CIBTech Journal of Surgery ISSN: 2319-3875 (Online)
An Open Access, Online International Journal Available at http://www.cibtech.org/cjs.htm
2014 Vol. 3 (2) May-August, pp.4-6/Karthikeyan et al.
Case Report

A RARE CASE PRESENTATION OF CARCINOID TUMOUR: A CASE REPORT

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ABSTRACT
Carcinoid tumors are of neuroendocrine origin and derived from primitive stem cells in the gut wall. Carcinoids are the most common neuroendocrine tumors, with an estimated 1.5 clinical cases per 100,000 populations. The majority of patients present with metastasis to the lymph nodes or liver, and 5% to 7% of patients present with the carcinoid syndrome. Appendicular carcinoid tumors are most common. Carcinoid tumors can be associated with concentric and elastic vascular sclerosis that results in obliteration of vascular lumina and ischemia. Approximately 80% of appendicial tumors are incidentally discovered during surgery for other indications, but some cause or coexist with acute appendicitis. Though surgical resection is curative for isolated lesions, metastatic and residual lesion need octreotide therapy and other chemotherapeutic agents. Carcinoid tumour can have any mode of presentation and is good mimicker of other diseases

Keywords: *Neuroendocrine Tumour, Carcinoid, Appendicites*

INTRODUCTION
Carcinoid tumors are of neuroendocrine origin and derived from primitive stem cells in the gut wall. Carcinoids are the most common neuroendocrine tumors, with an estimated 1.5 clinical cases per 100,000 population. Most carcinoid tumors are slow growing and indolent without symptoms. Depending on the size and location, carcinoid tumors can cause various symptoms, including carcinoid syndrome. Carcinoid tumors of the ileum and jejunum, especially those larger than 1 cm, are most prone to produce symptoms. Midgut carcinoids are argentaffin positive and can produce high levels of serotonin 5-hydroxytryptamine (5-HT), kinins, prostaglandins, substance P (SP), and other vasoactive peptides. The majority of patients present with metastasis to the lymph nodes or liver, and 5% to 7% of patients present with the carcinoid syndrome (flushing, diarrhea, wheezing, heart valvular dysfunction, and pellagra). It poses a diagnostic challenge because imaging techniques, such as computerized tomography and small bowel contrast studies, rarely identify the primary tumor preoperatively. OctreoScan is reported to be 80% to 100% sensitive for the detection of carcinoid tumors Measurement of the serotonin metabolite 5-hydroxyindolacetic acid is the most common and reproducible test for detection of carcinoid. Small bowel surgical resection, together with resection of the associated mesentery, is the treatment of choice and is curative in limited early stage. The 5-year survival rate is 65% among patients with localized or regional disease and decreases to 36% in those with distant metastasis

CASES
A 59 year old male patient a chronic smoker with no known co-morbidities presented with history of right sided abdominal pain for 2 days duration, colicky in nature radiating to the umbilicus with associated history of obstipation. Patient had a significant past history of diarrhoea since 10 days. On examination - abdomen distended, generalized tenderness present, guarding and rigidity present in right lower quadrant, bowel sounds were sluggish and left hydrocoele was present. P/R- showed collapsed rectum and prostatomegaly. WBC counts were increased, renal, liver, and coagulation profile were within normal limits. X- ray abdomen was suggestive of intestinal obstruction. USG abdomen showed liver cyst, RIF probe tenderness present. In view of clinical suspicion of perforated appendicites patient was planned for diagnostic laparoscopy and proceed, Intraop findings (Small bowel ischaemia extending from 11/2 feet from DJ flexure to ileocaecal valve, mesenteric thickening) following which resection and end

jejunostomy was done. Post operatively, patient was started on anticoagulants, patient was started on diet which he tolerated well and was conservatively managed. Postoperative CT scan showed (Irregular enhancing lesion in the mesentery located in central abdomen slightly to the right of midline -likely residual carcinoid /neuroendocrine tumor, Few enlarged mesentric and paraaortic lymphnodes, encasement and occlusion of SMA and SMV by the lesion with preserved jejunal branches and right colic branch of the mesenteric artery. Biopsy was suggestive of well differentiated neuroendocrine carcinoma/tumour of low grade malignant potential - grade III (tumour >2cm of size with extension beyond submucosa),extent of tumour involves full thickness muscularis propria, margins free of tumour,1 lymphnode identified and is free of tumour, vascular/perineural/lymphatic invasion absent and no evidence of meastasis. Medical oncology opinion was sought and was advised to start on octreotide.

