

A huge pelvic-abdominal malignant gist tumor in a patient with neurofibromatosis type1: A case report and literature review

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Gastrointestinal stromal tumors are rare tumors of the GIT accounting for 0.1%-3% of all gastrointestinal tumors. The most common location is the stomach (55%) followed by small bowel (31.8%), colon (6%), other various locations (5.5%), and esophagus (0.7%). They may also occur in extra-intestinal locations. The signs and symptoms of GIST depend on tumor location and size. Gastrointestinal bleeding is one of the most common symptoms. Other signs and symptoms include abdominal discomfort, pain, or distention; intestinal obstruction; and weight loss. The association between the development of GISTs and Neurofibromatosis 1 (NF1) has been established. NF1-associated GISTs tend to have a distinct phenotype and absence of KIT/PDGRF mutations which in turn has an implication on further management where they do not respond well to Imatinib treatment.

Case Presentation: Here we present one of the largest GISTs reported in the currently available literature with a total volume of (25.3 x 20 x 14 cm+ 27.9 x 23 x 8 cm) and an overall weight of 7.3 Kgs, which developed in a 43-year-old female patient with NF1 and was resected on an emergency basis due to the rapid deterioration and development of abdominal compartment syndrome. Pathology assessment showed a malignant GIST composed of spindle cells with elongated nuclei with necrosis, marked pleomorphism and numerous tumor giant cells. The mitotic count was >15/50 HPF, Ki-67 was 80%, and the lymphovascular invasion was clear. Immunohistochemistry investigations showed that Vimentin, CD117, and DOG1 were positive, while BCL-2 and CD99 were focal positives. Pan-CK, S-100, CD34, Desmin, SMA and HMB-45 were negatives.

Biography

Islam Omar, General Surgery Specialist Registrar in Furness, General Hospital, NHS, UK. He has a clinical Master degree in General Surgery from Alexandria University and he is a member of the Royal College of Surgeons of Ireland. He is a Fellow of the European Board of Surgery. He has an active research recently and managed to publish some of his recent papers in recognized international journals

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