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Gastrointestinal Stromal Tumor and Thrombocytosis

Case Report

Gastrointestinal Stromal Tümör ve Trombositozis

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ABSTRACT

Gastrointestinal stromal tumors (GISTs) represent rare neoplasms of the gastrointestinal tract. Here we describe a case with GIST and thrombocytosis presenting as an acute abdomen. Our knowledge, the co-existence of GIST and thrombocytosis has not been reported so far. A 66-year old female was admitted to the emergency room with epigastric pain and vomiting over duration of 3 days. Physical examination showed abdominal distension, rebound tenderness, and a palpable RLQ mass. The abdominal exploration showed that a 6x6x6 cm mass was located on small intestine. The mass was completely resected and enteroenterostomy was performed. Specific immunophenotype studies results showed GIST. During the post operative follow up, platelets were above normal levels 400x109/l. Therefore, bone marrow biopsy was performed and hiperplasia in megakaryocytes were found. The current treatment for localized disease is surgical resection. Co-existence of thrombocytosis and GIST has never been reported.

Key Words: *Gastrointestinal Stromal Tumour, Thrombocytosis*

ÖZET

Gastrointestinal stromal tümör (GIST) gastrointestinal traktusun nadir görülen neoplamlarındandır. Biz bu yazıda akut batınla seyreden trombosiztozisle birlikte görülen gastrointestinal stromal tümörü tanımladık. Literatür bilgilerine göre bugüne kadar trombositozisle birlikte seyreden GIST tanımlanmamıştır. 66 yaşında bayan hasta, acil servise 3 gündür devam eden epigastrik ağrı ve kusma ile başvurdu. Fizik muayenede abdominal distansiyon, rebound ve sağ alt kadranda palpabl kitle mevcuttu.

Abdominal eksplorasyonda ince barsakta lokalize 6*6*6 cm lik kitle saptandı. Kitle total olarak çıkarıldı ve enteroenterostomi yapıldı. Spesifik immunfenotipik çalışmalar GIST gösterdi. Ameliyat sonrası takipte trombosit değerlerinin 400.000 in üstünde olduğu görüldü. Onun için alınan kemik iliği biopsisinde megakaryosit hiperplazisi saptandı. Lokalize hastalığın şuanki tedavisi cerrahi rezeksiyondur. GIST ve trombositozis birlikteliği bugüne kadar rapor edilmemiştir.

Anahtar Kelimeler: *Gastrointestinal Stromal Tümör, Trombositozis*

INTRODUCTION

Gastrointestinal stromal tumors (GISTs) represent rare neoplasms of the gastrointestinal tract, which are characterized by a specific histological and immunohistochemical pattern (1). In some rare cases coexistence of GIST with hematologic disorders has been reported 1-4. Here we describe a case with GIST and thrombocytosis. To the best of our knowledge, the co-existence of GIST and thrombocytosis has not been reported so far.

CASE REPORT

A 66-year old female was admitted to the emergency room with epigastric pain, constipation and bilious vomiting over duration of 3 days.

Physical examination showed a marked abdominal distension with rebound tenderness, and a palpable RLQ mass. She was in distress, but afebrile and haemodynamically stable.

The laboratory findings were within normal limits except for white blood cells 16.740/µ with left shifted, PLT 574x109/L and C-reactive protein 289.4 mg/l.

A computed tomography scan of the abdomen showed conglomerate of jejunal and ileal loops showing significant dilatation at the center of the abdomen and mild fluid collection among the loops was also remarkable.

The patient underwent an emergency laparotomy under a diagnosis of a strangulated intestinal obstruction. The abdominal exploration showed that a 6x6x6 cm mass was located on jejunoileal junction of small intestine and was adhered on sigmoid colon (Figure 1). The mass was dissected free from the sigmoid colon and a part of the small intestine which the mass was located on was completely resected and enteroenterostomy was performed.

