

ANALES MEDICOS

Volumen **11**
Volume

Número **1**
Number

Enero-Abril **2005**
January-April

Artículo:

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stromal tumor (GIST)

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Intussusception due to gastro-intestinal stromal tumor (GIST): A case report

Mauricio CM de Almeida,*
Sun Rey Lin,* Ana Paula S
Carvalho,* Ricardo
Artigiani Neto,* Laércio
Gomes Lourenço,* José
Carlos Del Grande*

* Department of Surgery, Emergency
Service, Federal University of São
Paulo, São Paulo, Brazil.

Correspondence:

Dr. Mauricio CM de Almeida
Alameda dos Picas 799 # 71
São Paulo, Brazil, C.P. 04086-002
Fax: +5511 5737669
Tel. +5511 5713626

Abstract

Objective: The authors present a rare manifestations of stromal tumors; an ileal tumor leading to intussusception. **Material and methods:** (Case report), a 39 years old female patient: Present colicky abdominal pain and constipation for 15 days; seven days prior to the admission she had a single episode of diarrhea with blood, physical examination revealed mild pallor; an abdominal mass measuring 10 cm at the right flank. Plain abdominal x-ray demonstrated a soft tissue mass effect and atypical absence of gas at the right flank. Ct scanning was performed, revealing thickening of the right colon and an image of concentric layers, highly suggestive of intussusception, barium enema confirmed the diagnosis. Treatment: Exploratory laparotomy was performed and surgical findings were compatible with ileocolic intussusception, a right colectomy with resection of the invaginated ileum was done followed by primary anastomosis. **Results:** Histopathological studies confirmed a 2.4 cm gastro-intestinal stromal tumor (gist), 21 cm from the ileocecal valve compromising only the submucosae. Immunohistochemical test were positive for CD 117 (C-KIT), CD 34, 1a4 and vimentin. Mitotic rate was lower than 5/50 high-power fields (HPF). **Discussion:** Intussusception occurs rarely in adults (less than 5%) and is responsible for only 1% of the all cases of the obstruction in this group, in 95% of occurrences, etiology is identifiable. Symptoms are related to the location of the tumor. Gastric lesions more often present with bleeding (60-70%).

Key words: Gastro-intestinal stromal tumor, intussusception, abdominal pain, constipation.

Resumen

Objetivo: Los autores presentan unas manifestaciones raras de tumores del estroma. Un tumor ileal que lleva a la intususcepción. **Material y métodos:** (Caso reportado) Paciente femenino de 39 años de edad que presentaba; dolor abdominal tipo cólico y constipación, durante 15 días. Siete días antes de su internamiento presentó un periodo único de diarrea con sangre. En el examen físico revela palidez leve y masa abdominal de 10 centímetros en flanco derecho. La radiografía abdominal demostró una masa de tejido suave y ausencia de gas en el mismo lado. Se practicó un escaneo que reveló un colon derecho más espeso y una imagen de capas concéntricas altamente sugestiva de intususcepción. El enema de bario, confirmó el diagnóstico. **Tratamiento:** Laparotomía exploradora fue realizada, y los hallazgos quirúrgicos fueron compatibles con una intususcepción ileocólica. Fue realizada una colectomía derecha con resección del íleo invaginado, seguido por una anastomosis primaria. **Resultados:** Los estudios histopatológicos revelaron: Tumor estromal gastrointestinal (GIST), a 21 centímetros de la válvula ileocecal; comprometiendo sólo la submucosa. Los tests inmunohistoquímicos fueron positivos para CD 117 (C-KIT), CD, 34, 1 a 4 y vimentin. El tiempo mitótico fue menor que 5/50 de campos de alto poder (HPF). **Discusión:** Intususcepción ocurre raramente en adultos (menos de 5%), y es responsable de solamente del 1% de todos los casos de obstrucción en este grupo. En un 95% de ocurrencia, la etiología es identificable. Los síntomas están relacionados con la localización del tumor. Las lesiones gástricas, más a menudo se presentan con sangrado (60-70%).

Palabras clave: Tumor estromal gastrointestinal, intususcepción, dolor abdominal, constipación.

The scientific community has recently been paying increased attention to the gastro-intestinal stromal tumors (GIST). Previously described as tumors arising from muscular tissue (Stout, 1962),¹ neural tissue or mixed tumors, it is nowadays believed that they originate from a specific interstitial cell called "cell of Cajal".² It is common consent that stromal tumors belong to a particular group of neoplasias, characterized by expressing the tyrosine kinase receptor KIT (CD117) at immunohistochemical tests.³

Surgical resection has been so far the treatment of choice, but innovative use of a tyrosine kinase inhibitor, imatinib mesylate (STI-571, Gleevec or Glivec) for patients with metastatic or inoperative disease, has been showing promising results.^{4,5}

Stromal tumors are more often found in stomach (60%-70%), followed by the small intestine in second place (20%-30%). Clinical features are unspecified and depend basically on the size and location of the tumor. Usually asymptomatic, these tumors are commonly discovered incidentally in necropsies or during laparotomies for other causes.

