Inflammatory myofibroblastic tumour

Ozgur Firat, MD*
Safak Ozturk, MD*
Taner Akalin, MD†
Ahmet Coker, MD*

From the Departments of *General Surgery and †Pathology, Ege University Hospital, Izmir, Turkey

Correspondence to:
Dr. O. Firat
Department of General Surgery
Ege University Hospital, Floor 3
Bornova 35100
Izmir, Turkey
fax 00 90 232 339 88 38
ozgur.firat@ege.edu.tr

Inflammatory myofibroblastic tumours (IMTs), previously accepted as a subtype of the group of tumours called inflammatory pseudotumours, are now recognized to comprise their own, discrete diagnosis. Since IMTs can arise from various anatomic locations, they concern almost every subspecialty in surgical oncology. The management of these tumours can be challenging because there are no established medical treatment protocols, and tumours can be irresectable owing to their proximity to vital structures. We present the case of a patient whose IMT was removed after 2 trials of chemotherapy after 2 unsuccessful surgeries.

CASE REPORT

A 55-year-old woman was admitted to our department with a retroperitoneal mass. The tumour had been incidentally discovered 3 years previously at $4.9 \times 5.5$ cm in size. At that time, she underwent surgery in a state hospital; however, the surgeons deemed the tumour irresectable because of fixation to the interaortocaval region. Based on the intraoperative biopsy specimen, they diagnosed an undifferentiated cancerous tumour. After recovery, the patient received 3 cycles of chemotherapy (cyclophosphamide 500 mg/m², doxorubicin 50 mg/m² and cisplatin 50 mg/m²). Four months later, the patient qualified for reoperation; however, surgeons again deemed the tumour irresectable. She received an additional 3 cycles of chemotherapy with the same protocol and was followed-up for 30 months. She was subsequently referred to our department for a possible third surgery.

We found nothing remarkable on physical examination except for the previous incision scars. Laboratory test results revealed hypochromic, microcytic anemia accompanying high levels of C-reactive protein and antistreptolysin O. The patient’s erythrocyte sedimentation rate and tumour markers (carcinoembryonic antigen, Ca 19–9, Ca 15–3, Ca 125) were within normal ranges. Positron emission and computed tomography scans showed a hypermetabolic retroperitoneal mass, now $7.5 \times 7.5 \times 8$ cm in size, located in the right para-vertebral region above the iliac bifurcation (Fig. 1), with no invasion to the surrounding vascular structures. The patient underwent a third surgery, and we resected the tumour completely. The postoperative course was uneventful. We detected no sign of recurrence on follow-up at 7 months.

On gross examination, the tumour was a well circumscribed nodular mass with a grey-whitish cut surface. Histopathology findings showed it to be composed of a mesenchymal arrangement of spindle cells with evident cellularity and necrosis, accompanied by mononuclear cell infiltration consisting of dense lymphocytes and plasma cells (Fig. 2). Immunohistochemical stains were negative. We diagnosed an inflammatory myofibroblastic tumour.

DISCUSSION

On presentation, IMTs can constitute a formidable challenge, from diagnosis...
through to treatment. In this report, we present the case of a patient with a retroperitoneal IMT that challenged radiologists, pathologists, oncologists and, inevitably, surgeons. Most of these tumours are discovered incidentally during radiological studies. There are no established decisive criteria for differential diagnosis. The current histopathological definition of an IMT is a distinctive neoplasm composed of myofibroblastic mesenchymal spindle cells accompanied by an inflammatory infiltrate of plasma cells. Nevertheless, tissue samples obtained by computed tomography–guided fine-needle or tru-cut biopsies, and even analysis of perioperative biopsies, are occasionally not enough to establish a diagnosis, and there is no specific immunohistochemical staining for IMTs. As a result, the pathologist usually asks for the whole specimen. In this case, the initial histopathological diagnosis of undifferentiated malignant tumour led surgeons and oncologists astray. Our final diagnosis was only possible after the evaluation of the whole mass and the detection of neoplastic growth of myofibroblastic spindle cells on an inflammatory background.

Currently, surgery is the mainstay of the treatment for IMTs. Complete removal of the tumour generally provides resolution of all symptoms and laboratory abnormalities. However, tumours in intra- or retroperitoneal locations tend to invade adjacent structures, preventing curative resections and breeding local recurrences. These patients require further management. Unfortunately, chemotherapy and radiotherapy are not successful in most patients. Recently, researchers have published promising results with anti-inflammatory agents and anti-tumour necrosis factor-α binding antibodies.

In the case of our patient, it is difficult to comment on the effects of chemotherapy as her tumour had enlarged in the follow-up period. Also, it is rare that after chemotherapy a third laparotomy would be necessary to achieve a complete resection; the decision to pronounce a tumour “irresectable” depends on the experience, skill and local conditions of the surgical team. Until the development and proven success of targeted therapies, these tumours will continue to challenge surgeons and other specialists.

Competing interests: None declared.

References