Rectal gastrointestinal stromal tumor mimicking a primary prostatic lesion

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The interstitial cells of Cajal have been identified in locations beyond the gastrointestinal tract, including the prostate, uterus and bladder. Indeed, there are reports of primary gastrointestinal stromal tumor (GIST) arising from each of these sites. We report the case of a 72-year-old male who presented with benign prostatic hypertrophy and was diagnosed on retropubic prostatectomy as having a GIST. While the initial clinical and radiologic impression was that of a primary prostatic GIST, subsequent imaging ultimately revealed a small rectal extension as the source of the lesion. The purpose of our report is to highlight the need to assiduously rule-out gastrointestinal sources of GIST prior to making the diagnosis of primary prostatic GIST.

Key Words: gastrointestinal stromal tumor, GIST, extragastrointestinal stromal tumor, EGIST, prostate, CD117

Introduction

Gastrointestinal stromal tumors (GISTs) are mesenchymal neoplasms that – based on ultrastructural and immunohistochemical profiles – are believed to be related to the interstitial cells of Cajal (ICC).[1] The latter are interspersed along the gastrointestinal tract and contribute to gastrointestinal motility.[2] Recently, cells bearing the morphology and immunophenotype of ICCs have been identified in multiple locations outside the luminal gastrointestinal tract; unsurprisingly, these observations have been met with reports of so-called extra-gastrointestinal stromal tumors (EGISTs), at sites including the soft tissue of the omentum and mesentary,[3,4]

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abdominal cavity,[5] retroperitoneum,[5,6] uterus,[7] vagina,[8-10] rectovaginal septum,[11] bladder,[12,13] and prostate.[14,15]

Here we report a case that was initially considered to be a primary prostatic GIST and subsequently found to have a rectal origin. The purpose of this report is to highlight this potential diagnostic pitfall by making urologists and pathologists aware of this important diagnostic consideration.

Case report

A 72-year-old Asian male presented with ongoing obstructive lower urinary tract symptoms. His past history included borderline hypertension and gout. A year earlier he twice underwent transuretheral resection of the prostate (TURP) at an outside facility, and was maintained with an indwelling catheter; histology from the first TURP was interpreted as benign glandular and fibromuscular hyperplasia, the second TURP contained fragments of benign smooth muscle and blood vessels with focal necrosis.

CT examination of the abdomen and pelvis revealed an enlarged prostate, with a low centre of attenuation that was considered to represent an abscess, although necrotic tumor was also considered in the differential; no other significant findings were observed. Transrectal ultrasound revealed an enlarged prostate (12 cm x 11 cm x 10.7 cm; volume > 700 cc) with a central region of cystic degeneration. Owing to his ongoing symptoms the patient underwent a simple retropubic prostatectomy.

Gross examination of the resected specimen revealed a tan, hemorrhagic and friable tumor. Histology showed a cellular spindle cell tumor with a fascicular architecture, Figure 1a and 1b. Multiple serpentine regions of necrosis were present. The cytoplasm was eosinophilic and focally vacuolated; the nuclei oval and elongated, with occasional mitotic activity (7/50 HPF). No residual prostate glandular tissue was identified. Immunohistochemistry showed expression of CD34, CD117, Figure 1c and vimentin; there was no staining for cytokeratin, smooth muscle actin, desmin, S100, or ER/PR. Molecular studies confirmed an 18bp deletion (1653del18), resulting in an in frame deletion of six amino acids, in c-Kit exon 11. On the basis of the morphologic, immunohistochemical and molecular findings, the diagnosis of GIST was rendered. The previous outside TURP specimens were reviewed: the first was found to show fragments of benign glandular and fibromuscular hyperplasia, and GIST; the second contained only fragments of GIST.

