Aegean Pathology Journal **1**, 22–25, 2004
Available on line at http://www.epd.org.tr

Inflammatory myofibroblastic tumor of the spleen a case report

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Accepted for publication 12.01.2004

Background: Inflammatory myofibroblastic tumor, also known as inflammatory pseudotumor or plasma cell granuloma, is a benign lesion. Although this tumor reported to be more common in lung, it can be detected in extrapulmonary sites. Inflammatory myofibroblastic tumor of the spleen is quite rare.
Case report: Herein, we describe a case of the spleen in a 62-year-old patient treated with surgery.
Conclusion: Most of IMT are benign. But, clinical behavior may vary depending on the site of the tumor and cellular atypia. Thus, it is important to be aware of malignant transformation, and evaluate the localization and cellular features.

Keywords: Spleen, inflammatory pseudotumor, inflammatory myofibroblastic tumor

Introduction

Inflammatory myofibroblastic tumor (IMT) of the spleen is an uncommon benign lesion with unknown etiology. The terms of plasma cell granuloma, inflammatory pseudotumor, pseudosarcomatous myofibroblastic proliferation, xanthamatous pseudotumor, inflammatory myohistiocytic proliferation have all been named to similar lesion with varying amount of inflammatory cells, particularly plasma cells, macrophages, and mesenchymal elements.[1,2] The clinical implication of this farraginous lesion is that the inflammatory pseudotumor is not a single clinicopathologic entity, however, it is an umbrella term to encompass any nonspecific or not otherwise classifiable chronic inflammatory, expansile lesion. They are reported to occur in diverse anatomic sites, but the lung is the organ of apparent predilection. Extrapulmonary inflammatory tumefactions have a tendency for the abdomen, head and neck, and central nervous system. Within the abdomen, areas of predominant involvement include the gastrointestinal tract, urinary bladder, retroperitoneum, mesentery, and liver. On the other hand, IMT of the spleen is a quite rare condition. Many authors suggested that the development of this peculiar tumor is due to response to infectious agents, particularly Epstein-Barr virus or trauma.[3,4]

Recognition of this lesion is important, since the

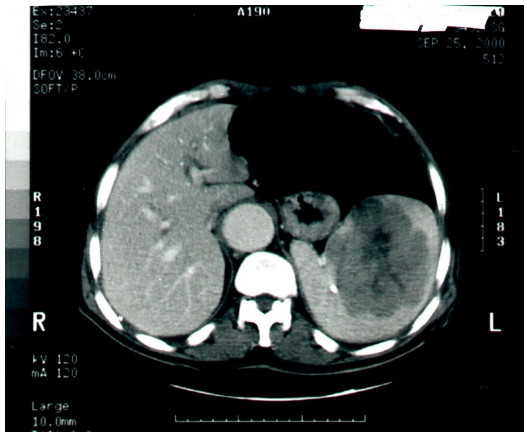

Figure 1. Abdominal computed tomography was demonstrated a large solid tumor mass located in the enlarged spleen.

clinical manifestations and radiographic features may mimic a malignant lymphoproliferative disorder.[5]

We presented an unusual case of IMT of the spleen and reviewed the relevant literature.

Case Report

A 62-years-old man was evaluated for nonspecific abdominal complaints at emergency unit. The physical examination was unremarkable. Computed tomography of the abdomen showed a large solid mass arising from the spleen (Fig. 1). Splenectomy was performed.

On gross pathologic examination, the spleen was enlarged and weighed 855 gr. The cut surface revealed a firm, single, gray-white, well-circumscribed tumor-like mass with focal areas of necrosis and haemorrhage that compressed the adjacent splenic parenchyma. The mass measured 7x6 cm in diameter.

Microscopically, the splenic parenchyma was replaced by a proliferation of spindle-shaped cells (Fig. 2). In some areas these spindle cells had large vesicular nuclei with prominent nucleoli. Focally, they were arranged in a storiform pattern. Admixed with the spindle cells, there was a mixed inflammatory cell infiltrate of lymphocytes, mature plasma cells, eosinophils, histiocytes, and few neutrophils.

Immunostaining for immunoglobulin light chains kappa and lambda, disclosed a polyclonal proliferation of plasma cells. The spindle cells were stained positively with the antibodies to smooth muscle actin and vimentin (Fig. 3). In addition, a mixed population of B and T lymphocytes were also seen.