DISCUSSION

Carcinoid tumors are of neuroendocrine origin and derived from primitive stem cells in the gut wall, but they can be seen in other organs, including the lungs, mediastinum, thymus, liver, pancreas, bronchus, ovaries, prostate, and kidneys. Carcinoid tumors have high potential for metastasis. Midgut tumors are derived from the second portion of the duodenum, the jejunum, the ileum, and the right colon. These account for 60-80% of all carcinoid tumors (especially those of the appendix and distal ileum) in adults and are also seen in children. Appendicular carcinoid tumors are most common. More than 70% of these tumors occur at the tip of the appendix and are often an incidental finding in appendectomy specimens. Midgut carcinoids are argentaffin positive and can produce high levels of serotonin 5-hydroxytryptamine (5-HT), kinins, prostaglandins, substance P (SP), and other vasoactive peptides. These tumors have a rare potential to produce corticotropic hormone (previously Adrenocorticotropic Hormone [ACTH]). Bone metastasis is uncommon.

In the intestinal tract, these tumors develop deep in the mucosa, growing slowly and extending into the underlying submucosa and mucosal surface. This results in the formation of small firm nodules, which bulge into the intestinal lumen. These tumors have a yellow, tan, or gray-brown appearance that can be observed through the intact mucosa. The yellow color is a result of cholesterol and lipid accumulation within the tumor. Tumors can have a polypoid appearance and occasionally become ulcerated. With expansion and infiltration through the submucosa into the muscularis propria and serosa, carcinoid tumors can involve the mesentery. Lesions associated with diffuse wall thickening are relatively uncommon. The size of the tumor can be correlated with outcome of the disease; tumors smaller than 1.5 cm in diameter rarely result in distant metastases or recurrences. Carcinoid tumors can be associated with concentric and elastic vascular sclerosis that results in obliteration of vascular lumina and ischemia. A common finding is elastosis and fibrosis that surround nests of the tumor cells and that result in matting of the involved tissues and lymph nodes.

Approximately 80% of appendicial tumors are incidentally discovered during surgery for other indications, but some cause or coexist with acute appendicitis. The most common clinical presentation for a small intestinal carcinoid is periodic abdominal pain, which can be caused by fibrosis of the mesentery, kinking of the bowel, or intestinal obstruction. A constellation of symptoms called malignant carcinoid syndrome is often associated with this tumor. Diarrhea and malabsorption occur in as many as 84% of patients. Diarrhea may or may not be associated with abdominal pain, flushing, and cramps. It may be profuse and often colicky. Though various vague clinical presentations occur the exact etiology of this disease is unknown.

Urinary 5-HIAA levels are usually increased and aid in the assessment of carcinoid tumors. Measurement of urinary 5-HIAA levels can help in diagnosing carcinoid syndrome but may not help in detecting tumors at an early stage of development when they are potentially curable with resection. CT, MRI, angiography, positron emission tomography (PET), scintigraphy with Metaiodobenzylguanidine (MIBG) and octreotide, radionuclide imaging with somatostatin analogs attached to the radioactive tracer, and technetium-99m bone scanning are available but the sensitivity and specificity is varying and warrants a high clinical suspicion of carcinoid tumour.

Conclusion

Carcinoid tumors are of neuroendocrine origin and derived from primitive stem cells in the gut wall. The incidence is 1.5 clinical cases per 100,000 population. Carcinoid tumours have various modalities of clinical presentation and are not easy to diagnose clinically. Though various laboratory and imaging modalities are present for its detection, one should have a high clinical suspicion of carcinoid as it is usually encountered intraoperatively or histopathologically. Though surgical resection is curative for isolated lesions, metastatic and residual lesion need octreotide therapy and other chemotherapeutic agents. With this case report we conclude that carcinoid tumour can have any mode of presentation and is good mimicker of other diseases.

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