

Carcinoid of the Caecum with Osseous and Soft Tissue Metastases: A Rare Case Report

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ABSTRACT

Background: The carcinoid tumour arises from the enterochromaffin cells of Kulchitsky at the base of the crypts of Lieberkuhn. Gastrointestinal (GI) carcinoids account for 95 % of all the carcinoids and caecal carcinoids account for 5 % of all the carcinoids. They remain silent and asymptomatic for years and many a times, are diagnosed intraoperatively, endoscopically or during autopsy, based on their histopathological findings. Malignant carcinoids metastasise to the liver and the lymph nodes, while bone and soft tissue metastasis have rarely been reported. We present here, a rare case of caecal carcinoid with bone and soft tissue metastasis.

Case Report: A 47 year old female was admitted with pain in the abdomen in our hospital, 3 years back. Colonoscopy showed caecal pathology, for which resection and anastomosis was done. Her post operative histopathology revealed carcinoid

of the caecum which invaded the subserosa with lymph node metastasis. She received adjuvant chemotherapy and presented to the OPD 3 years later, with swellings over the medial end of the right clavicle, the anterior chest wall and the node on the right side of the neck. FNAC from all these sites revealed metastatic carcinoid. She was treated with chemotherapy and palliative radiotherapy and has been asymptomatic for the past two years.

Conclusion: Carcinoids are slow growing, indolent tumours and they may remain asymptomatic for years. Malignant carcinoids rarely metastasise to the bone and soft tissues. The present report adds to the literature, an interesting case of caecal carcinoid with osseous and soft tissue metastasis, besides highlighting the importance of histopathology in its diagnosis, as well as the role of chemotherapy and palliative radiotherapy in controlling the symptoms.

Key Words: Caecal, Carcinoid, Metastases

INTRODUCTION

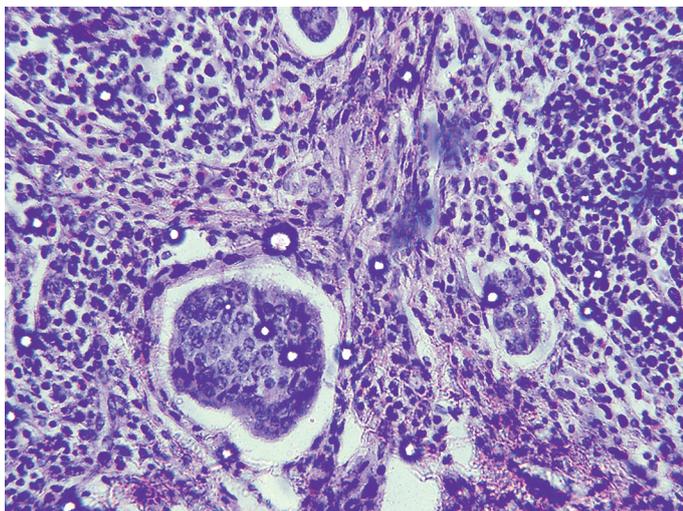
The carcinoid tumour, which is also called argentaffinoma, is a member of the exclusive neoplastic family which is called NET (neuroendocrine tumours) or APUD (amine precursor uptake and decarboxylation) tumours [1]. This tumour arises from the enterochromaffin cells of Kulchitsky which are situated at the base of the crypts of Lieberkuhn. Gastrointestinal carcinoids account for 95% of all the carcinoids and caecal carcinoids account for 5% of all the carcinoids [1]. They remain silent and asymptomatic for years and many a times, are diagnosed intraoperatively, endoscopically or during autopsy, based on their histopathological findings. Malignant carcinoids are known to metastasise to the liver and the lymph nodes, while bone and soft tissue metastasis are very rarely observed; only a single case has been reported so far. We present here, a rare case of carcinoid of the caecum with metastasis to the bone and soft tissues.

CASE REPORT

A 47 year old female with recurrent attacks of abdominal pain, constipation and weight loss for a period of 2 years, was admitted to our hospital in June 2006. Her general examination was essentially normal. On local examination, her abdomen was found to be scaphoid and her umbilicus was found to be centrally located and inverted. A vague, ill defined, non-tender, mobile mass which measured 3 × 3 cm was found, which was palpable in the right iliac fossa. Her digital rectal examination did not reveal any palpable mass through the rectal wall in the pelvis. There was no visible peristalsis and the overlying skin was healthy. Her liver was palpable

2 cm below the costal margin and there was no splenomegaly or ascites. Her routine laboratory and biochemical examinations were normal. The ultrasonogram of her abdomen showed a hypoechoic mass in the abdomen, with thickening of the adjacent gut loops and colonoscopy revealed an annular growth in the caecum which obstructed the ileocaecal junction, for which a laparotomy was done. Intraoperatively, a caecal growth was observed and a hemicolectomy with end to end anastomosis was performed. Grossly, the segment of the terminal ileum and caecum was about 20 cm in length, a nodular, yellowish pink mass, about 3 cm in diameter was present and two smaller nodules of sizes 0.7 and 1 cm in the adjacent colonic mucosa were noticed.

