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Presacral GIST. Case Review.

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Abstract:

Retro-rectal tumors are a very rare entity in the adult population, with an incidence of approximately 1 in every 40.000-63.000 patients. Gastrointestinal Stromal Tumors (GIST) are the most common mesenchymal tumors of the gastrointestinal tract.

Clinical Case: 51-years-old patient with a clinical scenario of dysuria, vesical urgency and tenesmus, urinary retention, and constipation. Computer Tomography (CT) and Magnetic Resonance Imaging (MRI) shows a presacral mass. Pathology studies report GIST. Treated with abdominal resection and adjuvant therapy.

Discussion: In 1983 the GIST was described with its origin in interstitial cells of Cajal, this tumor is more frequently found in the stomach, appearing in the 4th decade of life and being abdominal pain the most common symptom, the more used diagnostic tools are still the CT and MRI, they let the physician decide which of the three approaching surgical methods is used. There are 3 histologic types GIST and the stratification of risk for each one is given based on tumor site, size, number of mitosis, and rupture.

Keywords: presacral; tumor; constipation.

Introduction

Retro-rectal tumors are a very rare entity in the adult population, with an incidence of approximately 1 in every 40.000-63.000 patients. The majority of the retro-rectal tumors are benign, they are more common in females and generally asymptomatic, being diagnosed incidentally during the proctologic examination. When symptoms appear, their presentation is variable, being pain the most common one. They are divided in congenital (55-65%), neurogenic (10-12%), osseous (5-11%), inflammatory (5%), and others (12-16%) (1). Gastrointestinal Stromal Tumors (GIST) are the most common mesenchymal tumors of the gastrointestinal tract, they are found in adults with more than 50 years old, and its main location is in the stomach (60%) (2). The main treatment is resection of the tumor and also tyrosine kinase inhibitors (Imatinib or Sunitinib) may help with unresectable primary malignancies, metastatic disease or as adjuvant therapy to reduce the grade of recurrence (3).

A case is presented about a presacral GIST in a woman, she was treated with resection of the tumor and adjuvant therapy.

Clinical Case

51-years-old patient with medical history of uterine myomatosis treated with a total abdominal hysterectomy and bilateral adnexectomy, four years ago. She refers 7 months of dysuria, vesical urgency and tenesmus, urinary retention, and constipation with fine bowel movements, occasional rectal bleeding, dyspareunia, rectal tenesmus, and feeling of hypogastric hardening for the last two months.

At the abdominal physical examination, there is a non-mobile, non-tender mass with regular borders at the palpation of the hypogastrium. At the rectal examination, there is normal tone with no masses. Normal laboratory exams. At the MRI there is a voluminous mass with heterogenic signal that occupies the whole extension of the pelvic region with diameters of 112x149 mm in the sagittal sequence, 113x127 mm in coronal sequence, and 113x 123 mm in the axial sequence, bladder displacement, extension of the mass protruding by the rectal canal, in the floor and region of the anal sphincter (Fig 1). In the CT there is a 13 cm pelvic mass with well-defined borders that produces compression and right anterolateral displacement of the rectosigmoid muscle with anterior shift of the bladder (Fig. 2). After a colonoscopy there is evidence of an extrinsic compression of the rectum, in the posterior section (5-15 cm from the anal margin) (Fig. 3). With presumptive diagnosis of retro-rectal (presacral) tumor, surgical resolution is decided and she accepts.

