Solitary fibrous tumor

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Abstract

Solitary fibrous tumor (SFT) is a rare mesenchymal neoplasm which may be found everywhere in the body. It is now distinguished into two forms, pleural and extrapleural, which morphologically resemble each other. Abdominal localizations are quite rare, with 10 cases only reported in bladder; rarely they can be source of paraneoplastic syndromes (i.e., hypoglycemia secondary to insulin-like growth factor). In April 2006 a 74-year-old white male presented with chills, diaphoresis and acute abdominal pain with hematuria. At admission in emergency he underwent an abdominal X-ray (no pathological findings) and an ultrasound examination of the kidneys and urinary tract, which revealed a pelvic hyperechogenic mass measuring approximately 10×8×7 cm, compressing the bladder. Blood chemistry at admission revealed only a mild neutrophilic leucocytosis (WBC 16600, N 80%, L 11%), elevated fibrinogen and ESR, and hypoglycemia (38 mg/dL). Macroscopic hematuria was evident, while urinoculture was negative. Contrast enhanced CT scan of the abdomen and pelvic region revealed a large round neof ormation dislocating the bladder, with an evident contrast-enhanced periphery and a central necrotic area. Continuous infusion of glucose 5% solution was necessary in order to maintain blood glucose levels above 50 mg/dL. The patient underwent complete surgical resection of an ovoidal mass coated by adipose tissue and with well delimited margins; histological findings were consistent with solitary fibrous tumor (SFT). Hypoglycemia resolved completely with removal of the neoplasm.

Case Report

In April 2006 a 74-year-old white male presented with chills, diaphoresis and acute abdominal pain with hematuria. He was not diabetic, was a smoker (about 30 cigarettes/day) and in 2005 had undergone a complete surgical resection of an ovoidal mass coated by adipose tissue and with well delimited margins. Contrast enhanced CT scan of the abdomen and pelvic region revealed a large round neof ormation dislocating the bladder, with an evident contrast-enhanced periphery and a central necrotic area (Figure 1). Continuous infusion of glucose 5% solution was necessary in order to maintain blood glucose levels above 50 mg/dL. The patient underwent complete surgical resection of an ovoidal mass coated by adipose tissue and with well delimited margins (Figure 2). Histological findings were consistent with SFT. The patient underwent radical excision of the neoplasm and has since not been treated with any further therapies. He is still alive without any clinical-radiological evidence of recurrence. Hypoglycemia resolved completely with removal of the neoplasm.

Discussion

The histological appearance of SFT may resemble and overlap with other benign and malignant diagnostic entities, such as heman giopericytoma, leiomyoma, nodular fasciitis, inflammatory myofibroblastic tumor, fibromatosis, and benign peripheral nerve sheet tumor.1 It must be noted that tumors with similar features have been described in literature with different names, thereby creating some confusion among pathologists and clinicians.

The unifying morphological criterion of all these lesions is represented by a well-circumscribed, tan-colored, rubbery mass, which is often tethered by a pedicle and partially encap-
Sected. Microscopically it is described as a patternless proliferation of bland-looking spindle to oval epithelioid cells that form short fascicles and/or clusters, admixed with thick or thin collagen bands, and a prominent branching vasculature. Mature adipocytes and giant multinucleated stromal cells may be present. Mitoses are generally scarce, rarely exceeding 3 mitoses per 10 hp fields.

Malignant counterparts are usually hypercellular lesions, showing variable cytologic (locally moderate to marked) atypia, tumor necrosis, infiltrative margins and higher frequency of mitoses. Immunohistochemical staining for vimentin, Bcl2, CD 99 and CD34 immunoreactivity is performed in order to discriminate SFT from other neoplasms; it is usually completely negative for S-100 protein, cytokeratin AE1/AE3 and neurofilaments. In some cases immunohistochemical negativity has been reported for c-kit, CAM 5.2, factor VIII, HMB-45, AE-1, SMA, CD31, and Fli-1.6,7

One classification distinguishes SFT into two subcategories: fibroblastic and myofibroblastic. The latter strongly expresses alpha smooth muscle actin and desmin, while the former do not. Solitary fibrous tumors are karyotypically characterized by the wild type platelet-derived growth factor receptor-β. its in vitro inhibitory activity in chemo- and radioresistant malignant SFT has recently been reported.

Conclusions

In this case report we describe a SFT growing in the bladder, a quite infrequent localization, which presented a unique associated hypoglycemia.

In contrast with the majority of cases reported in the literature, the behavior of this SFT was not aggressive. Surgical resection was considered conclusive, given that the patient is still alive, despite the initial presentation with paraneoplastic syndrome.

In view of the recent introduction of targeted therapies in cancer treatment and the rarity of this neoplasm, further efforts are needed in order to collect a critical mass of biopsy specimens and to perform more exhaustive anatomicopathological and molecular examinations.

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Figure 1. Computed tomography scan of the abdomen with the mass.

Figure 2. The mass after surgery.
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