1130-0108/2006/98/5/387-389
Revista Española de Enfermedades Digestivas

Rev Esp Enferm Dig (Madrid)
Vol. 98. N.° 5, pp. 387-389, 2006

PICTURES IN DIGESTIVE PATHOLOGY

Lower GI bleeding secondary to a stromal rectal tumor (rectal GIST)

R. de Francisco, G. Díaz, V. Cadahía, R. F. Velázquez, F. Giganto, O. González and L. Rodrigo

Departament of Gastroenterology. Hospital Central de Asturias. Oviedo, Spain

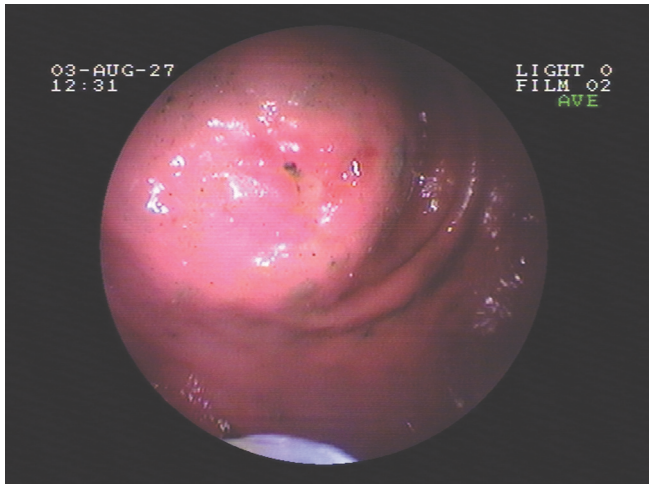

Fig. 1.- Endoscopic picture of a rectal GIST showing an ulcerative lesion on its vertex.
Aspecto endoscópico del GIST rectal, ulcerado en su cúspide.

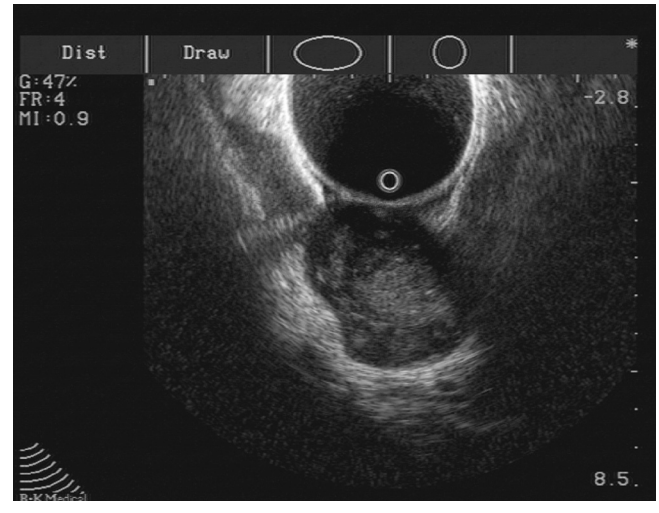

Fig. 2.- Endoanal ultrasonography showing a large, submucosal lesion, corresponding to a rectal GIST.
Ecografía endoanal mostrando una gran lesión submucosa correspondiente al GIST rectal.

CASE STUDY

We present the case of a 64-year-old woman without any known allergies to drugs, who had smoked 20 cigarettes a day for more than thirty years, and with no other relevant history, who presented to our Department of Gastroenterology complaining of rectal bleeding and secondary ferropenic anemia with a hemodynamic repercussion. A colonoscopy was made to reveal the presence of a polypoid, submucous, ulcerated lesion in its vertex (8 cm from the anal margin) (Fig. 1).

