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Case Report

Duodenal leiomyosarcoma: a rare entity

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ABSTRACT

Tumours of the small intestine are among the rarer conditions as compared to other tumours of the gastrointestinal tract. Among these, sarcomas hold a much lesser number in proportion to adenocarcinomas. Leiomyosarcomas occurring in the small intestine constitute an even smaller number, accounting for about 1% of malignant mesenchymal lesions in the gastrointestinal tract. Within the spectrum of LMS of the small intestine, the more common location encountered is the ileum, followed by the duodenum. The second part of the duodenum is usually implicated in most studied and reported cases of duodenal LMS. These tumours are known for their aggressive nature and their often-delayed presentation due to ill- defined symptomatology and diagnostic challenges. As a result of the above, even a century after the first reported case, their prognosis is poor. Here we report a case of a 64-year-old female who presented with vague complaints of abdominal discomfort and dyspepsia for a month and was diagnosed to have a mass lesion in the 2nd and 3rd part of the duodenum. Intraoperatively, she was found to have two separate mass lesions in D1 and D3 and was subjected to a pancreaticoduodenectomy. The mass lesions were pathologically categorized as leiomyosarcomas of the first and third part of the duodenum.

Keywords: Duodenum, tumor, Leiomyosarcoma, Pancreaticoduodenectomy

INTRODUCTION

Vague abdominal pain or discomfort is often dismissed by most patients as a minor ailment, and once a certain degree of symptomatic relief is obtained, it is altogether forgotten. However, this may be an innocuous manifestation of certain insidious entities, including very rare diseases such as leiomyosarcomas.[1-3]

A leiomyosarcoma is a rare malignancy arising from the smooth muscles, mostly encountered in the retroperitoneal space, vascular wall, and soft tissues of the lower extremities.[4] GISTs are the most common of the malignant mesenchymal tumours, while these tumours, a type of non-GIST sarcomas, are rare tumours that occur anywhere in the gastrointestinal tract, though mostly described in the stomach and small bowel. Among gastrointestinal malignancies, these constitute a rare subset, and mostly occur in the ileum. When occurring in the duodenum, they usually involve the second part most commonly (53%) followed by 27% in the third part, and 5-10% in the first and fourth part.[5] Because these usually have non-specific symptoms and endoscopic biopsies do not yield definitive diagnoses, the presentation and hence diagnosis is often delayed and challenging. We present a case of a patient with vague abdominal discomfort plaguing her for a period of 2 months which was later found to be caused by multiple duodenal leiomyosarcomas.

CASE REPORT

A 64-year-old female with no known previous illnesses presented to us with complaints of obscure abdominal

discomfort and symptoms suggestive of dyspepsia, in the last two months. She had complaints of on and off upper abdominal pain that showed no periodicity or relation to food intake. She did not complain of abdominal distention or vomiting, nor had she perceived any mass per abdomen. She reported no weight loss. Despite two months of symptomatic management, her symptoms had not abated, hence she came for further evaluation. On admission, her blood pressure was 130/80, pulse rate was 78/min, and she was afebrile. General physical examination revealed pallor. Abdominal examination revealed a vague mass of 5×7 cm in the right upper quadrant. Laboratory investigations revealed a hemoglobin of 8 g/dl, and albumin of 2.8 g/dl. Other lab values were within the normal range. Peripheral smear revealed a microcytic hypochromic pattern of anemia. Contrast enhanced computed tomography of the abdomen and pelvis revealed a well-defined hypodense mass measuring 4.9 (AP) × 7.9 (T) × 5.4 (CC) cm arising from the lateral wall of the 2nd part of the duodenum and 3rd part of the duodenum, the lesion showing heterogenous enhancement on contrast administration. The lesion was causing compression of the duodenal lumen with the duodenum appearing dilated. Concurrently, a posterior wall uterine fibroid and a hepatic cyst were noted. The finding was radiologically opined to be submucosal in origin and was noted to be causing aneurysmal dilatation of the duodenum. An upper GI endoscopy was undertaken, where a semi-circumferential extraneous impression with minimal luminal narrowing of the overlying mucosa was noted at the level of the 2nd part of the duodenum. A working diagnosis of duodenal GIST was arrived at.