Medical Procedure

Patient under general anesthesia in Lloyd-Davis position, asepsis and antisepsis is done and after that, surgical fields are placed. Starting with a Peri-infraumbilical incision, delving is done by planes through the abdominal cavity, the team observed and palpated a hard mass in the left side of the pelvic region. An incision of the posterior part of the rectum in the presacral space is done, freeing the tumor posteriorly, lateral to the pelvic floor, the vascular plane and left ureter are identified. During dissection, the internal iliac artery is damaged accidentally, but the bleeding is controlled and the mistake is repaired by the vascular surgeon. The ureter was temporally severed, but posteriorly reimplantation by the urologist and placing of double J catheter was done. Hemostasis of the cavity is verified, pneumatic test was done twice to confirm that there were no perforations of the rectum or anal canal, was negative on both occasions. Because of the manipulation of the rectum in the pelvic cavity, it was decided to leave a colostomy in a loop by the left iliac fossa,

hemostasis is verified, and tubular drain was left in the tumor area (15x12 cm) (Fig. 4). Closure by planes was done with conventional technique.

Microscopy indicated constitutional fragments of mesenchymal neoplasia constituted by proliferation of spindle cells with mitotic index of 6m/10cga, necrotic areas and hemorrhages in a 25% of the tumor that formed an ulcer focally to the intestinal mucosa of one of the examined fragments. Surgical margins were compromised by the neoplasia. Immuno-histochemical studies were done, reflecting immunoreactivity for CD117, strong and positive in 80% of extension. Desmin, cytokeratin and S-10 were negative. Diagnosis of GIST of intermedium grade is made. Surgical margins compromised (Fig. 5).

Oncologic treatment with Imatinib for 25 cycles, closure of colostomy was done without complications, satisfactory evolution until the moment.

Discussion

The term GIST was described by Mazur and Clark in 1983, to help with the differentiation between Stromal Tumors with Leiomyomas (3) (4). The origin of these tumors is believe to come from the interstitial cells of Cajal (5) (6).

The GIST can be found in all the extension of the gastrointestinal system, but its most common location is in the stomach (50-60%) and the small bowel (20-30%), additionally these tumors can be found in the colon (10%) and in the esophagus (5%) (7). In the case presented, the patient had a tumor in the presacral space, a rare localization.

In general, the GIST are seen after the 4th decade of life, and the mean age of diagnosis is 60 years old, in the reviewed case, the patient was 54 years old. The most common symptom is upper abdominal pain (50-70%), gastrointestinal bleeding (20-50%), and abdominal mass (5%) (8). The patient in this case had different symptoms: dysuria, vesical urgency and tenesmus, urinary retention, and constipation with fine bowel movements, occasional rectal bleeding, dyspareunia, rectal tenesmus, and feeling of hypogastric hardening.

The diagnostic methods used, were the ones described for presacral lesions: Computed Tomography used to distinguished if a tumor is cystic, solid or mixed, and to evaluate if there is sacral or pelvic invasion. The Magnetic Resonance Imaging is useful to specify the soft tissues planes and assess with certainty its relations with the bones, muscles and nerves. These parameters allow to determine the level and extension of the resection and help to decide the more appropriate surgical approach. In this case the CT with the report of a 13 cm, solid pelvic mass with defined borders, that produces compression and right anterolateral displacement of the rectosigmoid muscle with anterior shift of the bladder and the MRI that reported voluminous mass with heterogenic signal that occupies the whole extension of the pelvic region with diameters of 112x149 mm in the sagittal sequence, 113x127 mm in coronal sequence, and 113x 123 mm in the axial sequence, bladder displacement, extension of the mass protruding by the rectal canal, in the floor and region of the anal sphincter.

As a cellular histological type, GIST are divided in three groups: spindle cell type (70%), epithelioid type (20%), and a rare mixed type (9), in this case review it was reported a tumor constituted by spindle cells. The CD117 (protein) is detected in 98-100% of GIST cases, as in this patient (10).

The National Institute of Health (NIH) shows the GIST risk classification system, based on the tumor location, size, number of mitosis and rupture. It includes very low risk, low risk, intermedium risk and high risk (11). The case presented had a 15x12 cm tumor in a rare location with mitotic index of 6m/10cga and it was classified as a GIST with intermedium risk.