The authors present a rare manifestation of stromal tumors: an ileal tumor leading to intussusception.

CASE REPORT

A 39-year-old female patient presented to the emergency department of Hospital São Paulo, São Paulo, Brazil, with colicky abdominal pain and constipation for 15 days. She denied fever or weight loss and referred normal intestinal habit until then. Seven days prior to the admission she had a single episode of diarrhea with blood.

Physical examination revealed mild pallor; an abdominal mass measuring 10 centimeters at the right flank and normal bowel sounds. Digital rectal examination showed no abnormalities.

Plain abdominal X-ray demonstrated a soft tissue mass effect and atypical absence of gas at the right flank. CT scanning was performed, revealing thickening of the right colon and an image of concentric layers, highly suggestive of intussusception (*Figure 1*). Barium enema confirmed the diagnosis (*Figure 2*).

TREATMENT

Exploratory laparotomy was performed through a median incision. Surgical findings were compatible with ileocolic intussusception (*Figure 3*), and reduction was not possible. A right colectomy with resection of the invaginated ileum was done, followed by primary anastomosis. Gross pathology showed an ileal tumor of approximately 3 cm (*Figure 4*).

Histopathological studies confirmed a 2.4 cm gastro-intestinal stromal tumor (GIST), 21 cm from the ileocecal valve compromising only the submucosa. Immunohistochemical tests were positive for CD 117 (C-KIT) (*Figure 5*), CD 34, 1a4 and vimentin. Mitotic rate was lower than 5/50 high-power fields (HPF).

Based on these features the tumor was classified as a very low risk GIST, according to Fletcher et al. The patient recovered with no further complications and was sent home on the seventh postoperative day.

DISCUSSION

Intussusception occurs rarely in adults (less than 5%) and is responsible for only 1% of all cases of bowel obstruction in this group. In 95% of occurrences, etiology is identifiable.⁶

Symptoms due to GIST are related to the location of the tumor. Gastric lesions more often present with bleeding (60 to 70%), and the presence of hematemesis or melena implies urgent evaluation and indication of endoscopy.

Other symptoms such as anorexia, weight loss, postprandial fullness and palpable mass are also described. Acute abdomen, either perforative, hemorrhagic or obstructive, and intussusception are rare.

Among a significant number of asymptomatic patients, the diagnosis happens to be incidental, for example, during laparotomies or laparoscopies for other reasons.

Gastrointestinal tumors leading to intussusception are unusual.⁷ Definitive diagnosis of intussusception is normally reached only during surgical procedure. The most specific and sensitive complementary methods are the CT-scan and the ultrasonography. Although less used lately, barium enema remains as an interesting option for the diagnosis of intussusception.

The gastro-intestinal stromal tumors (GIST) have been target of a great interest. Consensus over the classification of these tumors based on the presence of the tyrosine kinase receptor KIT and its answer to treatment with imatinib mesylate has brought a whole new perspective to these patients.

GIST's are potentially malignant tumors, classified mainly according to their size and mitotic rate. Nevertheless, even very low risk tumors ≤ 2 cm and < 5 mitoses per 50 high-power-fields – can metastasize. Therefore, every GIST should be followed up for local and distant recurrence for at least 5 years,⁸ though it is known that they could happen even within 10 years.

GIST expresses the c-kit through CD-117 in 95% of cases. The CD-34, another marker associated with en-

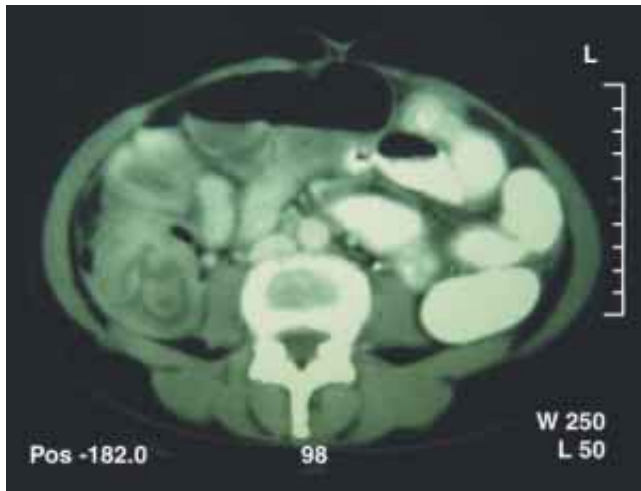


Figure 1. CT scan at the level of the intussusception.