Discussion

Inflammatory pseudotumor is an uncommon and enigmatic lesion. The spindle cells found in this tumor have features of myofibroblasts. Because of the indefinite relationship of these lesions with

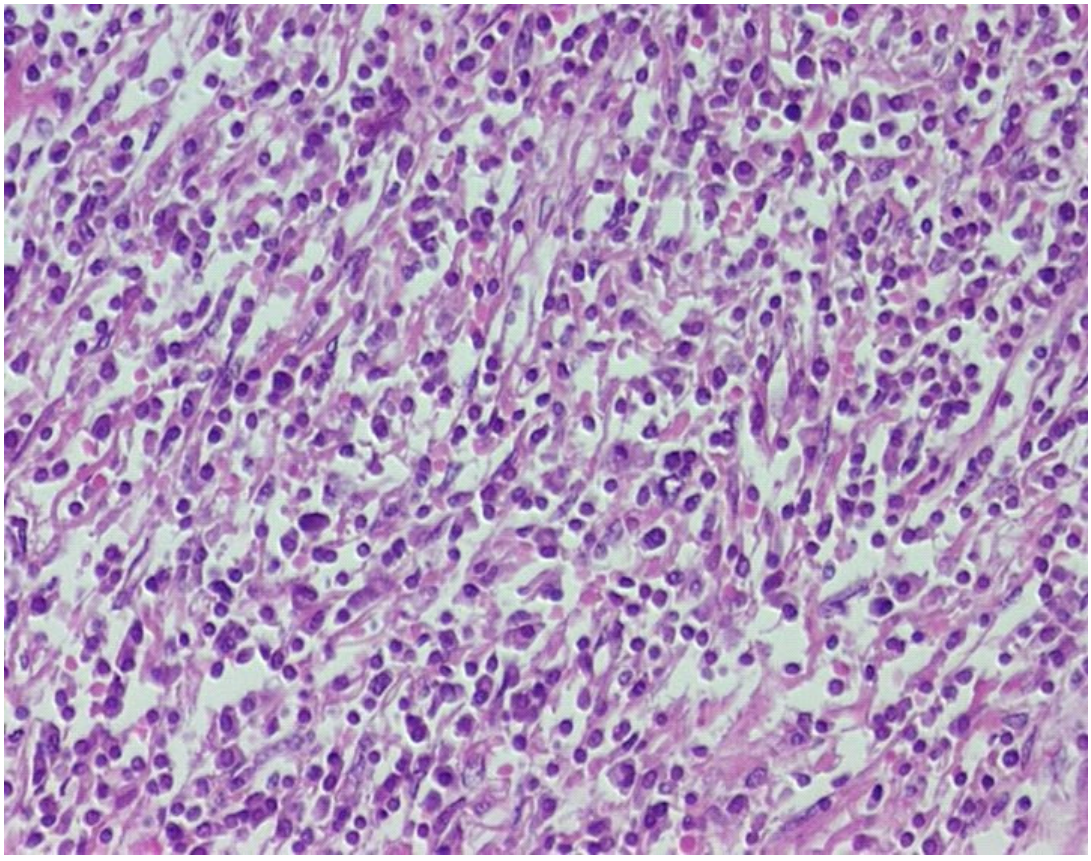

Figure 2. A proliferation of spindle-shaped cells was seen replacing the splenic parenchyma. These spindle cells having large vesicular nuclei were arranged in a storiform pattern (Haematoxylin and eosin, original magnification x 100).

