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LEIOMYOSARCOMA OF THE RECTUM: CASE REPORT AND REVIEW OF LITERATURE

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ABSTRACT

Leiomyosarcoma of the rectum is rare tumor occurring at all ages and in both sexes, but it is more common beyond the sixth decade of life and represents approximately 0.3% of rectal malignancies. The standard treatment is surgical resection with or without radiotherapy & chemotherapy for patients with operable tumors. Prognosis is poor. Survival rates vary and depend of tumor staging. We report a patient case complaining of recurrent episodes of rectal bleeding and pelvic mass due to a rectal leiomyosarcoma, he had surgery with sphincter preserving. We discuss particularities of this rare histological type of tumor.

INTRODUCTION

Rectal leiomyosarcoma is a rare tumor of smooth muscle origin, accounting 0.3% of all rectal malignancies., those tumors occur at all ages and in both sexes but They are more common in females and beyond the sixth decade of life. Diagnosis is difficult without proper immunohistochemistry, Leiomyosarcoma classically stain with, smooth muscle actin, vimentin and desmin. Bleeding, constipation, and rectal pain are the most common symptoms of this disease. Leiomyosarcoma of the rectum grows slowly and metastasizes late. Spread is by direct extension into contiguous structures and by bloodstream to liver, lungs, brain, and bone. Lymphatic metastases are rare. Blood-borne metastases to liver and lung are the most common cause of death. Besides its rarity, optimal treatment of rectal leiomyosarcoma is still not clear; abdominoperineal resection is still the procedure of choice. Local excision might be considered for small lesions of low-grade malignancy in hight-risk patients. Radiotherapy and chemotherapy are of limited benefit. Herein we report a case of rectal leiomyosarcoma with unusual presentation.

Case Report

A 54-year-old female was admitted to hospital presenting with a low abdominal mass, bleeding the 4 days preceding her admission, a feeling of incomplete evacuation after a bowel movement, tenesmus, and a weight loss of 10 kg in the previous month. Rectal digital examination found a hard, non-mobile mass in the anus at 4 cm of the anal margin. General examination, a complete blood cell count, blood chemistry and coagulation studies were normal, except for anemia (haematocrit, 32%) and an increase of white blood cell count. The erythrocyte sedimentation rate was also elevated (81 mm/h).

The findings of preoperative ultrasonography tomography were suggestive of a malignant pelvic mass due to intestinal tract, CTscan of the abdomen and pelvis revealed an approximately 13x10 cm heterogeneous solid mass occupying the entire pelvis (photos 1, 2). Colonoscopy was unsuccessful because of stenosis of the lumen at 4 cm from the anal margin; there is also a protrusion of the rectal mucosa into the lumen, probably because of extrinsic compression. Biopsy shows spindle cells proliferation with malignancy. The patient underwent curative radical surgical resection, including extended low anterior resection whith colo-anal anastomosis; ileostomy was also performed.

The postoperative course was uneventful. Microscopically, the tumor was composed of spindle cells with enlarged hyper chromatic nuclei and increased mitotic activity. The tumor was immunoreactive for vimetin, weakly reactive for actin and desmin.

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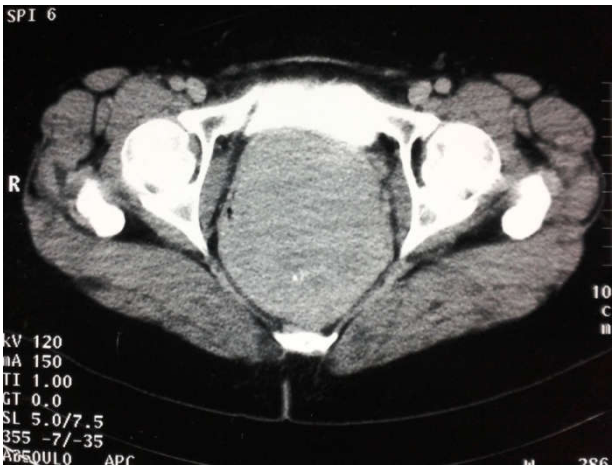

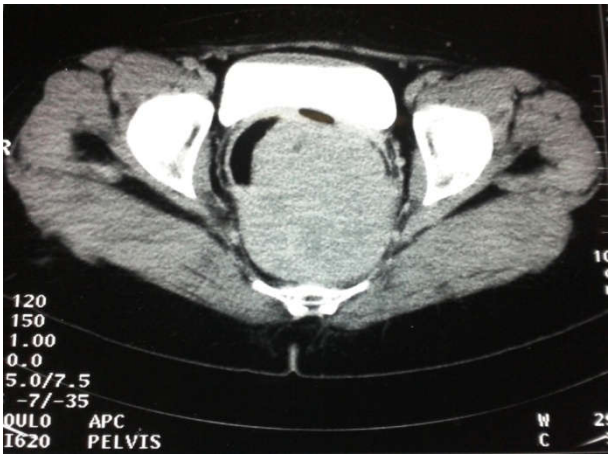

Photos 1 and 2. CT scan of pelvic zone showing heterogeneous solid mass occupying the entire pelvis

DISCUSSION

Primary gastrointestinal (GI) sarcomas are a very rare entity, accounting for 1-2% of GI malignancies. leiomyosarcoma is the most common histiotype in this group. It is a mesenchymal tumor of smooth muscle origin. Leiomyosarcoma of the rectum represents approximately 0.07% to 0.1% of gastro-intestinal malignancies (Khalifa *et al*., 1986; Moore and Hilbun, 1986). In a review of 81,000 cases of malignancy entered into the Armed Forces Central Medical Registry, Feldtman *et al*. (1981) found seven cases of leiomyosarcoma of the rectum. This neoplasm accounted for 0.06% of all gastrointestinal malignancies and 0.3% of all rectal malignancies. Bleeding, constipation, and rectal pain are the most common symptoms. In addition, the patient may complain of tenesmus, diarrhea, and dysuria, (Khalifa *et al*., 1986). The majority of lesions are palpable on digital examination. They vary in size and are predominantly endorectal, in the more malignant forms they have extensive perirectal invasion (Khalifa *et al*., 1986; Moore and Hilbun, 1986; Feldtman *et al*., 1981; Labow and Hoexter, 1977).

