An inflammatory myofibroblastic tumor (IMT), or inflammatory pseudotumor, is a relatively rare quasineoplastic lesion consisting of inflammatory cells and myofibroblastic spindle cells. The exact etiology of an IMT is unknown and whether an IMT represents a reactive or neoplastic process remains unclear. An IMT was initially described in the lung, but it has subsequently been found to potentially involve nearly all areas of the body (1-3). An IMT usually presents as a single mass within a single organ or sometimes as multifocal lesions within a single anatomic region. An IMT involving noncontiguous multi-organs within different anatomic regions is extremely rare. We present a case of an aggressive IMT that involved the musculoskeletal system and multiple abdominal visceral organs.

Index words: Computed tomography (CT)
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We present a case of an aggressive IMT that involves multiple organs within different anatomic regions.

Case Report

A 68-year-old man presented with a one-week history of right lower extremity weakness and lower back pain. The patient had no trauma history. The laboratory data revealed only a mild elevation of ESR (18 mm/hr) and a minor degree of thrombocytosis with a platelet count of 525,000 platelets/$\mu l$. MR imaging showed irregular bony destructive soft tissue masses involving the body and right pedicle of the L2 vertebra, with expansion to the epidural space and the ipsilateral neural foramen (Fig. 1A-C). Since the lesion was considered a metastasis, we performed a diagnostic work-up to find the primary malignancy. A contrast-enhanced abdominopelvic CT scan demonstrated the presence of multiple nodules in the spleen, both kidneys, pancreas, right paravertebral, and bladder wall (Fig. 1D-H). From brain MR imaging, a 1.5 cm, well enhanced mass was noted that extended from the hypothalamus to the pituitary stalk.
The patient underwent an excisional biopsy for the lesion in the L2 vertebra. A microscopic examination revealed diffuse dense collagenous fibrosis admixed with intervening, mature looking lymphoid cells and plasma cells, which is consistent with an IMT (Fig. 1I, J). An additional ultrasound-guided biopsy of the splenic mass revealed the same microscopic findings as the L2 vertebral lesion, therefore enabling the conclusion of the same diagnosis of an IMT. Thirteen months after discharge, the patient received a follow-up abdominopelvic CT and brain MRI. A contrast enhanced CT scan showed a decrease in the size of the nodules in the spleen, and no residual masses were noted in the kidneys and pancreas (Fig. 1K, L). From the brain MRI, no mass was demonstrable in the hypothalamic area.

Discussion

An inflammatory myofibroblastic tumor, also known as an inflammatory pseudotumor, is defined as “a tumor composed of differentiated myofibroblastic spindle cells usually accompanied by numerous plasma cells and/or lymphocytes” by the 1994 WHO Classification of Soft Tissue Tumors (6). Coffin et al. (1) subdivided this rare tumor type into three histological subtypes: myxoid-vascular, hypocellular fibrous, and the compact spindle cell type. The tumor mainly affects young patients and most often involves the lung and the orbit, but may occur in virtually any anatomic location and at any age.

Whereas the exact etiology of an IMT is unknown, an IMT has been thought to result from inflammation following trauma, surgery, or acute infection. Other investigators argue that an IMT is a true neoplastic lesion. Some IMT cases with malignant transformation or metastasis to distant organs have been demonstrated in the literature (2). There are only sporadic descriptions of the radiological findings for IMT in the medical literature and the reported findings are variable depending on

Fig. 1. A-C. MR imaging demonstrates the mass destroying the L2 vertebral body and the right pedicle, and expanding to the epidural space and the neural foramen. The mass shows heterogeneous hypo- and hyperintensity on the axial T2-weighted image (A) and isointensity on the T1-weighted image (B). The post-contrast T1-weighted image (C) reveals a well-enhanced lesion.
the involved organ, histological subtype, and are even inconsistent within the same organ (3, 4).

IMT bone involvement is extremely rare, and the general imaging features of this involvement have not been well described (7, 8). In our case, a bony lesion demonstrated heterogeneous signal intensity on T2WI, homogeneous iso-signal intensity on T1WI, and heterogeneously well-enhanced intensity on gadolinium-enhanced T1WI with expansion to the epidural space and the neural foramen. These findings cannot be differenti-

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Fig. 1. D-H. Contrast enhanced CT images show variable-sized, multiple, hypodense masses (arrows) in the spleen (D, E), pancreas head (F), right kidney (F), left kidney (G), right paravertebral area (G), and bladder wall (H).
ated from a metastasis or other primary bone tumors. The MR findings of IMT may be low to intermediate signal intensity on T1- and variable signal intensity on T2-weighted images and possible strong enhancement with gadopentetate dimeglumine (3). Han et al. (9) suggested that the T2 hypointensity of a soft-tissue lesion might be explained by a relative lack of both free water and mobile protons within the fibrotic lesions. Hoger et al. (4) reported a case of synchronous IMTs with different subtypes. In that case, the MRI signal on T2-weighted sequence was lower in the spindle cell variant than that in the myxoid-vascular subtype, but reliable differentiation by MRI was not possible. Therefore, MR signal intensities may differ according to histological subtype and/or the degree of inflammation as well as the amount of fibrotic content within the tumor.

An IMT has been reported in various sites within the abdomen, including the liver, spleen, pancreas, adrenal gland, kidney, retroperitoneum, diaphragm, mesentery, and the alimentary and urinary tracts. An IMT has a variable CT appearance. On un-enhanced scans, the mass may be hypoattenuated or isoattenuated relative to both free water and mobile protons within the fibrotic lesions. Hoger et al. (4) reported a case of synchronous IMTs with different subtypes. In that case, the MRI signal on T2-weighted sequence was lower in the spindle cell variant than that in the myxoid-vascular subtype, but reliable differentiation by MRI was not possible. Therefore, MR signal intensities may differ according to histological subtype and/or the degree of inflammation as well as the amount of fibrotic content within the tumor.

Fig. 1. I, J. Diffuse, dense collagenous fibrosis admixed with intervening, mature looking lymphoid cells and plasma cells, which are consistent with IMT histology [H & E staining, I: ×40, J: ×400].

K, L. Follow-up contrast enhanced CT images show a decrease in the size of the masses (arrows) in the spleen (K), left kidney (L), and pancreas (L).
we assume that the IMT also involved the kidneys, pancreas, paravertebral area, bladder wall, and brain. In addition, all of the lesions involving the intra-abdominal organs were homogeneous iso- or hypo-attenuated masses relative to the muscle on contrast-enhanced CT. The density and enhancement patterns of this case were similar to those described in previous reports. However, the radiological findings in this case were nonspecific and the lesions could not be differentiated from a lymphoma, melanoma, or other metastatic or multi-organ involved lesions.

In conclusion, this case certifies that this rare entity may demonstrate non-contiguous, multi-organ involvement, aggressive growth patterns, and variable signal intensities. In addition, this case shows inconsistent enhancement with MR and CT imaging. Therefore, when dealing with such cases, an IMT should be included as a potential differential diagnosis, even though these cases are very rare.

References
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