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Case report of a gastrointestinal tumour (Carcinoid Tumour): A diagnostic delima

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ABSTRACT

Background: Carcinoid tumors are rare gastrointestinal malignancies and are slow growing compared with adenocarcinomas. Because they rarely invade the intestinal mucosa, they are extremely difficult to diagnose. The aim of this case report is to emphasize the difficulty in making a clinical diagnosis of this tumor and require a high index of suspension.

Method: The case records of a 42-year-old widow who present with a history of intermittent profuse diarrhea for one year. Each episode was trigger by eating either Bananas, pineapple, avocado pears or food containing too much of tomatoes and was diagnosed to have ileal carcinoid tumors on laparotomy. A review of literature was done using med line and manual library search.

Results: The patient was admitted in the female ward for 2 days and was treated conservatively with good intravenous fluid of 6 liters of normal saline. She got better and was discharge home. Two weeks later, she came back with similar complain after she ate bananas and pineapple fruit drink. An abdominal ultrasound scan was ordered which was of no use to the diagnosis. An exploratory laparotomy was carried out where a small mass was found located in the medium. Ileal resection and end-to-end anastomosis were done. There was good wound healing and was discharged 12 days after laparotomy. She was well clinically at follow up review one month after discharge.

Conclusion: The case illustrates the difficulty in making a diagnosis and therefore requires a high index of suspicion before surgery.

Keywords: ileal carcinoid, difficult clinical diagnosis, high index of suspicion

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INTRODUCTION

Carcinoids tumors are rare and slow growing as compared with adenocarcinomas they can arise from several places throughout the body, carcinoid tumor is a subset of neuroendocrine tumor commomly affected organ is the digestive system or the lungs1.2 The neuroendocrine tumor is derive from the enterochromaffin cell [1]. Carcinoid tumors don't often cause sign and symptoms, until late in the cause of disease. Carcinoid tumors are scare tumors; with only 10% of neuroendocrine tumors constitute carcinoid syndrome [2]. Epidemiology, the age-adjusted incidence of nonpancreatic neuroendocrine tumors is 4.7 per 100,000. The incidence of neuroendocrine tumors, including nonpancreatic and pancreatic neuroendocrine tumors, increased from 1.09 to 5.25/100,000 between 1973 through 2004 (per data from SEER). This increase in incidence in the past decades is likely due to an increased number of endoscopic and radio-imaging studies. Incidence varies by gender and race. Recent data suggest the incidence of neuroendocrine tumors is higher in black males compared to Caucasians (6.46 versus 4.6/100,000). The ratio of tumor incidence among males and females is almost equal, with slightly higher in males. The median age of diagnosis of neuroendocrine tumors is between 55 and 60 [2]. In the gastrointestinal tract, they arise from foregut, midgut and hindgut [3, 4]. They arise most commonly in the ileum (25%) followed by the rectum (14%) and appendix (12%). in these sites, they may present with a mass which may bleed, perforate or obstruct the intestine. A clinical diagnosis is difficult to make except in the presence of the carcinoid syndrome. The diagnosis is sometimes made incidentally during an emergency laparotomy when there is intestinal obstruction. Early detection and resection offer the best chance of cure. The larger the tumor greater than 2 cm in diameter the worse the prognosis. The pathophysiology of carcinoid syndrome is based on biologically active amines and peptides entering the systemic circulation and escaping the first-pass metabolism of the liver. Usually, these bioactive products are inactivated in the liver. However, in the cases of neuroendocrine tumors with liver metastasis, either these bioactive products directly enter into the systemic circulation or are not inactivated due to deranged liver function. Less frequently, carcinoid syndrome can occur without liver metastasis in conditions such as primary gut tumors with widespread retroperitoneal nodal metastases, ovarian tumors, or bronchial carcinoids, which release bioactive amines directly into the systemic circulation [1]. Neuroendocrine tumors release approximately 40 types of

biologically active amines and peptides. The most common ones are serotonin, histamine, tachykinins, kallikrein, and prostaglandins [3]. Most of the clinical features are due to serotonin, which is an end product of tryptophan metabolism. Usually, only 1% of dietary tryptophan is metabolized into serotonin. However, in the cases of neuroendocrine tumors, up to 70% of tryptophan is metabolized into serotonin. Serotonin undergoes oxidative reactions that lead to the formation of 5-Hydroxy Indoleacetic Acid (5-HIAA) with the help of aldehyde dehydrogenase, which is then eliminated in the urine. Serotonin causes increased motility and excessive secretion of the gastrointestinal tract, leading to diarrhea. As most of the tryptophan is diverted to the serotonin formation pathway by neuroendocrine tumors, it results in a deficiency of tryptophan, which is needed for the synthesis of niacin. Consequently, deficiency of niacin leads to Pellagra, which manifests as a triad of dermatitis, dementia, and diarrhea. Prostaglandins also play their part in increased intestinal motility and fluid secretion in the gastrointestinal tract, causing diarrhea. Neuroendocrine tumors of the foregut and lungs lack the enzyme aromatic L-amino acid decarboxylase, which metabolizes 5-hydroxytryptophan to serotonin. Thus, the neuroendocrine tumors of the lungs and foregut do not produce serotonin. On the other hand, hindgut neuroendocrine tumors usually do not produce any bioactive hormone. Pulmonary neuroendocrine tumors mainly produce histamine, which can cause atypical flushing and pruritus. Tachykinins (substance p, neurokinin A, neuropeptide k) are also responsible for causing flushing due to their vasodilatory effect. Carcinoid tumors are majorly classified according to either embryologic origin or the vascular supply of the gastrointestinal tract (foregut midgut hindgut)

CASE PRESENTATION

A 42-year-old widow attended the family Medicine out-patient clinic of University of Calabar Teaching Hospital on account of intermittent profuse diarrhea for one year. She also mentioned general body weakness and abdominal discomfort. Each episode is triggered when she eats bananas, pineapple or avocado pear. She has taken many drugs known or suggested to stop diarrhea including herbal preparations. No other associated symptoms.

