypical spindle cell component [4], which is more or less fasciculated in a hyaline stroma, associated with many inflammatory lymphocytes/plasma cells. There is little mitotic activity. Anaplastic lymphoma kinase (ALK) gene expression, although not specific to IPT, is observed in approximately 50% of cases. The ALK gene present on 2p23, is involved in the pathogenesis of this lesion. The presence of clonal rearrangements of this gene in IMT in children and young adults indicates the tumoral character of the lesion rather than any reactive nature.

Abdominal locations are very variable: liver, spleen, pancreas, adrenal glands, retroperitoneal space, diaphragm, mesentery, gastrointestinal tract, etc. Mesenteric and omental forms are the most frequently encountered. Abdominal locations evolve less favourably than extraperitoneal locations, with 23 to 37% of recurrence [5].

Radiological signs are poorly defined because of the polymorphism of IMTs: only small series of cases have been reported and multiphase acquisitions allowing more complete study of the enhancement kinetics are rarely performed in these young people for obvious radiation protection reasons. There is usually a large single lesion, exerting a mass effect on adjacent organs. Its margins are more or less well defined. A CT scan readily shows a homogeneous or heterogeneous lesion, which is hypodense or isodense relative to the muscles. Calcifications and lymphadenomegalies are rare. Contrast uptake is variable [6]: enhancement may be absent, early or late, homogeneous or not, and peripheral contrast enhancement may show central necrosis or fibrosis. The same signs are found with MRI. This can be explained by the variable composition of the tissue of the IMT, including a fibrous component corresponding to the scar tissue. The time for uptake of contrast agent depends on the vascular component: early contrast enhancement in the case of young, oedematous, inflammatory cellular fibrosis; delayed contrast enhancement if the fibrosis is mature—collagen. In theory, it is therefore desirable to make late acquisitions, to highlight late contrast enhancement, even if it is discreet. Given the problems associated with irradiation in young subjects, non-irradiating multiphase exploration is preferable, such as MRI. IMT is a diagnosis by exclusion. The differential diagnoses include non-tumour lesions such as abscesses or tuberculous granulomas, and benign or malignant tumours. A desmoplastic small round cell tumour is a malignant tumour in the young with a poor prognosis, characterised by a single or multiple peritoneal mass located in the omentum or mesentery. A lymphoma or GIST must also be eliminated.

The definitive diagnosis is histological and treatment is surgical. Local or regional recurrence is possible and metastases are rare.

**Disclosure of interest**

The authors declare that they have no conflicts of interest concerning this article.

**References**