An endoanal ultrasound scan showed a heterogeneous mass located in the posterior wall of the rectum, approximately 7 cm in size, with no infiltration of perirectal fat (Fig. 2). A biopsy was made with a tru-cut needle, and the pathological study showed a proliferation of fusiform cells, with no mitoses or atypias, and strongly positive for the CD-117 marker; staining by other markers (CD-34, desmine, actine, and S-100) was negative, except for one slight ki-67-related positivity, less than 10%. An abdominal CAT scan revealed no metastases at all levels. With a preoperative diagnosis of rectal stromal tumor, the mass was removed by local excision with preservation of the rectum. The patient is currently in the eighteenth month of follow-up, and has no signs or symptoms of relapse, neither locally nor distally.

DISCUSSION

The incidence of GISTs is greatest in the fifth and sixth decades of life. Histologically they are made up of fusiform, epitheloid, or mixed cells. The diagnosis is confirmed by immuno-histochemical techniques, and by the expression of a

tyrosine-kinase receptor (Kit or CD-117, the product of gene c-kit). The presence of ADN mutations may result in an abnormal receptor that is constantly activated, even in the absence of an appropriate ligand, which results in uncontrolled cellular proliferation. Sixty to seventy percent of GISTs also have concurrent positivity for CD-34. A group of GISTs exist that have a mutation in PDG-FRA (a receptor derived from platelet growth factor alpha), and these are negative for c-kit (1,2). A differential diagnosis with other mesenchymal tumors is necessary, and is based on negativity for desmine and actine (positive in leiomyomas), and a negative S-100 protein (positive in schwannomas) (3).

Approximately 30% of GISTs are malignant, but clear criteria for malignancy do not exist. Size (> 5 cm) and mitotic index (> 5 mitoses/50 fields) are most useful morphologic features. Other factors for malignancy include location (stomach, better prognosis), the presence of necrosis areas, hemorrhage, hypercellularity, nuclear atypias, etc. The monoclonal antibody Ki-67 is also used as a predictive marker on the basis of its capacity to detect the presence of a nuclear antigen that is only expressed in proliferative cells (if > 10% of cells, it is associated with a worse prognosis). Endoscopic appearance usually includes the presence of a submucosal lesion that is covered with normal mucosa, with an apical ulcerated area. This renders the usefulness of endoscopic biopsies low, and thus frequently insufficient to tell benign from malignant lesions. In echo-endoscopy these lesions appear as hypoechoic areas originating in the muscular layer itself or the muscularis propria. Endoscopic findings associated with a good prognosis include size smaller than 3 cm, lesion homogeneity, and presence of regular contours. Nevertheless, no definitive echo-endoscopic features allowing differentiation between GISTs and leiomyomas are currently available.

Treatment is mainly surgical. Survival at 5 years after surgery ranges between 20 and 80% (4). Relapse is frequent, both local and metastatic, mainly in the liver, followed by the lungs and bones.

The treatment of non-resectable tumors has changed in recent years as we now have a tyrosine-kinase STI-571 inhibitor (imatinib mesylate), which manages to reduce tumor size, or at least to control tumor progression, in 80-90% of cases (5).

In summary, we describe the case of a considerably sized rectal GIST with a low mitotic index, and without associated metastases, which was removed surgically. Clinical follow-up has to be necessarily prolonged in such a case, due to a high risk of relapse even in the long term.

REFERENCES

1. Fernández A, Aparicio J. Gastrointestinal Imatinib and stromal tumor (GIST): a selective targeted therapy. Rev Esp Enferm Dig 2004; 96: 723-9.
2. Heinrich MC, Corless CL, Duensing A, McGreevey L, Chen Cl, Joseph H, et al. Gastrointestinal PDGFRA activating mutations in stromal tumors. Science 2003; 299: 708-10.
3. Davila RE, Faigel DO. GI stromal tumors. Gastrointest Endosc 2003; 58: 80-8.
4. Lehnert T. Gastrointestinal sarcoma (GIST): a review of surgical management. Ann Chir Gynaecol 1998; 87: 297-305.
5. Demetri GD, Von Mehren M, Blanke CD, Van den Abbeele AD, Eisenberg B, Roberts PJ, et al. Gastrointestinal efficacy and safety of imatinib mesylate in advanced stromal tumors. N Engl J Med 2002; 347: 472-80.