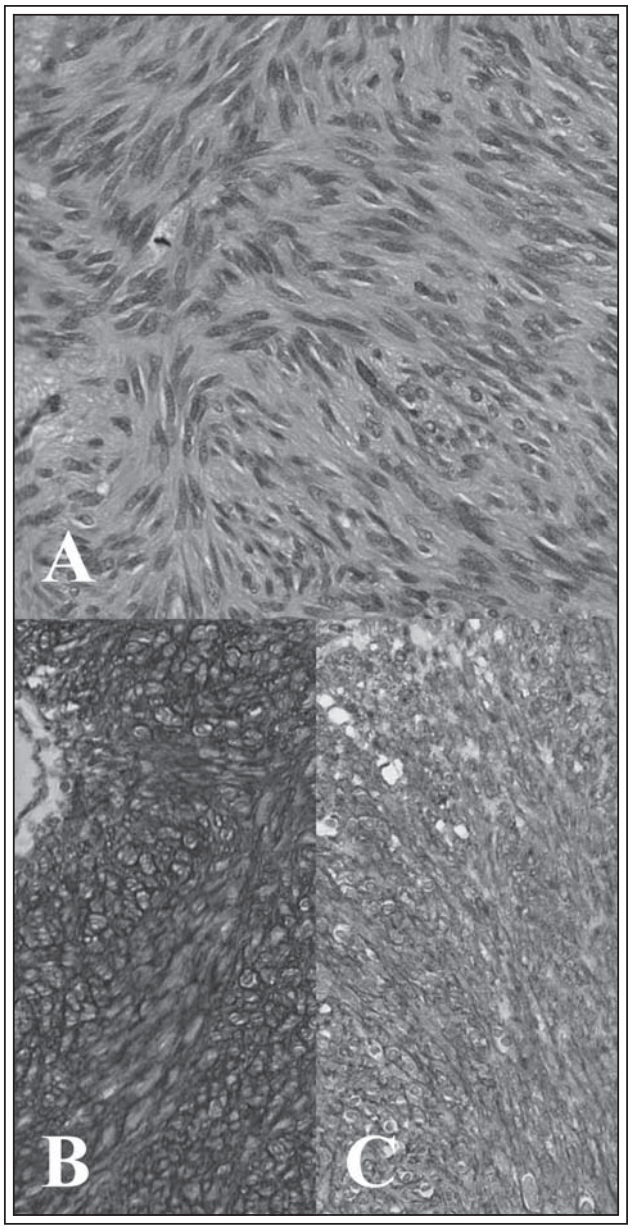

Figure 1a. Section showing effacement of prostate gland by fascicles of spindle cells containing fusiform nuclei (H&E, x200). **b,c.** Immunohistochemistry showing diffuse expression of CD34 (x200), and CD117 (x200), respectively.

The patient was referred to an oncologic centre where MRI confirmed extensive obliteration of the residual prostate by a lobulated mass, as well as significant involvement of the inferior rectal wall which was not appreciated on CT, ultrasound or retropubic prostatectomy, Figure 2. While he declined further surgical intervention, the patient exhibited a good response to medical management.

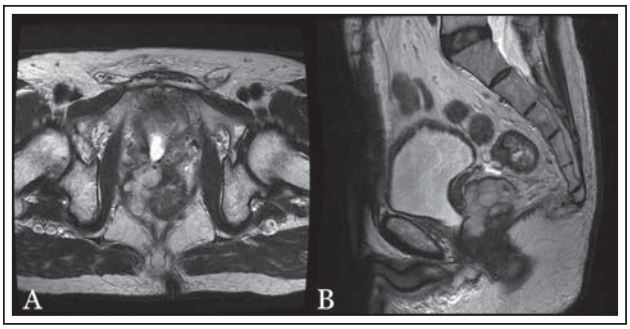

Figure 2. Magnetic resonance imaging (MRI) showing relationship between the prostatic lesion and the rectal wall; A. axial, B. sagittal.

Discussion

Increasingly it is recognized that cells similar to the ICC, or ICC-like cells, exist outside the luminal GI tract; indeed, such an observation would explain the origin of primary EGISTs. To date, there have been at least two reports of primary prostatic GIST.[14,15] In the first case a patient presented with both a large prostate mass and simultaneous liver metastases.[14] Primary prostatic GIST was diagnosed on the basis of imaging and transperineal biopsy without surgical resection of the gland, leading some to doubt whether the lesion is truly a prostatic primary.[16] In a second report, a prostatic GIST was confirmed on imaging and intraoperatively with radical prostatectomy to be confined to the prostate, thereby making the diagnosis of a primary EGIST more tenable.[15]

Upto 10% of GISTs occur in the anorectal region, with many of these lesions producing symptoms sufficient to warrant prostate needle core biopsy.[16] It is therefore necessary to consider GISTs – hence the application of immunohistochemical markers such as CD34 and CD117 – in the differential diagnosis of spindle cell lesions on prostatic biopsy. Differentiation of primary versus secondary prostatic GIST on the basis of biopsy alone is difficult as such tumors may represent (i) primary prostatic GISTs, (ii) metastatic GISTs, (iii) primary rectal tumors or (iv) neoplasms occurring within the space separating the rectum and prostate.

The initial radiologic and surgical impression in this case was that of a primary prostatic EGIST, and it was not until subsequent imaging with MRI that evidence of extension from a low rectal lesion was demonstrated. This case further highlights the need to rigorously exclude a rectal primary, which may have important therapeutic implications for the patient.[17] □

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