CASE REPORT

Ileal Carcinoid

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ABSTRACT

Background: *Gastrointestinal carcinoids are ill understood, enigmatic malignancies which are extremely difficult to diagnose. In spite of their slow growth rate, they can be aggressive. The aim of this report is to present a case of ileal carcinoid which produced a complete intestinal obstruction.*

Method: *The case records of a 78 year-old tailor who was treated over a period of one year and diagnosed with an ileal carcinoid and a review of literature using medline and manual library search were used.*

Result: *The patient was admitted and treated conservatively on two occasions in 2004 with features of subacute intestinal obstruction. He improved on each occasion and was discharged. He was admitted for the third time in 2005 with complete intestinal obstruction which required an exploratory laparotomy. A small, hard and completely obstructing tumor was found in midileum. No synchronous tumor was detected and there was no ascites. The liver felt and looked normal. Ileal resection and end-to-end anestomosis was done. Post operative recovery was complicated by a bout of severe diarrhea which required intravenous fluid therapy. There was good wound healing and was discharged 12 days after laparotomy. He was well clinically at follow-up review one month after discharge.*

Conclusion: *The case illustrates the difficulty in making a diagnosis of this condition before surgery.*

KEYWORDS: Ileal carcinoid; Difficult pre-operative diagnosis.

Paper accepted for publication 5th June 2006.

INTRODUCTION

Carcinoids are rare and interesting tumors[1]. Gastrointestinal carcinoids constitute ill understood and enigmatic malignancies[2]. They are slow growing compared to adenocarcinomas[2]. Their behaviour, however, can be very aggressive. 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%)[2]. In these sites, they may present with a mass, bleeding, obstruction or perforation. A pre-operative diagnosis is difficult to make except in the presence of the carcinoid syndrome. The diagnosis is usually made incidentally at emergency surgery.[1-4] Early detection and resection offer the best chance of cure and resection should be carried out even in the presence of metastases.[4] Generally the larger the lesion the worse the prognosis. The presence of hepatic metastases further worsens the outlook. A case of ileal carcinoid is presented to emphasize the difficulty in making a pre-operative diagnosis.

CASE REPORT

A 78 year-old tailor presented as an emergency in June 2005 in the emergency unit of the University of Port Harcourt teaching hospital with colicky abdominal pain, abdominal distension, vomiting and constipation for two days.

He had two similar episodes in the previous year which improved on conservative treatment (nasogastric suction and intravenous fluids). After resolution of the first attack sigmoidoscopy and barium enema were done which proved normal. He had bilateral inguinal hernia repair about 10 years previously. The intestinal obstructions on the two previous occasions were attributed to adhesions as he improved satisfactorily on conservative treatment.

Examination on this occasion showed an elderly man with mild dehydration who was not clinically anaemic. The abdomen was distended with visible peristalsis. There were bilateral inguinal scars without evidence of a recurrent hernia. Abdominal tenderness was generalised without rebound tenderness and the bowel sounds were exaggerated. Digital rectal examination revealed an empty rectum. The hemoglobin was 12.3g/dl and the white cell count, urea and electrolytes were within normal limits. Chest radiograph and ECG were normal. An abdominal ultrasound scan was unhelpful diagnostically.

After 36 hours of resuscitation by correction of fluid/electrolytes and nasogastric aspiration a laparotomy was performed under general anesthesia. There was a complete obstruction in the midileum by a small, hard tumor. No nodules were felt in the liver. Mesenteric nodes were not enlarged and no synchronous gastrointestinal tumor was detected. A resection with end-to-end ileo-ileal anastomosis was done. Post operative recovery was complicated by a severe bout of diarrhea which required intravenous fluid

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infusion. He was discharged home 12 days after laparotomy with good wound healing. The histopathological report read, "Carcinoid tumor cells extending through muscle layer to serosa". He was reviewed at 6 weeks and 3 months subsequently. On both occasions his clinical condition had improved without a recurrence of obstructive symptoms or diarrhoea.

DISCUSSION

Carcinoids occur commonly in the gastrointestinal tract. Midgut carcinoids are the most common[5]. In the midgut the commonest site is the ileum[2,6] . Carcinoid has been described in other sites such as the bronchus, ovary, breast, testis, cervix, pancreas, larynx and prostate[1,3,7-9]. The usual distribution in the gastrointestinal tract had been described as, apppendix 65%, ileum 25% and the rectum, colon, stomach and duodenum making up the rest[3]. Current statistics shows that they are most commonly found in the ileum (25%), followed by the rectum (14%) and the appendix (12%)[2]. In the ileum the majority are malignant, 40% are multiple and 30-35% are associated with other malignancies especially adenocarcinoma[3].

Intestinal carcinoids are usually small tumors with a mean diameter of 1cm; the largest tumors have measured about 3cm in diameter[10]. Tumors less than 1cm are rarely malignant, while tumors larger than 2cm are invasive and metastatic[3]. The tumor arises from the argentatffin cells (of KULTCHISKY) in the bases of the crypts of Lieberkuhn. The cells are uniform, round to oval and with a granular cytoplasm[10].

Patients with ileal carcinoids have a potential for other neoplasm's, especially in the colon. It is therefore important to search for synchronous, metachronous and metastatic growths in such cases[4].

Gastrointestinal carcinoids are notoriously difficult to diagnose pre-operatively in the absence of the carcinoid syndrome and therefore remain clinically silent[10,11]. The clinical presentation is usually non-specific, and patients go undiagnosed for years because of the slow growing nature of the tumor. In the presence of the carcinoid syndrome, the diagnosis can be suspected more easily. Extraintestinal sites like bronchus, ovary or testis are more likely to be associated with the carcinoid syndrome because their secretions enter he systemic circulation directly[10].