Hemorragia digestiva baja secundaria a tumor estromal rectal (GIST rectal)

R. de Francisco, G. Díaz, V. Cadahía, R. F. Velázquez, F. Giganto, O. González y L. Rodrigo

Servicio de Aparato Digestivo. Hospital Central de Asturias. Oviedo

CASO CLÍNICO

Presentamos el caso de una mujer de 64 años, sin alergias conocidas, ni antecedentes de interés, que ingresa en el Servicio de Digestivo, por presentar rectorragias de repetición, de 7 días de evolución, junto con anemización y ligera repercusión hemodinámica secundaria.

Se realizó colonoscopia visualizando en ampolla rectal, a ocho centímetros del margen anal, la existencia de una lesión polipoide, submucosa, ulcerada en su vértice (Fig. 1).

La ecografía endoanal mostró una masa situada a nivel de la pared posterior del recto, heterogénea, de 7 centímetros de diámetro, sin infiltración de la grasa perirrectal (Fig. 2).

Se realizó biopsia con tru-cut y el estudio A-P, mostró una proliferación de células fusiformes, sin mitosis, ni atipias, mostrando CD-117 (+), CD-34 (-), desmina (-), actina (-) S100 (-) y Ki-67 (+) (< 10%) por técnicas de inmuno-histoquímica. El TAC abdominal no mostró presencia de metástasis a ningún nivel. Con el diagnóstico preoperatorio de tumor estromal rectal fue intervenida, realizándole extirpación localizada de la lesión, con preservación del recto. Actualmente se encuentra asintomática al cabo de 1,5 años de seguimiento, sin signos de recidiva local, ni a distancia.

DISCUSIÓN

La incidencia de tumores GIST es mayor en la quinta-sexta décadas de la vida. Histológicamente están formados por células fusocelulares, epiteloides, o mixtas. El diagnóstico se confirma por técnicas de inmunohistoquímica, por la expresión de un receptor celular con actividad tirosin-kinasa (CD-117, producto del gen c-kit). El 60-70% de los GIST presentan además positividad para el CD-34. Existe un grupo de GIST, que presenta una mutación en el PDG-FRA (receptor derivado del factor de crecimiento alfa de las plaquetas) y son negativos para c-kit (1,2). El diagnóstico diferencial con otros tumores mesenquimales se realiza en base a la negatividad para desmina y actina (+) en los leiomiomas y de la proteína S100 (+) en los Schwannomas (3).

Aproximadamente el 30% de los GIST son malignos, pero no existen criterios claros de malignidad. El tamaño (> 5 cm) y el índice mitótico (> 5 mitosis/50 campos de gran aumento) son los rasgos morfológicos más útiles. Otros factores indicativos de malignidad son la localización (estómago, mejor pronóstico) y la presencia de áreas de necrosis, hemorragia, hipercelularidad o atipia nuclear. El aspecto endoscópico muestra, por lo general, la presencia de una lesión submucosa, recubierta por mucosa normal, con una zona umbilicada o ulcerada en su cúspide. Los hallazgos ecoendoscópicos que se asocian con benignidad son el tamaño menor de 3 cm, la homogeneidad de la lesión y la presencia de contornos regulares. Sin embargo, no existen hallazgos ecoendoscópicos definitivos, que nos permitan diferenciar claramente los GIST, de los leiomiomas.

El tratamiento se basa en la extirpación quirúrgica. La supervivencia tras la intervención a los 5 años, oscila entre el 20-78%. La recidiva es frecuente bien sea local o metastásica, de localización principalmente hepática seguida por el pulmón y óseas.

El tratamiento de los tumores no resecables, o con metástasis, consiste en la utilización de un inhibidor de la tirosin-kinasa denominado STI571 (Imatinib mesilato), que consigue una reducción del tamaño, o al menos control de la progresión tumoral, en el 80-90% de los casos (4,5).