Examination showed a middle age woman with moderate dehydration and was not clinically anemic. The watery stool was taken for laboratory test which include food poisoning Helminthics and other parasites. Blood was taken for urea and electrolytes and all the test results were all normal.

When she was resuscitated by correction of fluid and electrolytes, she became better and was discharged after staying in the ward for 2 days.

Two weeks later she came back to the clinic with similar symptoms of profuse diarrhea. This necessitated a more detailed investigation. A number of investigations were requested, stool analysis for parasites. HIV, cholera virus. Abdominal TB was also screen. Abdominal ultrasound scan was again ordered. The entire above test turns out to be normal and was not helpful for the diagnosis. I also thought of other differential diagnosis such as coeliae disease or tropical spruce but the facilities to test for them were not available. An exploratory laparotomy was carried out where a small mass was found in the midileum. A resection with end-to-end ileo-ileal anastomosis was done. The mass was sent to Histopathology laboratory for report. Post-operative condition was satisfactory. She was discharging home 12 days after laparotomy with good wound healing. Histopathological report read carcinoid tumors cells. She was reviewed at 6 weeks and 3 months subsequently. On both occasions her clinical condition improved without a recurrence of profuse diarrhea symptoms (Figures 1 and 2).

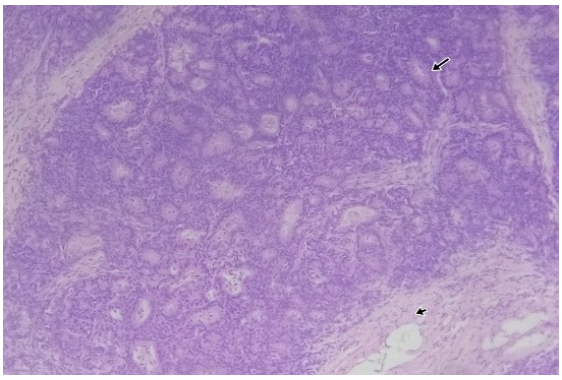

Fig. 1. Section shows a tumour displaying organoid, trabecular and nested pattern with rosette formation

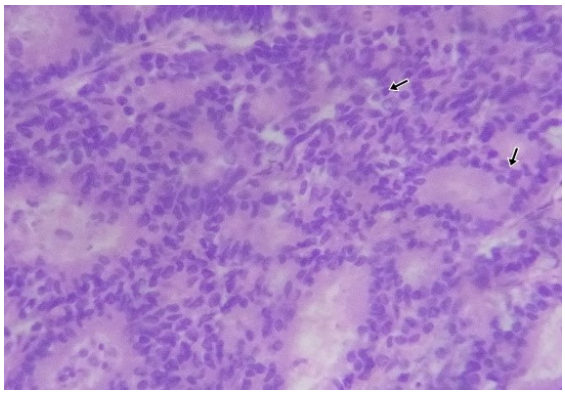

Fig. 2. The tumor cells are uniform polygonal shape having round to oval nuclei with salt and pepper chromatin (x 100, x 400)

DISCUSSION

Carcinoids tumors occur commonly in the gastrointestinal tract. The midgut is the most common site, specifically the ileum. These tumors have been described in other sites such as the bronchus, ovary, breast, testis, cervix, pancreas, larynx and prostate [5-9]. The usual distribution in the gastrointestinal tract had been described as appendix (65%) ileum (25%) and the rectum, colon, stomach and duodenum share the remaining (10%). Current statistic shows that they are most commonly found in the ileum (25%), rectum (14%) and the appendix (12%) [2]. Intestinal carcinoids are usually small tumors with a mean diameter of 1cm; the largest tumors have about 3 cm in diameters [10-12].

Gastrointestinal carcinoids are difficult to diagnose in the absence of the carcinoid syndrome. Because they are often covered with normal mucosa. Also, they are slow growing tumors. So that about 50% of individual with carcinoid tumors is asymptomatic just as our index patient. The clinical presentation is usually non-specific and patients go about undiagnosed for many years just as our index patient.

The patient in this case report exhibited the slow growing characteristic of the condition. Profuse diarrhea is one of the features encountered which were occasionally triggered by eating bananas, pineapple or avocado pear.

Apart from abdominal ultrasound scan, other investigations such as Computed Tomography (CT scan), Magnetic Resonance Imaging (MRI) and Urine 5- Hydroxyindole Acetic Acid (5-HIAA) were not done because of lack of facilities and finance. Just as our index patient has no money to carry out the above investigation so the diagnosis was relied on Histological report.

The primary treatment of carcinoid tumor is surgical excision of tumor and end-to-end anastomosis done. When such resection is done early enough it provides the best chance for a cure. Just as our index patient [4, 10].

The prognostic factors include the site and size of the primary tumor and the presence of metastases [13, 14]. Extra gastrointestinal carcinoid site such as the ovary, bronchus, testis, pancreas and prostate have worse prognosis. For early diagnosis there should be high index of suspicion for any patient who complains each time he or she eats bananas, pineapple or avocados pear has diarrhea.

Secondly, presence of intermittent diarrhea that is difficulty to manage with known antidiarrheal drugs.

Thirdly, presence of high concentration of urine 5-HIAA.

CONCLUSION

Carcinoid tumors are uncommon and therefore a high index of suspicion is required in making a diagnosis and early surgical excision of the tumor provides the best chance of cure or improved survival.

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