Research Article

iMedPub Journals
www.imedpub.com

Medical Case Reports
ISSN 2471-8041

2022
Vol.8 No.1:215

Gastrointestinal Stromal Tumour of Anal Canal; A Case Report and Literature Review

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Received date: May 2, 2021; **Accepted date:** January 3, 2022 **; Published date:** January 10, 2022

Citation: Muhammad Ezzra Shah RS (2022) Gastrointestinal Stromal Tumour of Anal Canal; A Case Report and Literature Review. Med Case Rep Vol.8 No.1.

Abstract

Gastrointestinal Stromal Tumour (GIST) is a mesenchymal neoplasm that expresses KIT or CD117 receptor. It constitutes 1% of all gastrointestinal tumours. Common anatomical distribution includes stomach (50-60%) and small bowel (30-40%). Less common location reported includes colon (7%), and esophagus (1%). GIST of anal canal is rare and only a few cases have been reported in literature. Surgical treatment is preferred in resectable tumours. Selective tyrosine kinase inhibitors is the current standard of care in non-resectable cases. Radiation therapy has been applied to non-resectable GIST with variable outcomes. We present a case of 50-year old male with perianal swelling and bleeding of 1 month duration. Colonoscopy showed circumferential growth 0.5cm from anal verge. Histopathology is consistent with mesenchymal tumour positive for CD117, CD34, and Caldesmon. Computer Tomography (CT) scan showed local infiltration to presacral space, mesorectal fascia and poor plane with prostate gland and levator ani. No distant metastasis noted. He underwent local radiotherapy and given Imitinib and is currently awaiting reassessment for response to therapy pending further intervention.

Keywords: Gastrointestinal Stromal Tumour, Anal GIST

Introduction

Case Presentation

A 50-year-old gentleman presented to us with complained of perianal swelling for 2 years duration but increasing in size for the past 1month. It was associated with perianal pain and fresh per rectal bleeding for 2weeks. He also complained of having mucoid stool, alternate bowel habit, and mild grade fever, loss of appetite and significant loss of weight. He has no family history of cancer and no other premorbids. He has been smoking cigarettes for the past 20years. Upon clinical examination, there was a perianal swelling at 5o'clock, which was hard in consistency, irregular surface but no contact bleeding. Per rectal examination noted hard mass presence circumferentially and tender on palpation. There was no enlarged inguinal lymph nodes and on abdominal examination there was no mass palpable and no hepatosplenomegaly.

Colonoscopy done on October 2016 showed circumferential mass 0.5cm from anal verge which was fungating in nature. Multiple biopsies were taken and histopathological examination showed that the tumour tissue is composed of spindle-shaped cells arranged in short fascicles. The spindle cells display oval, elongated uniform nuclei, fine chromatin pattern, paranuclear vacuoles and eosinophilic cytoplasm. Nuclear palisading is seen. Mitosis is 1/10 HPF. Immunohistochemistry showed positive CD117, CD34 and Caldesmon. Hence the features are consistent with GIST. Computed Tomography (CT) scan of Thorax, Abdomen and Pelvic noted circumferential enhancing bowel mass seen arising from the anal verge until rectum measuring 4.9cm (W) x 7.5cm (AP) x 5.1cm (CC) causing luminal narrowing. Presacral space, mesorectal fasciae and fat streakiness seen. No fat streakiness in both ishchioanal fossa. Engorged pelvic vessels. Poor fat plane with the prostate gland and levator ani at lower anal region. Multiple subcentimetre paraaortic, aortocaval, mesenteric and both inguinal lymph nodes. No distant metastasis.

He is currently receiving radiotherapy and was started on Imitinib. He is pending for another re-evaluation after radiotherapy for a definitive surgical intervention.