Nakamura et al found no difference in the survival rate between stomach and other locations of GIST (12). Some studies had reported an association between diameter of the mass and its malignancy (13) (14). Despite the large size of the tumor, it was classified as an intermedium risk. Mitosis is an important marker of prognosis, some studies show this (15) (16) and finally there are reports that epithelioid and mixed histological types, have the worst possible prognosis in comparison with spindle cells type (17).

The differential diagnosis of presacral tumors with similar histopathologic findings, includes hemangiopericytoma, malignant mesothelioma, synovial cell sarcoma, and fibrous solitary tumor (18).

Three different surgical approaches are known: abdominal, combined or posterior-abdominal, and trans-sacral. Choosing one of these depends on the size, location and relation of the cyst with the adjacent structures (19) (20). The abdominal procedure is recommended for cysts that are bigger than 8 cm and that have a cranially extension, surpassing the second sacral vertebral body (21) (22). The most utilized technique is the posterior Kraske (23) (24). Böhm and colleagues indicated that for bigger (than 4 or 5 cm) developmental cysts, is necessary the combined or posterior-abdominal procedure (25). Localio and colleagues reported that benign tumors that reach 8 cm of diameter can be resected posteriorly with removal of the coccyx or lower sacrum (26). Over the basis of these considerations in the reported case, it was performed an abdominal intervention because it was a 12 cm tumor. During the surgical act there was a vascular and ureteral infiltration, reason why it was necessary to repair them with success.

Imatinib mesylate is a tyrosine kinase inhibitor that has shown a tumoral response greater than 50% (27), and it has been stablished as standard therapy for patients in whom a primary GIST has been resected (28), that is exactly the same therapy that was applied for this patient in 25 cycles.

The patient is being controlled by the Oncology and Coloproctology Technical Units, in the studies carried out according to the stablished surveillance by the National Comprehensive Cancer Network (NCCN), there was no evidence of changes in the presacral region that could indicate local recurrence.

References

1. Sierra ME, Sierra LG, Leone DG, et al. (2009). Quiste epidermoide gigante del espacion presacro y posanal. Cir Cij. 77:69-72.
2. Demetri GD, Von Mehren, Antonescu CR, et al. (2010). NCCN Task Force report: update on the management of patients with gastrointestinal stromal tumors. J Natl Compr Canc New. 8 (Suppl 2): S1-S41
3. Miettinen M, Lasota J. (2006). Gastrointestinal stroma tumors: review on morphology, molecular pathology, prognosis, and differential diagnosis. Arch Pathol Lab Med, 130(10): 1466-1478.
4. Mazur MT, Clarks HB. (1983). Gastric stromal tumors. Reappraisal of histogénesis. Am J Surg Pathol, 7: 507-519
5. Melis M, Choi EA, Anders R, et al. (2007). Synchronous colorectal adenocarcinoma and gastrointestinal stromal tumor (GIST). Int J Colorectal Dis. 22: 109-14.
6. Efstahios P, Athanasios P, Papaconstantinou I, et al. (2007). Coexistence of gastrointestinal stromal tumor (GIST) and colorectal adenocarcinoma. A case report. World J Surg Oncol. 5:96
7. Dei Tos Ap. (2003). The reappraisal of gastrointestinal stromal tumors: from Stout to the Kit revolution. Virchows Arch. 442:421-8.
8. Sturgeon C, Chejfec G, Espar NJ. (2003). gastrointestinal stromal tumors: a spectrum of disease Surg Oncol, 12: 21-6.