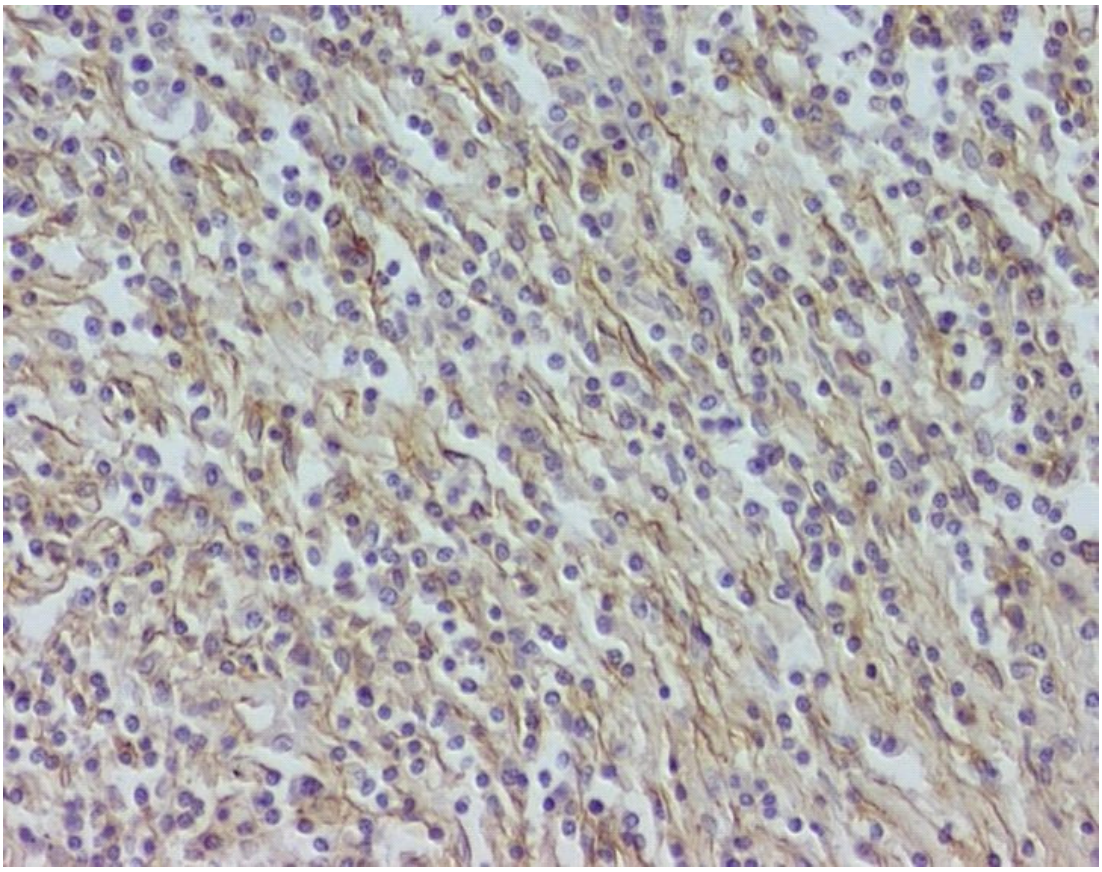

Figure 3. Strongly and diffuse positive cytoplasmic vimentin immunoreactivity in the spindle cells of IMT (Vimentin, x200).

inflammatory fibrosarcoma and their obscure biologic behavior, inflammatory pseudotumor is currently classified as inflammatory myofibroblastic tumor.

IMT is a benign tumor composed of mesenchymal cells intermixed with inflammatory cells. The recognition of a tumor-like lesion in the spleen having a similar morphology to IMT occurring in the lung and elsewhere was first reported by Cotelingam and Jaffe in 1984.[6] As far as we concern, only 39 cases of IMT of the spleen have been reported in the medical literature.[7] The cause and pathogenesis of IMT are unknown. It is thought to be a response to an infectious and noninfectious agents. Some authors considered the condition to be secondary to trauma or surgery. Cytokines are possibly involved in its pathogenesis.[5,8] In our patient no association has been found with infection, trauma or surgery. Recently, some studies have shown chromosomal rearrangements, involving 2p23 near or within anaplastic lymphoma kinase. These recurrent alterations in IMT supported the neoplastic nature of this lesion.[8–10]

IMT presented with clinically palpable mass or incidentally diagnosed during autopsy or routine physical examination, as it was in our case. Although the symptoms vary according to the location of tumor, symptoms such as fever, weight loss, pain were present in some of the patients.[1] In addition, most of these patients presented laboratory anomalies including anemia, thrombocytosis, polyclonal hypergammaglobulinemia and elevated erythrocyte sedimentation rate.[1,5] After surgical resection, all of these laboratory tests resolve dramatically. IMTs are generally solitary lesions, but there are a few reported cases of multifocal tumors.[2,8]

Macroscopically, IMTs were described as non-encapsulated, well- circumscribed firm mass that compressed the adjacent splenic parenchyma. Its cut surface is pale tan, yellow-white. There may be haemorrhagic and necrotic areas.[2] In our patient, a

single mass showing the typical macroscopic features was observed.

Microscopically, IMT is composed of mixed inflammatory cells and spindle cells within a connective tissue stroma. The spindle cells have the characteristics of myofibroblasts. They are reactive immunohistochemically to vimentin, smooth muscle actin and muscle specific actin. Electron microscopic studies have confirmed the myofibroblastic character of the spindle cells.[1,8,11] The plasma cells in IMT stain for kappa and lambda light chains. The presence of the mixed inflammatory infiltrate and the polyclonality of the plasma cells are indicative of the benign nature of IMT.[11]

