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Gastrointestinal stromal tumor (GIST) - medical rarities?

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Abstract
Although their overall incidence is low, GISTs are distinctive subgroup of gastrointestinal mesenchymal tumors which express CD117 or platelet derived growth factor receptor alpha (PDGFRA). Considered as rare digestive cancers, tumors like schwannomas, neurofibromas, gastrointestinal leiomiomas are now reclassified as GIST based on immunohistochemistry studies. GIST are more frequent in stomach (40-70%), small bowel (20-40%), colon (5-15%), meanwhile locations such as mesentery, omentum, retro peritoneum in less of 5%. 10 GIST patients were surgically managed during 2004-2009. 5 gastric and 5 small bowel GIST. Most with symptomatic disease: palpable tumor, abdominal pain, anemia, fatigue, superior digestive hemorrhage or occlusion. Imagistic diagnosis consisted of: barium swallow, abdominal sonography, CT and PET-CT. Confirmation was made by hystopathological exam and immunohistochemistry. All patients had more or less wide surgical resections. For some patients there was also a specific adjuvant treatment. All patients survived after surgery. The principle of surgery for GIST is R0 resection of the tumor. Tumor rupture or R1 resection of the primary tumor has a negative impact on disease free survival. Some patients (great volume tumors, R1 or R2 resection) had adjuvant treatment. Imatinib mesilat and derivates showed a significant improvement of recurrence free survival with one condition: permanent treatment. Surgery remains the mainstay of treatment in patients with localized, resectable GIST. Recurrence rate of 17-21% and 5 years survival rate of 48-70%, even in resectable GIST, impose an adjuvant treatment.
Keywords: rare, immunohistochemistry, R0 resection, imatinib mesilate

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