9. Canbak T, Bayraktar B, Acar A, Yigitbasi R. Evaluation of the gastrointetinal stromal tumors for clinical features, histopathological findings and prognostic criteria: A case control-study. North Clin Istanb 2020; 7(2):161-6.
10. Jiang Z-X, Zhang SI, Peng WJ, et al. (2013). Rectal gastrointestinal stromal tumors: imaging features with clinical and pathologicalcorrelation. World J Gastroenterol. 19(20): 3108-3116
11. Li J, Ye Y, Wang J, et al. (2017). Chinese consensus guidelines for diagnosis and management of gastrointestinal stromal tumors. Chin J Cancer Res. 29: 281.
12. Nakamura N, Yamamoto H, Yao T, et al. (2006). Prognostic significance of expresions of cell-cycle regulatory proteins in gastrointestinal stromal tumors and the revelance of the risk grade. Hum Pathol. 37: 503
13. Rudolph P, Chiaravalli AM, Pauser U, et al. (2002). Gastrointestinal mesenchymal tumors inmunophenotypic classification and survival analysis. Virchows Arch. 441: 238-48.
14. Carrillo R, Candia A, Rodriguez-Peralto Jl, et al. (1997). Prognostic significance of DNA ploidy and proliferative index (MIB-1 index) in gastrointestinal stromal tumors. Hum Pathol 28: 160-5.
15. Wang X, Mori I, Tang W, et al. (2001). Gastrointestinal stromal tumors: clinicopathological study of Chinese cases. Pathol Int, 51: 701-6.
16. Emory TS, Sobin LH, Lukes L, et al. (1999). Prognosis of gastrointestinal smooth-muscle (stroma) tumors: dependence on anatomic size. Am J Surg Pathol. 23: 82-7.
17. Miettinen M, Makhlouf H, Sobin LH, et al. (2006). Gastrointestinal stromal tumors of the jejunum and ileum: a clinicopathologic, immunohistochemical, and molecular genetic study of 996 cases before imatinib with long-term follow- up. AM J Surg Pathol. 30: 447-89.
18. Cristi E, Perrone G, Battista C, et al. (2005). A rare case of Solitary Fibrous Tumour of the Pre-sacral Space: Morphological and Immunohistochemical Features. In Vivo. 19(4): 777-80
19. Theodoropoulos D. Gastrointestinal tumors of the Colon and rectum. Clin Colon Rectal Surg 2011; 24(03): 161-70.
20. Martin ME, Pérez SJC, Cotano UJR, Atín del Campo V, Aguinagalde PM,Salas OJA, et al. (2002). Quistes del desarrollo en el espacio presacro en el adulto. Gastroenterol Hepatol. 25:601-604.
21. Malafosse M, Gallot D, Douvin D, Hervé De Sigalony JP. Kystes et. (1977). tumeurs péri-ano-rectaux d´origine vestigiale chez l´adulte. J Chir. 113:351-360.
22. Shimomura M, Goshima H, Katsumine Y, Miyahara S, Usui M. (1996). A case of presacral epidermoid cyst. Nihon Rinshou Geka Gakka Zasshi. 57:727-731.
23. Abel ME, Nelson R, Prasad ML, Pearl RK, Orsay CP, Abcalian H. (1985). Parasacrococcygeal approach for the resection of retro-rectal developmental cysts. Dis Colon Rectum. 28:855-858.
24. Hjermstad BM, Helgwig EB. Tailgut cysts. (1988). Report of 53 cases. Am J Clin Pathol. 89:139-147.
25. Böhm B, Milsom JW, Fazio VW, Lavery IC, Church JM, Oakley JR. (1993). Our approach to the management of congenital presacral tumors in adults. Int J Colorectal Dis. 8:134-138.
26. Localio SA, Eng K, Ranson JHC. (1980). Abdominosacral approach for retrorectal tumors. Ann Surg. 191:555-560.
27. Spinelli GP, Miele E, Tomao F, et al. (2008). The synchronous occurrnence of squamous cell carcinoma and gastrointestinal stromal tumor (GIST) at esophageal site. Worl J Surg Oncol. 6: 116.
28. De Matteo RP, Ballman KV, Antonescu CR, et al. (2009). Adjuvant imatinib mesylate after resection of localised primary gastrointestinal stromal tumor: a randomised doublé-blind placebo-controlled trial. Lancet. 373: 1097: 104