IMT can usually be treated successfully with surgery.[3] Although the overall prognosis of these patients is generally good, some cases are reported to be locally aggressive and recurred. Furthermore, also invasive and malignant transformation showing cases are reported.[8,9,11,12] Aggressive and recurrent clinical course of IMT was related to the site of the tumor, multinodularity and proximity to the vital structures. Atypical cells with vesicular nuclei, prominent nucleoli, and mitoses are the morphological features of malignant transformation. Some investigators have reported abdominal and sinonasal tumors to show more aggressive behaviour.[1,8,12] Meis and Enzinger have reported a local recurrence rate of 37% and metastases in a series of abdominal cavity IMTs and suggested that these tumors better classified as inflammatory fibrosarcoma.[3]

IMT must be differentiated from inflammatory fibrosarcoma as it is more agressive than IMT.[12] It is not clear whether this is a distinct entity or a heterogeneous process representing the more aggressive type of an IMT. However, IMT is considered as a separate entity in the current WHO classification of Soft Tissue Tumors.[8,12] Further investigations in large series are needed to clarify whether the IMT and inflammatory fibrosarcoma represent the same lesion or closely related entities.

Since clinical and radiographic features may simulate a malignant lymphoproliferative disorder in the spleen, an IMT must be differentiated from hematologic neoplasms such as Hodgkin and non-Hodgkin lymphomas.[2,5,13] Lymph node or bone marrow involvement was not seen in IMT.[2]

IMT is a benign lesion and can be treated with conservative surgery. However, clinical behavior of IMT may vary with the location of the lesion, and cellular atypia, and diagnostic studies may not be specific. Thus, it is important to be aware of malignant transformation, and evaluate the localization and cellular features of the tumor.

References

1. Coffin CM, Watterson J, Priest JR, Dehner LP. Extrapulmonary inflammatory myofibroblastic tumor (Inflammatory pseudotumor). Am J Surg Pathol 1995;19:859-872.
2. Monforte-Munoz H, Ro JY, Manning JT Jr, Landon G, Del Junco G, Carlson TS, Ayala AG. Inflammatory pseudotumor of the spleen: Report of two cases with a review of the literature. Am J Clin Pathol 1991;96:491-495.
3. Batsakis JG, Luna MA, El-Naggar AK, Goepfert H. Inflammatory pseudotumor: what is it? How does it behave? Ann Otol Rhinol Laryngol 1995;104:329-331.
4. Edoute Y, Roguin A, Gallimidi Z, Ben-Izhak O, Nagachandram P, Ben-Ami H. Splenic inflammatory pseudotumor mimicking primary splenic malignancy. Haematologica 1999;84:765-766.
5. McHenry CR, Perzy-Gall HB, Mardini G, Chung-Park M. Inflammatory pseudotumor of the spleen: a rare entity that may mimic hematopoetic malignancy. Am Surg 1995;61:1067-1071.
6. Cotelingam JD, Jaffe ES. Inflammatory pseudotumorof the spleen. Am J Surg Pathol 1984;8:375-380.
7. Noguchi H, Kondo H, Kondo M, Shiraiwa M,Monobe Y. Inflammatory pseudotumor of the spleen: a case report. JPN J Clin Oncol 2000;30:196-203.
8. Coffin CM, Humphrey PA, Dehner LP. Extrapulmonary inflammatory myofibroblastic tumor: a clinical and pathological survey. Semin Diagn Pathol 1998;15:85-101.
9. Griffin CA, Hawkins AL, Dvorak C, Henkle C, Ellingham T, Perlman EJ. Recurrent ınvolvement of 2p23 in inflammatory myofibroblastic tumors. Cancer Res 1999;12:2776-2780.
10. Lawrence B, Perez-Atayde A, Hibbard MK, Rubbin BP, Dal Cin P, Pinkus JL, Pinkus GS, Xiao S, Yi ES, Fletcher CD, Fletcher JA. TPM3-ALK and TPM4-ALK oncogenes in inflammatory myofibroblastic tumors. Am J Pathol 2000;157:377-384.
11. Pettinato G, Manivel JC, de Rosa N, Dehner LP. Inflammatory myofibroblastic tumor (Plasma cell granuloma). Am J Clin Pathol 1990;94:538-546.
12. Coffin CM, Dehner LP, Meis-Kindblom JM. Inflammatory myofibroblastic tumor, inflammatory fibrosarcoma, and related lesions: an historical review with differential diagnostic considerations. Semin Diagn Pathol 1998;15:102-110.
13. Hekimgil M, Isisag A, Ayhan S, Yegen R, Soydan S. Inflammatory pseudotumor of the spleen: A case with clinicopathologic and immunohistochemical characteristics. Turkish Journal of Neoplasia 1996;4:150-153.