Figure 2. Barium enema confirmed the diagnosis of intussusception.

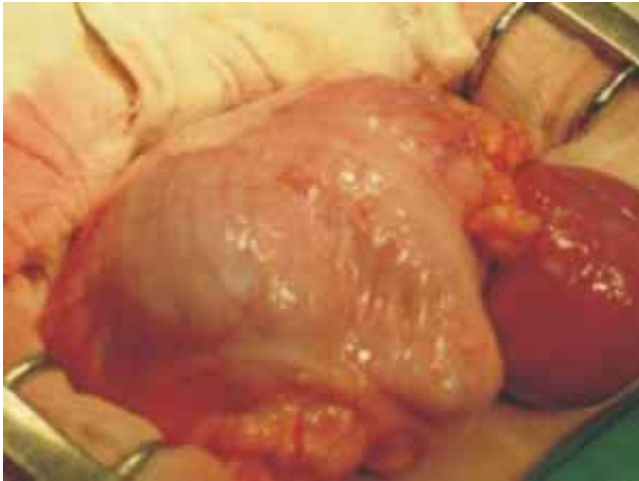


Figure 3. Surgical finding: a ileocolic intussusception.



Figure 4. Gross pathology after cecum opening (arrow) showed an ileal tumor of approximately 3 cm (arrowhead).



Figure 5. Positive immunohistochemical CD 117 (C-KIT).

dothelial cells, is also positive in 60 to 70% of cases, but is not considered specific, once it can be found in a great number of tumors as well. Several other markers such as SMA (smooth muscle actin), desmin, vimentin and S-100 are normally negative.

Differential diagnosis is made with leiomyomas, leiomyosarcomas, schwannomas, fibromatosis and desmoid tumors. Melanomas, angiosarcomas, small cell lung cancer, ovarian carcinoma, seminoma, neuroblastoma and mastocytoma should also be considered, once they all express the c-kit.

Surgery is the treatment of choice, but in inoperative cases or metastatic disease, the imatinib mesylate can be used with good results, showing partial response as much as in 83,5% of cases, depending on immunogenetic features.⁴

Treatment with imatinib mesylate was not used in this patient since there was no indication to do so (neither metastatic nor inoperative disease). The immunogenetic study for exon 11, besides bombesine and colecistoquinine, could be useful to predict malignancy on this case. Reubi and col (2004) have suggested that malignant GIST expresses more bombesine, colecistoquinine and vasoactive intestinal peptide (VPAC2) receptors.⁹ These receptors are also present in tumors resistant to therapy with imatinib mesylate.

BIBLIOGRAPHY

1. Stout AP. Bizarre smooth muscle tumor of the stomach. *Cancer* 1962; 15: 400-409.
2. Kindblom LG, Remotti HE, Aldenborg F et al. Gastrointestinal pacemaker cell tumor (GIPACT): Gastrointestinal stromal tumors show phenotypic characteristics of the interstitial cells of Cajal. *Am J Pathol* 1998; 152: 1259-1269.
3. Fletcher C, Berman J, Corless C et al. Diagnosis of gastrointestinal stromal tumors: A consensus approach. *Hum Pathol* 2002; 33: 459.
4. Henrich MC, Corless CL et al. Kinase mutations and imatinib response in patients with metastatic gastrointestinal stromal tumor. *J Clin Oncol* 2003; 21: 4342-4349.
5. Eisenberg BL. Imatinib Mesylate: a molecularly targeted therapy for gastrointestinal stromal tumors. *Oncol* 2003.
6. Azar T, Berger DL. Adult intussusception. *Ann Surg* 1997; 226(2): 134-138.
7. Crowter KS, Wyld L, Yamani Q, Jacob G. Case report: Gastroduodenal intussusception of a gastrointestinal stromal tumor. *Br J Radiol* 2002; 75(900): 987-9.
8. DeMatteo RP, Lewis JJ, Leung D et al. Two hundred gastrointestinal stromal tumors: Recurrence patterns and prognostic factors for survival. *Ann Surg* 2000; 231: 51-58.
9. Reubi JC, Korner M, Waser B, Mazzuchelli L, Guillou L. High expression of peptide receptors as a novel target in gastrointestinal stromal tumors. *Eur J Nucl Med Mol Imaging* 2004; 31(6): 803-10.