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An isolated inflammatory myofibroblastic tumor of adrenal gland

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ABSTRACT

Inflammatory myofibroblastic tumor (IMT) is an uncommon lesion that shows a wide range of anatomic distribution. The adrenal gland, however, is a distinctly rare site of occurrence. To date, only a few cases of IMT arising in the adrenal gland have been reported in the English literature. Here, we report another case of isolated adrenal IMT.

A 34-year-old man presented to the emergency department with a complaint of a sudden severe right-sided back pain. Subsequent computed tomographic scan imaging studies demonstrated a large right adrenal mass associated with a hematoma. The right adrenal gland was resected. Microscopic examination revealed an encapsulated cellular spindle cell proliferation with a prominent inflammatory infiltrate. Immunohistochemically, those spindle cells were diffusely and strongly positive for anaplastic lymphoma kinase-1, and focally and weakly positive for smooth muscle actin. S-100 protein and cytokeratin were negative. The findings were consistent with IMT arising from the adrenal gland.

Although IMTs in the adrenal gland are rare, they should be considered in the differential diagnosis of adrenal masses. The clinical behavior of IMTs in general is currently indeterminate and a close clinical follow-up is recommended. The behavior of adrenal IMTs remains uncertain because of rare reported cases and lack of long-term follow-up. Further follow-up of reported cases and recognition of additional new cases is warranted to unmask the true biological behavior of adrenal IMTs.

1. Introduction

Inflammatory myofibroblastic tumor (IMT) is an uncommon mesenchymal proliferation that demonstrates evidence of myofibroblastic/fibroblastic differentiation. It is characterized by the presence of variable component of inflammatory cells [1]. Since its initial recognition in the lung, cases of IMT have been described in various anatomic locations [2]. The adrenal gland, however, is a distinctly rare site of occurrence. To the best of our knowledge, only a few cases of IMT arising in the adrenal gland have been reported in the English literature [3-7]. Here, we report an additional case of an isolated adrenal IMT.

2. Report of a case

A 34-year-old gentleman presented to the emergency department with a complaint of a sudden severe right-sided back pain. Subsequent computed tomographic scan imaging studies demonstrated a large right adrenal mass associated with a substantial retroperitoneal hematoma (Fig. 1). The size of the mass and the hematoma combined was approximately 11 cm. These findings raised the clinical suspicion of an adrenal cortical carcinoma with associated hemorrhage. Other possibilities, including benign tumors such as adrenal vascular cyst, myelolipoma and leiomyoma, were also considered. Functional adrenal workup was found to be within normal range. The right adrenal gland was resected and the hematoma was evacuated. On gross examination, the mass measured 8 cm, weighed 615 g, and was well circumscribed. It had a variegated tan-yellow to pink cut surface with focal cystic, hemorrhagic, and myxoid areas. In addition, small foci of calcifications were grossly evident. Representative tissue sections were submitted for microscopic studies and subsequent immunohistochemical analysis (Fig. 2). Microscopic examination revealed an encapsulated cellular spindle cell proliferation with fascicular and focally whorling patterns and a prominent inflammatory infiltrate that comprised lymphocytes, plasma cells (<10/high-power fields [HPF]), and histiocytes. There were areas of myxoid stroma, calcifications, and ossifications. The proliferating spindle cells were generally bland looking; however, they exhibited focal mild nuclear pleomorphism and rare mitotic figures (<3/10 HPF). Immunohistochemically, these spindle cells were diffusely and strongly positive for anaplastic large cell lymphoma kinase 1 (ALK1), and focally and weakly positive for smooth muscle actin. S-100 protein was negative. IgG4 immunostain demonstrated less than 10 IgG4-positive plasma cells/HPF. The findings were consistent with IMT arising from the adrenal gland. Upon follow-up, the patient remained recurrence-free more than 4 years after surgical resection of the neoplasm.

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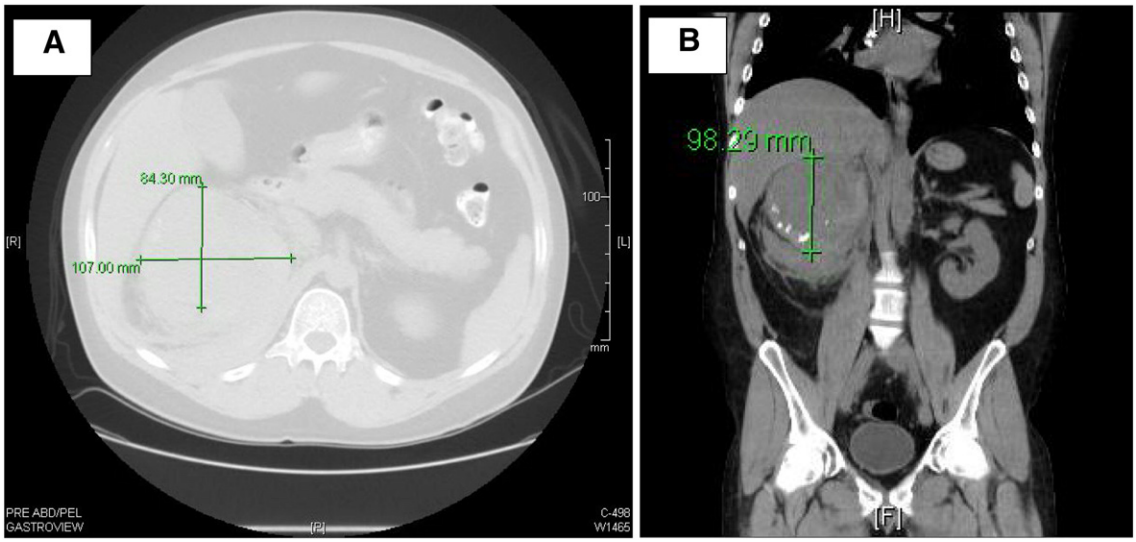