Because these tumors are intramural in origin and tend to grow extraluminally, biopsy findings from the luminal wall obtained from over the tumor often will be reported as benign mucosa. Endoscopic ultrasound with guided biopsy may lead to diagnosis; Leiomyosarcomas are identified as having inhomogeneous echoes and irregular borders.

Unfortunately, early detection of the tumor with CT scanning depends on the size and may not be helpful, but MRI can make the diagnosis even for leiomyosarcoma with small size.

The development of intrarectal ultrasonography has introduced a new dimension in the treatment of patients with rectal leiomyosarcomas. This diagnostic modality has proven to be a useful tool in the preoperative assessment of patients who might be candidates for local forms of therapy. The layers of the rectal wall can be identified and the depth of penetration determined. The status of lymph nodes also can be assessed. Most lesions appear as submucosal masses and approximately 44% are ulcerated. The consistency may be firm, soft, or hard (Khalifa *et al*., 1986; Moore and Hilbun, 1986; Feldtman *et al*., 1981; Labow and Hoexter, 1977; Akawri *et al*., 1978; Kessler *et al*., 1996; Randleman *et al*., 1989; Yeh *et al*., 2000; Thalheimer *et al*., 2008). Histologically, interlacing bands of distinct, spindle shaped cells are seen. The criterion of malignancy is simple in the high-grade forms. It is based upon the number of mitoses per high-power field (Akawri *et al*., 1978). It is on this definition that the decision for a more definitive operation is based.

Histologic features under light microscopy are the most important factors in making the diagnosis of leiomyosarcoma. However, adjunctive modalities including immunohisto chemisty and electron microscopy play an important confirmatory role. Immunohisto chemistry helps support the diagnosis by demonstrating the presence of muscle specific markers including: desmin, muscle specific antigen (HHF35), cytokeratin (CK) and S100. While not required to make the diagnosis, one or more of these markers is usually found in specimens of leiomyosarcoma.

Electron microscopy is useful in further elucidating the classic nuclear morphology seen in this tumor. Cytogenetic analysis of large series of soft tissue sarcoma, including leiomyosarcoma, has not shown a consistent chromosomal aberration or translocation. CD117 and CD34 markers are negative eliminating stromal tumor (GIST). Leiomyosarcoma of the rectum grows slowly and metastasizes late. Spread is by direct extension into contiguous structures and by bloodstream to liver, lungs, brain, and bone and are the most common cause of death. Lymphatic metastases are rare (Moore and Hilbun, 1986; Campos *et al*., 2004).

Attempts have been made to modify the treatment according to the histologic classification of the neoplasm. Because of the difficulty in determining malignancy, supposedly benign lesions treated by local excision have a high recurrence rate. When local recurrence develops and in lesions diagnosed primarily as malignant, the treatment of choice is wide local excision rather than lymphatic clearance, but abdominoperineal resection may be necessary. Radiation therapy is an important additional treatment for improving rates of local control when surgical margins are close, especially in high-grade sarcomas. Radiation therapy can be delivered either pre-operatively (neoadjuvant) or post-operatively (adjuvant). Radiation therapy can also be utilized as a means of palliative local control in cases where extensive metastasis has already occurred.

Survival is generally poor, with a five-year survival rate between 20% and 40% and local recurrence rates as high as 20% to 86% (Kessler *et al*., 1996). In a review of the literature, Labow and Hoexter (1977) calculated a 20% five-year survival rate. Randleman *et al*. (1989) reported 5-year and 10-year survival rates: 62% and 40%, respectively. Adjuvant therapy had no effect on survival. The authors concluded that lesions less than 2.5 cm in size can be treated by wide local excision, whereas larger lesions require radical resection. In a review of the literature involving 135 cases, Khalifa *et al*. (1986) concluded that abdominoperineal resection is still the procedure of choice. Local excision might be considered for small lesions of low-grade malignancy in poor-risk patients. Radiotherapy is of limited benefit. Chemotherapy [doxorubicin (formerly adriamycin)] may prove effective in one-third of cases (Khalifa *et al*., 1986). Yeh *et al*. (2000) presented the prognosis after resection of 40 patients with rectal leiomyosarcoma: 48% developed recurrence or metastasis postoperatively. There was a strong trend toward higher local recurrence rates for the local excision group than for the radical resection (55% vs. 24%). There was no difference in the incidence of distant metastasis between the two groups with different operation types. The metastasis rates of the wide local excision and radical excision groups were 27% and 38%, respectively.

Conclusion

Sarcoma is a rare subgroup in colorectal neoplasm and leiomyosarcoma is presently the most common type of colorectal sarcoma. Leiomyosarcoma has a tendency of recurrence. No reliable tumor markers or useful adjuvant chemotherapy are as yet available, making close image follow-up a necessity for the management of this disease. Survival is generally poor, with a five-year survival rate between 20% and 40% and local recurrence rates as high as 20% to 86%.

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