Discussion

Gastrointestinal Stromal Tumour (GIST), is a mesenchymal tumor that expressing KIT tyrosine kinase and showing presence of platelet-derived growth factor alpha, (PDGFRa). It was initially was classified as leiomyomas/leiomyosarcomas because they possessed smooth muscle features when examined under microscope. Not until 1983 when it was then described as stromal tumour. In 1998, Hirota et al discovered C-kit proto-oncogenes mutations in these tumours which lead to a new classification of GIST. Later in 2003, Heinrich et al found out that GIST too contained mutation of PDGFRa gene. These advances have led to the classification of GIST as an entity separate from smooth muscle tumours. These new understanding have also helped explicate their etiology and pathogenesis at molecular

level and led to the development of molecular-targeted; Imitinib therapy for this disease.

GIST accounts for less than 1% of GI tumours, but they are the most common mesenchymal neoplasm of the GI tract. It is commonly found in stomach or small intestines, and incidence of anal GIST is very rare with only few publications noted in English literature. Because of its rare entity, there was no proper guidelines in managing anal GIST.

Ten cases of anal gist reviewed from 9 case reports published in English literature. Male has more preponderance with 7 of them and 3 of the cases are females. Age ranges from 60 to 80years old. However, in our case, the patient is only 50years old which makes the incidence in the younger age group is rare. Clinical presentations are varies and they share similarity to those with anal carcinoma. Per rectal bleeding is the most common presentations followed by anal mass and anal pain. Other presentations include constipation and alternate bowel habit. Most GIST are benign but it does carries malignant potential. A prediction scheme has been constructed by Flectcher et al which helps in predicting tumour's behaviours and its risk of recurrence. Tumour size of <2cm and mitotic rate of <5 in 50 high power field (HPF) has a very low risk. It is widely accepted that criterias to predict GIST malignancy are tumout size of >5cm with mitotic rate of >5/50HPF or size > 10cm with any mitotic rate as proposed by Fletcher. Changchien CR et al has included age of less than 50 of age, higher histologic tumour grade, and larger tumour of >5cm as factors that contributes to poor prognosis. In the series of case reports, there are 2 cases were noted to have recurrence. Both cases have tumour size of more than 5cm and one case noted to have high mitotic rate of 5/50HPF. One case had pelvic exanteration and noted to have recurrence after 24months and another case had only transanal excision and recurrence noted after 30months of follow up.

Conclusion

In all case reports, all patients undergone surgical resection. Most cases had undergone abdominoperineal resection. In all three cases who had non radical resection, none of them had local-regional invasion. One case who had non radical resection had recurrence as stated and the other 2 cases have no recurrence after 1-2years follow up. Only 3 cases stated that patients received imitinib as adjuvant therapy and none of the them received any radiotherapy.

For our case, who is a young 50 year old gentleman, who has tumour size of 4.9cm (W) x 7.5cm (AP) x 5.1cm (CC), with locoregional invasion to surrounding tissue and subcentimer nodal involvement, confirmed diagnosis of GIST with positive CD117 and has mitotic rate of 1/10HPF, he has a poor prognosis and high rate for recurrence. The options for him would be pelvic exanteration in view of localregional invasion to prostate. So far there is no literature noted for the use of radiotherapy for anal GIST. However the use of radiotherapy for rectal GIST has been discussed in literatures to may have benefited to post resection patients who had involved margins. Studies done at Princess Margaret Hospital by Crosby JA et al on patients of rectal GIST provides evidence that GIST is a radiosensitive tumour. However contradicting to this finding is a study by Pierrie JP et al in Massasuchates General Hospital in which 20patients who have intermediate and high grade tumour following incomplete resection received radiotherapy and they concluded that there is no advantage in disease specific survival. Although, the authors note that those who received radiotherapy were likely had more aggressive disease. Another 2 publications noted the use of radiotherapy and imitinib as adjuvant for rectal GIST by Boruban et al and Ciresa et al. Both cases showed tumour regression and marked clinical responses following radiotherapy and Imitinib.

Currently our patient is undergoing radiotherapy and also on imitinib. He will be follow up to determine its response and will be planned for definitive surgery. Hopefully if it is responded well with radiotherapy, he can avoid a radical surgical resection.

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