The patient in this case report exhibited the slow-growth characteristic of the condition. Intestinal obstruction is one of the features encountered. The obstruction which is usually chronic initially later becomes acute[3] as typified by this case report. Occasionally massive gastrointestinal bleeding and perforation may occur.

Apart from chest radiograph, ECG and abdominal ultrasound scan, other investigations such as computed tomography (CT), magnetic resonance imaging (MRI), endoscopic ultrasound and nuclear scintigraphy were not done because of lack of facilities. For the same reason, plasma chromogranin A (CgA), serotonin (5HT) or urinary 5- hydroxyindoleacetic acid (5-HIAA)[2] were not done.

The primary treatment of carcinoid is surgical excision in order to avert local manifestations and reduce secretion[2]. When such resection is done early it provides the best chance for a cure[3,4,10]. Resection should also be carried out even in the presence of distant metastases in order to prevent future intestinal obstruction[4,12]. Boudreaux *et al* further state that the "wait and see" method of management of this slow-growing cancer no longer has merit[12]. Hepatic metastases are amenable to cytoreduction, radiofrequency ablation, embolization alone, or with cytotoxics[2]. Hepatic transplantation may rarely be found beneficial[2]. Modlin *et al* are of the opinion that chemotherapy and radiotherapy substantially reduce the patient's quality of life and the therapy is not of satisfactory effectiveness. Zuetenhorst and Taal feel that because carcinoid tumors are uncommon, such patients should be treated in specialized centres[6].

Prognostic factors are complex and multifaceted[13]. These include site of origin within the gastrointestinal tract, size of the primary tumor and the anatomical extent of the lesion (ie localized, regional or metastatic)[13]. High concentrations of the tumor markers, urinary 5-HIAA and plasma chromogranin A, malignant carcinoid syndrome, carcinoid heart disease are all indicators of poor prognosis. The overall 5 year survival for ileal carcinoid is 60%[2]. Extra gastrointestinal carcinoid sites (eg ovary, bronchus, testis, pancreas and prostate) have much worse prognosis because of the presence of the carcinoid syndrome[10].

As matters stand the way to an early diagnosis is a high index of suspicion. Even then, the presentation may be nonspecific. When it is possible to establish an increase in plasma chromogranin A (CgA) levels the diagnosis may become clearer. However such investigative tool may not be readily available, and where available may be too expensive to carry out in our setting.

In conclusion, carcinoids are uncommon and interesting tumors that can rarely be clinically

diagnosed pre-operatively in the absence of the carcinoid syndrome. A high index of suspicion is required in making a diagnosis while early surgical excision provides the best chance of improved survival or possible cure.

REFERENCES

1. Archampong EQ, Tandoh JFK, Nwako FA, Badoe EA, Foli AK, Akande B, Obianyo NEN Small and large intestines (including rectum and anus). In Badoe EA, Archampong EQ, Jaja MOA (Ed). Principles and Practice of Surgery including Pathology in the Tropics. 2nd edn. Tema Ghana Publishing Corporation 1994: 575 - 656.
2. Modlin IM, Kidd M, Latich I, Zikusoka MN, Shapiro MD. Current status of gastrointestinal carcinoids. Gastroenterology 2005; 128 (6) : 1717 - 51.
3. Cuschieri A. The Small Intestine and Vermiform Appendix. In: Cuschieri A, Giles GR, Moosa AR (Ed) Essential Surgical Practice. 3rd edn. Oxford Butterworth-Heinemann. 1995: 1297 - 1328.
4. Debas HT, Orloff SL. Carcinoid Tumors and the Carcinoid Syndrome. In: Sabistin DC (Ed). Textbook of Surgery 14th edn. Philadelphia WB Saunders 1991: 869 - 873.
5. Akerstrom G, Hellman P, Hessman O, Osmak L. Management of midgut carcinoids. J Surg Oncol 2005; 89(3): 16 - 9.
6. Zuetenhorst JM, Taal BG. Metastatic carcinoid tumors: a clinical review. Oncologist 2005 ; 10(2): 123-31.
7. Lim KH, Huang MJ, Yang S, Hsieh RK, Lin J. Primary carcinoid tumor of prostate presenting with bone marrow metastases. Urology 2005; 65(1): 174.
8. Fujita K, Wada R, Sakurai T, Sashide K, Fujime M. Primary carcinoid tumor of the testis with teratoma metastatic to the para-aortic lymph node. Int J Urol 2005; P(3): 328 - 31.
9. Abdulkareen FB, Banigo AAF, Atoyebi OA, Adesanya AA, Elesha SO. Carcinoid tumor A clinicopathological review. The Nig Postgrad Med J 2000; 7(1): 29 - 32.
10. Adotey JM. Primary malignant tumors of the small intestine. The Brit J Clin Pract 1985; 54 - 58.
11. Gore RM, Berlin JW, Mehta UK, Newmark GM, Yaghmai V. GI carcinoid tumors; appearance of the primary and detecting metastases. Best Pract Res Clin Endocrinol Metab 2005; 19(2): 245-63.
13. Boudreaux JP. Surgical treatment of advanced-stage carcinoid tumors: lessons learned. Ann Surg 2005; 241(6): 839-45.
14. Rorstad O. Prognostic indicators for carcinoid neuroendocrine tumors of the gastrointestinal tract. J Surg Oncol 2005; 89(3): 151 - 60.