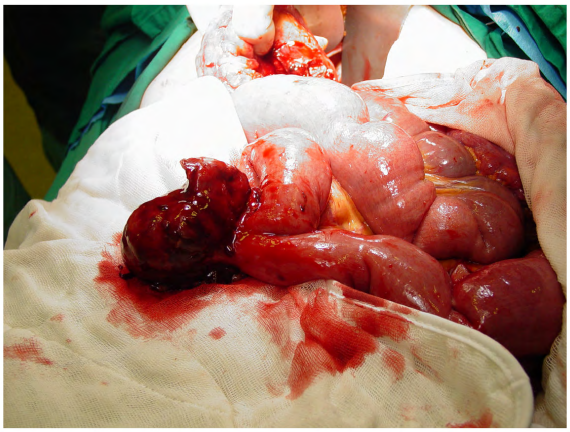

Figure 1: *GIST located on small intestine*

The histological examination demonstrated whirling sheets of spindle cells which were stained positively for CD 117 (c-kit) and CD34, mitotic index> 10/50 HPF, while smooth muscle actin and vimentin were focally positive, and keratine, desmin, S-100 protein were negative (Figure 2).

This specific immunophenotype characterized GIST. During the post operative follow up, the CBC work ups showed that platelets were above normal levels 400x109 /l. Therefore, the patient was consulted to the hematology oncology unit and bone marrow biopsy was performed. The histopathological examination of bone marrow revealed hyperplasia in megakaryocytes characterized with increase in both mature

and immature megakaryocytes with hypolobulated nuclei and scant cytoplasm. The patient was negative for BCR-ABL and Philadelphia chromosome. PET CT and whole body CT scan were perform to evaluate for GIST.

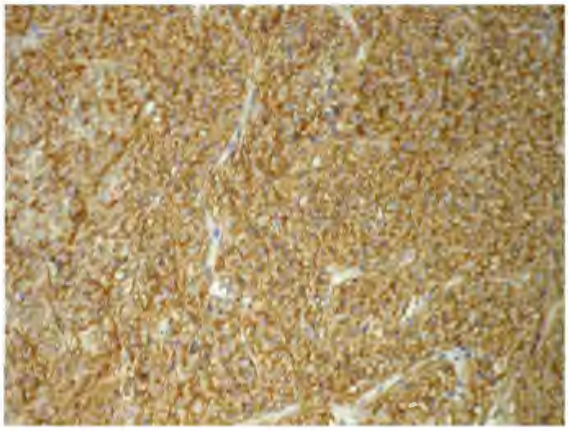

Figure 2: *Immunohistochemical CD117 positivity in tumoral cells(C-kitx100).*

DISCUSSION

GISTs are rare tumors of the gastrointestinal tract, with an annual incidence of 10 to 15 cases per million in Caucasian populations and are the most common mesenchymal tumors arising from the small intestine and comprise up to 15% of all small bowel malignancies (2). GISTs comprise the vast majority of tumors that were formerly classified as leiomyomas, leiomyosarcomas and smooth muscle tumors of the intestine. Jejunal leiomyosarcoma is a rare variety of malignant small bowel tumor and accounts for less than 15% of the malignant small bowel tumors (2).

The GIST is a neoplasm of older individuals that has been the focus of much interest owing to its presumed origin from the same precursor cells as the interstitial cells of Cajal and its typical expression of c-Kit protein (CD117), which is a growth factor receptor with tyrosine kinase activity (3-4). The positive staining for CD117 is thought to confirm the diagnosis of GIST. Ten percent to 30% of these tumors are biologically aggressive; signs of malignant potential are metastases, invasion, greater than 2 to 5 mitoses per 10 high-power fields, scanty stroma, and coagulative necrosis (3-4).

The current treatment for localized disease is surgical resection whereas therapy with imatinib mesylate can be applied in non-operable and /or metastatic tumor (1).

GISTs are most notably associated with pulmonary chondromas and gastric adenocarcinoma, mucosa-associated lymphoid tissue lymphoma,and Burkitt lymphoma (1-4).

Thrombocytosis refers to platelet counts above 400x109 /l. Thrombocytosis has been reported in a variety of solid tumors, including lung, renal, gastric, breast, pancreas, liver and colon malignancies (5). Malignant cells produce certain cytokines such as interleukin-6 and other growth factors, which are capable of inducing platelet production5. While some studies point out that thrombocytosis may reflect tumor burden, other studies suggest that platelets may contribute to tumor growth and metastases (5). Platelets release both platelet-derived growth factor (PDGF) and thrombospondin. PDGF functions as a potent mitogen for different cell types and thrombospondin is an adhesive glycoprotein that may cause the adhesion of tumor cells and promote metastases (5).

Co-existence of thrombocytosis and GIST has never been reported. The present case raises the question of whether this is an incidental finding for our patient or the two conditions have any biological relevance. Molecular studies investigating the expression of platelet secretory factors in patients with GIST are required to elucidate the differences in clinical outcomes and findings in this co-existence.

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