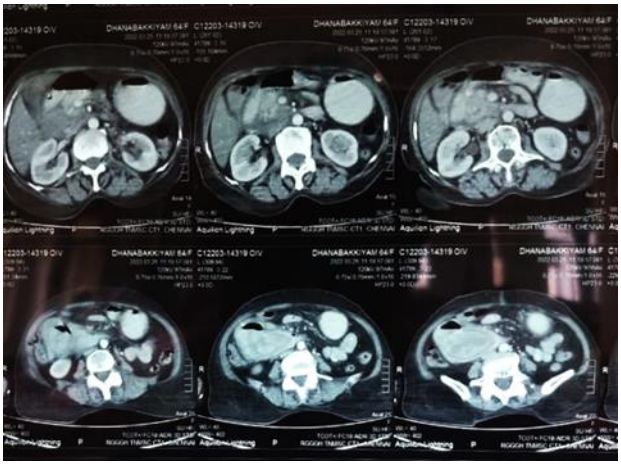

Figure 1: CECT images showing mass lesion in the 2nd and 3rd part of the duodenum.

The patient was optimized and taken up for surgery. Intraoperatively, two lesions were noted in the duodenum, the larger in D3 and the smaller in D1. Due to segmental resection not being feasible, the operative team proceeded with a pancreaticoduodenectomy followed by a pancreaticojejunostomy, hepaticojejunostomy and gastrojejunostomy with a feeding jejunostomy.

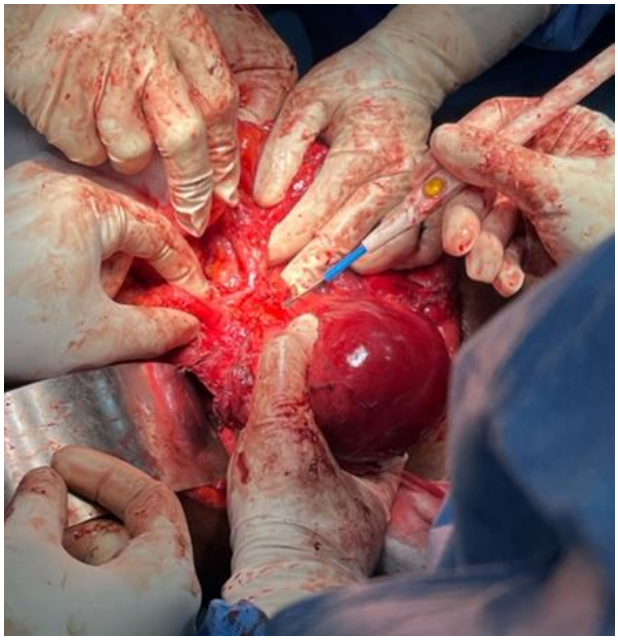

Figure 2: Larger mass lesion arising from the 2nd part of the duodenum.

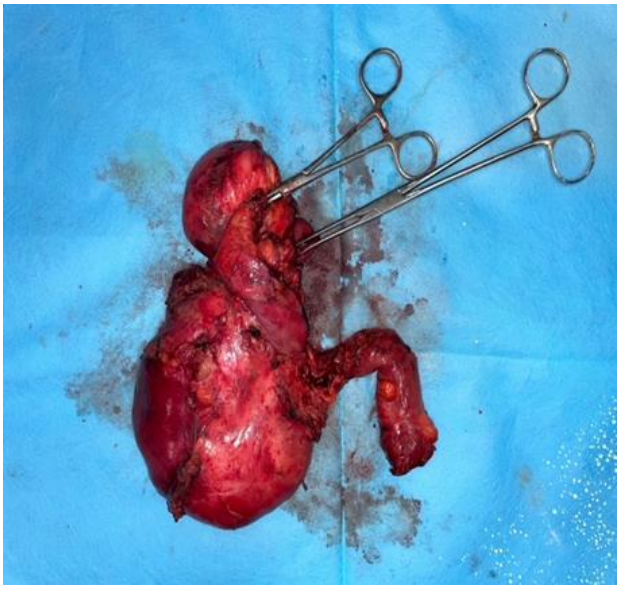

Figure 3: Specimen post Whipple's procedure with lesions in D1 and D2.