Fig. 1. (A) Computed tomographic scan of the abdomen shows a right adrenal mass with hyperdensity centrally within the mass, indicating intratumoral hemorrhage. (B) Magnetic resonance imaging of the mass demonstrates peripheral calcifications.

3. Discussion

Inflammatory myofibroblastic tumor is an uncommon mesenchymal proliferation characterized by the presence of myofibroblastic/fibroblastic proliferation admixed with variable amount of inflammatory cells [1]. The tumor was initially reported by Brunn [2] in 1939, who described "two interesting benign lung tumors of contradictory histopathology." Subsequently, various nomenclatures were proposed to designate this lesion, namely, inflammatory pseudotumor [8,9], plasma cell granuloma [10], pseudosarcomatous fibromyxoid tumor [11], and inflammatory myofibrohistiocytic proliferation [12]. The term *inflammatory pseudotumor* was most commonly used to describe this enigmatic entity until the name *inflammatory myofibroblastic tumor* was introduced by Day et al [13] in 1986. Inflammatory myofibroblastic tumor was once regarded as a nonneoplastic fibroinflammatory lesion [8,14,15]. The reactive inflammatory theory was supported by the finding of areas of organizing pneumonia in cases of pulmonary IMT [8]. Currently, it is recognized that IMT has distinctive histologic and molecular features, suggesting its neoplastic nature [16-18]. The clinical presentation of patients affected with IMT is naturally determined by the anatomic location of the tumor. Pain, fever, weight loss, and a mass are the most common presenting complaints. Interestingly, a clinical syndrome comprising fever, growth failure (in children), malaise, weight loss, anemia, thrombocytosis, polyclonal hyperglobulinemia, and elevated erythrocyte sedimentation rate (ESR) is encountered in up to one-third of patients. The clinical status normalizes when the mass is surgically resected, but symptoms return once recurrence takes place [19,20].

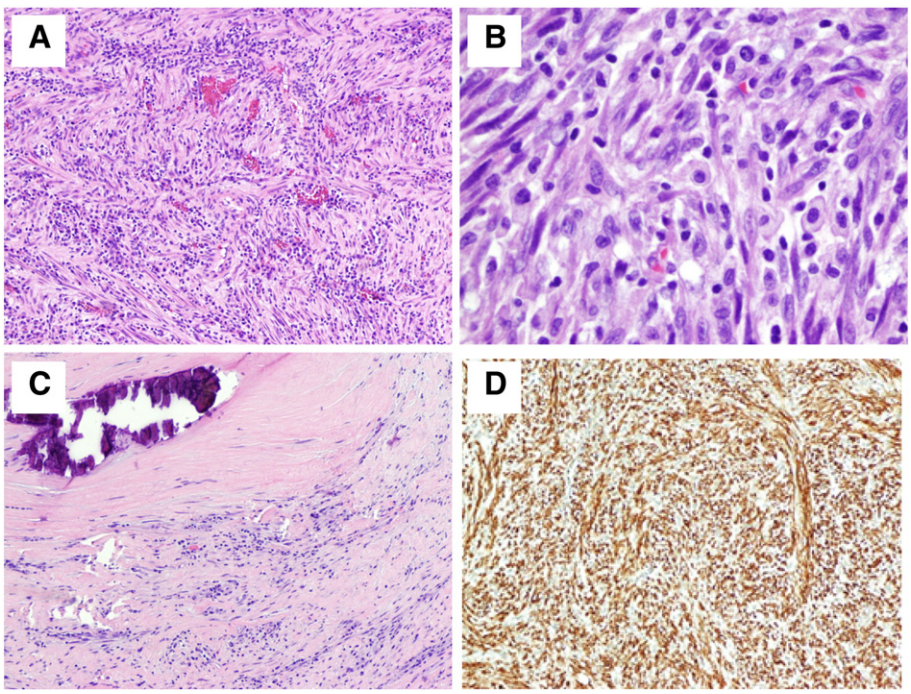

Fig. 2. (A) The neoplasm is composed of fascicles of spindle cells admixed with inflammatory component. (B) The neoplastic spindle cells exhibit bland nuclear features. See scattered inflammatory cells. (C) Hypocellular area with calcification is seen. (D) The neoplastic spindle cells demonstrate diffuse and strong ALK1 expression.