Solitary fibrous tumor with giant multinucleated cells in the retroperitoneum - a case report

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Abstract

Context: Solitary fibrous tumor is a distinct neoplasm, rarely recognized in extrathoracic sites. Case report: The article reports a new case in the retroperitoneum in a 55 year-old man, who presented with urinary symptoms. Tumor was completely excised, and the solitary fibrous tumor was diagnosed after pathologic examination and immunohistochemistry. The patient has remained free of disease for five years since surgery. Conclusion: An uncommon variant of retroperitoneal solitary fibrous tumor with giant multinucleated cells was documented in the study.

Keywords: solitary fibrous tumor, histopathology, retroperitoneum.

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Introduction

Solitary fibrous tumor (SFT) is a distinct neoplasm, which generally arises in the pleura, and is rarely recognized in extrapleural sites such as soft tissue, salivary glands, urinary bladder, adrenals and other system [1]. In the present article, we report a new case with SFT in the retroperitoneum that contains giant multinucleated stromal cells.

Case report

A 55 year-old man presented with urinary symptoms. A heterogeneous retrourinary bladder tumor was suggested by computed tomography scan (Fig 1). Laparotomy was performed, and revealed a circumscribed retrourinary bladder mass without other tumors in the pelvis or abdomen, this mass was surgically removed.

Macroscopically, it was a well circumscribed and firm mass which measured 15 x 10 x 5 cm. The cut surface was solid, brownish and white with hemorrhagic areas.

Microscopically, it was unencapsulated tumor that is composed predominantly of spindle cells. These cells were arranged in short fascicular, storiform and irregular patterns that were separated by collagenous bundles (Fig 2) with numerous branching vascular channels. Spindle cells had eosinophilic cytoplasm and fusiform nuclei. Giant multinucleated stromal cells were focally observed (Fig 3). Cellular atypia was minimal, and mitotic activity was seen in one mitosis per 10 high power fields. Tumor cells were immunoreactive for CD34 (Fig 4) and were negative for cytokeratin, epithelial membrane antigen, c-kit, desmin, S100 protein and actin smooth muscle. Expression of bcl2 and CD99 was focally detected.

The diagnosis of SFT of the retroperitoneum was performed and the patient remained free of disease for five years after diagnosis.
Discussion

Retroperitoneal SFT are rare, about 50 cases have been reported [2, 3, 4]. Most of patients had no leading symptoms and the disease was discovered by imaging studies during the follow-up of other disorders or by medical checkup [1]. Some cases were diagnosed after urinary symptoms or hypoglycemia due to insulin-like growth factor II secreted by the tumor cells [5].

Macroscopically, most tumors are well circumscribed, and show a yellowish-white cut surface, sometimes with necrosis and hemorrhagic areas.

Histologically, typical SFT shows a patternless architecture with an alternating hypocellular and hypercellular areas separated by thick collagenous bundles and branching haemangiopericytoma-like vessels. Tumor cells are round to spindle with little cytoplasm and vesicular nuclei. Cellular atypia is minimal and mitoses are generally scarce rarely exceeding 3 mitoses per 10 high power fields. Some SFT may contain giant multinucleated stromal cells [6], like our case. Typically, tumor cells stain strongly and diffusely for CD34. Expression of CD99 and bcl2 is focally positive. Retroperitoneal SFT should be differentiated from the other solid retroperitoneal masses including gastrointestinal stromal tumors (GIST), desmoid tumor, smooth muscle tumors and giant cell angiofibroma [2].

The confrontation of the pathological examination with immunohistochemistry may be helpful for the diagnosis, particularly in retroperitoneal GIST that is specifically positive for c-kit (CD117) [2]. Giant cell angiofibroma is a benign neoplasm containing multinucleated giant stromal cells and vascular spaces and can be confused with SFT rich in multinucleated giant stromal cells, like our case [1]. Histological and immunohistochemical examinations allow the distinction between these two tumors [2]. Moreover, giant cell angiofibroma may belong to the solitary fibrous tumor group [1]. Although most extrapleural SFT have been reported to be benign histologically, the behavior of extrapleural SFT is unpredictable [4]. Large tumor sizes (> 10 cm), increased cellularity with a high mitotic index (> 4 mitoses per 10 high power fields) have been proposed as the predictors of a poor outcome [4]. Moreover, immunoreactivity for CD34 and bcl2 suggests a favorable outcome [2]. However, there is a poor relationship between morphology and outcome in SFT. Most of retroperitoneal SFT have been treated with complete surgical excision without any adjuvant treatment [2].

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