

# Extraintestinal gastrointestinal stromal tumor of undetermined origin: Is the mass resection a wrong approach? A case report and review of the literature

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## DISCUSSION

GISTs are generally mesenchymal tumors (70%) originating from gastrointestinal tract, which constitute less than 2% of GIS malignancies. They are usually observed during the 6th and 8th decades with an equal male/female ratio (7). They can be observed in any place of the gastrointestinal system. However they were observed in the stomach and in the rectum with a frequency of 60-70% and 4%, respectively (7).

Thuneberg, proposed a revolutionary hypothesis in 1982, which asserted that ICCs are intestinal cells with a potential pacemaker activity just like cardiac cells (8). Although these cells demonstrate similar morphological characteristics in various tissue types, and were called as ICCs, they are stained positively with methylene blue, and silver stains, while ICCs express a tyrosine kinase enzyme c-kit (D 117 receptor protein), and stain accordingly. Tyrosine kinase receptor c-kit contributes to cell migration, and proliferation in melanoblastomas, hematopoietic precursors, and primordial germ cells (9). C-kit, which is a membrane receptor protein, functions as a growth factor and protooncogen. Abnormal increase in the functions of C-kit is hazardous for GIS. C-kit positive cells undergo transformation and induce formation of GIST.

Presence of EGISTs has been reported in various organs including mesentery, omentum, retroperitoneum, liver, gallbladder, vagina, uterus, urinary bladder or prostate. Besides, cases of EGIST with undetermined origin have been also defined. When we review the literature, dysuria, hematuria, urinary retention, transient rectal bleeding and painful defecation have been reportedly associated with prostatic EGISTs. However definitive diagnosis is made based on radiological findings, and biopsy results. In the pathology, C-KIT (CD117) and platelet-derived growth factor receptor (PDGFRA) mutation carries importance (10). In recent years, the place of GIST-1 (DOG-1) antibody in the diagnosis of GIST and EGIST has been investigated in many studies (7).

Morphological, immunophenotypic and molecular genetic characteristics of EGISTs are comparable.

Therefore, enteroscopy and radiological imaging play important roles in the differential diagnosis. Also, biological characteristics of these tumors resemble each other,

only 10-30% of GISTs demonstrate malignant aggressive behaviour (11). Patients with tumors demonstrating aggressive behaviour are generally admitted with recurrences within 2 years (12).

Rectal GIST rarely invades directly prostate (13). Limited accumulation of information is available in the literature about prostatic GIST, which is thought to originate from prostate. When we reviewed the literature, we observed that patients with prostatic GIST usually were treated with aggressive surgical interventions, and even that these patients had been subjected to radical cystoprostatectomy because of histopathological diagnosis of malignant mesenchymal tumor (14-16). These patients had also been prescribed adjuvant imatinib therapy. Mean follow-up periods of these patients were comparable to ours. Based on prostate biopsy, our patient was diagnosed as gastrointestinal stromal tumor, and the patient was scheduled for radical surgery. However encapsulated mass was easily dissected away from the rectum and the prostate and the mass was extirpated. Since the frozen section of the specimen excised from the surgical margin was tumor-negative, the procedure was terminated. Our patient is presently at 24 month follow-up and he is still receiving adjuvant imatinib therapy without any recurrence.

The present case is the first case with EGIST who underwent extirpation of the mass instead of radical surgery.

In conclusion, the current study presents a rare case of EGIST arising from the prostate and the following assertions are proposed: i) in mass lesions originating from extraintestinal and retroprostatic structures, biopsy can fail to define the origin of these formations, and presence of Cajal cells in the mesenchymal layer of the rectoprostatic septum can be suspected, In fact, it is not surprising that prostatic biopsy specimens also contain rectal tissue in addition to prostatic tissue. ii) during surgical interventions performed for masses with undetermined origin, inadvertently tissues and organs in the vicinity of the lesion may be also excised. iii) as was the case with our patient, based on inadequacy of data, we can not assert that with extirpation of the mass, survival rates comparable to those obtained with radical surgery can be achieved. In the light of all this information, we think

that in consideration of applicability of local resection during perioperative period, and general oncological principles, novel treatment approaches to the cases with EGIST should be investigated.

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