Macroscopic examination of the specimen a mass arising from the submucosa of the duodenum measuring 9.5×6×4.5 cm and another mass noted in the serosal aspect of the duodenum of 6×5×3.5 cm. Histological type was noted to be Leiomyosarcoma, Grade 2, with all margins free from tumour and no nodal positivity. IHC showed strong MSA positivity. Post-operatively, the patient suffered from a dehiscence of the GJ anastomosis and was hence taken up for re-exploration, where the GJ was re-done. However, patient continued to deteriorate due to poor general condition and succumbed 3 months after the initial Whipple's resection.

DISCUSSION

Duodenal leiomyosarcomas primarily occur in the 50-60 age group with a slight female preponderance. The first case of such a disease entity was observed by Von Salis in 1920.[3] Since then, until today, only close to 200 cases have been observed in literature.[2] Out of these, most occur in the 2nd part of the duodenum. Simultaneous occurrence of two lesions in the first and third part has not been published yet. The symptomatology of these tumours continues to be very imprecise, those being abdominal pain, weight loss, or that related to gastrointestinal bleeding.[6] Diarrhea related to pancreatic dysfunction, fever secondary to central necrosis of tumour, obstruction of the GIT or the biliary tree are very infrequent presentations.[6] An abdominal mass is palpable only in 40% of the cases. The symptoms were noted to be of such nature due to the tendency of the tumour to grow extraneously. These tumours also demonstrate high vascularity.

The diagnosis of these tumours continues to be a task, partly because there are no specific radiological signs. Radiology may demonstrate a mass lesion arising from the submucosa, however that is highly non-specific. Another constraint in terms of diagnosis is that even if accessible endoscopically, due to the extraluminal nature of the lesion, biopsies often return inconclusive. Hence it is exceedingly difficult to make a pre-operative diagnosis of this rare disease.

It often presents as a locally advanced disease with involvement of adjacent organs, with spread to distant organs noted only in 20% of the cases.[1] The spread is most often hematogenous, with lymphatic spread almost non-existent. The spread occurs mostly to the liver. Due to the lack of proven effective adjuvant therapy, surgery is the mainstay in the management of said lesions. The type of surgery undertaken depends upon the location of the tumour and the size of the tumour, rather than on the need to perform a nodal dissection. For small tumours, and those located in the distal duodenum (D3 and D4), tumour excision or segmental resection of the involved portion of the duodenum with primary anastomosis is recommended. For larger tumours which were not amenable to the above- mentioned approaches, or for tumours involving the proximal duodenum, nothing short of a pancreaticoduodenectomy has to be undertaken.[5] In our case, since the duodenum was diffusely involved by large lesions, we had to proceed with a Whipple's resection. Post-operative tumour recurrence has been noted in both segmental resections as well as pancreaticoduodenectomy, but less commonly noted with simple excision, probably accrued to the stage of the disease. However, this goes on to highlight that the impetus remains on the attainment of negative margins rather than a thorough lymphadenectomy.[7] Post-operative care is of paramount importance in patients undergoing surgery for this disease, which constitutes a major surgical procedure. Nutritional supplementation needs to be adequate and more to allow the patient to recuperate.

CONCLUSION

Duodenal leiomyosarcomas are rare tumours that, even after all the time elapsed since the first reported case, continue to be diagnostic and therapeutic dilemmas. These are often diagnosed at advanced stages due to the indolent course of the illness. An aggressive surgery remains the dictum in the management of this disease entity.

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