Microscopically, the caecal wall showed a partly distorted lining mucosa and villi with residual mucosal crypts. Well differentiated nests of uniformly round to polygonal cells, having round nuclei with stippled chromatin cleft like retraction spaces around the cell nests were seen and these neoplastic cell nests extended into the muscle coat. The cells appeared in monotonous sheets and were arranged in clusters and nests with retraction artifacts, intense desmoplasia and thin delicate fibrovascular stroma. The nuclei appeared round to oval with scanty cytoplasm and no mitotic activity. The lymph nodes showed nests of neoplastic cells which were suggestive of a caecal carcinoid which invaded the muscles and the pericolic nodes. [Table/Fig-1]. She received 3 cycles of injection 5 FU and was closely monitored. 3 years later, during her follow up, we detected swellings at the medial end of the clavicle, over the anterior chest wall and a node at the right side of the neck [Table/Fig-2]. FNAC (Fine Needle Aspiration Cytology) was



[Table/Fig-1]: Histopathology: H&E stained HP picture of caecal wall showing well differentiated nests of uniform round to polygonal cells having round nuclei with stippled chromatin and infiltration of these neoplastic cell nests into muscle coat. The neoplastic cells are also seen invading the pericolic lymph nodes.



[Table/Fig-2]: Clinical: Picture showing well defined swellings over the medial end of the right clavicle, anterior chest wall just above the sternum and a clinically significant node over the right supraclavicular region.

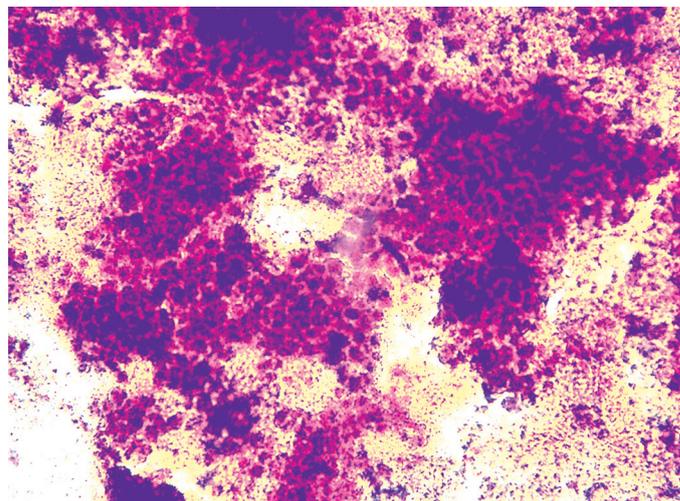
advised from the above three sites; the cytosmear showed small cells in clusters and nests with intense desmoplasia and retraction artifacts. The cells were arranged in monotonous sheets, which was suggestive of a metastatic carcinoid [Table/Fig-3].

The sonography of her abdomen and pelvis revealed hepatomegaly with hyperechoic lesions of variable sizes in both the lobes of the liver, which were suggestive of hepatic metastasis.

Her chest radiograph showed normal lung parenchyma and a soft tissue mass around the medial end of the clavicle, along with destruction of the medial end of the right clavicle, which were suggestive of bone and soft tissue metastasis [Table/Fig-4]. Lab investigations were done for serum serotonin and tryptamine, which were found to be raised. With a diagnosis of metastatic caecal carcinoid, we treated her with three cycles of injection Cisplatin and 5 FU, followed by palliative radiotherapy to the right clavicle at a total dose of 30 Gy in 10 #. 2 years post treatment, she is currently asymptomatic and her disease has been clinically controlled.

DISCUSSION

The carcinoid tumour or argentaffinoma arises from the enterochromaffin cells of Kulchitsky, which are present at the base of



[Table/Fig-3]: FNAC: cytosmear of soft tissue swelling showing small cells in clusters and discretely in Leishman's stain suggestive of metastatic carcinoid.



[Table/Fig-4]: Chest radiograph: (P/A view) showing soft tissue opacity over the medial end of the right clavicle and an osteolytic lesion with destruction of the right clavicle.

the crypts of Lieberkuhn. GI carcinoids are the most common primary tumours of the small bowel and comprise 95% of all the carcinoids [1]. 95 % of all the GI carcinoids are located in the appendix, the rectum and the small intestine. The percentage of the location of the carcinoids is appendix (30-45 %), small bowel (25-35%), rectum (10-15%), caecum (2-3%), and stomach (0.5%) [2]. The carcinoid tumours have been traditionally classified according to their anatomic site of origin as foregut, midgut and hindgut tumours. However, currently, there is a shift away from this system to a histology-based classification system, in which the tumour subtype is determined according to the cellular differentiation [3]. These tumours are very indolent, asymptomatic and slow growing tumours and they may remain clinically silent for years [4]. The hallmark of these tumours is the secretion or production of serotonin, gastrin, kinin- peptide, hydroxyl tryptamine, catecholamine and glucagon, which are responsible for the varied clinical manifestations and they may sometimes induce the carcinoid syndrome which is characterized by flushing, diarrhoea, right sided heart disease and wheezing. These tumours present with a male to female ratio of 2:1. Most of the carcinoids occur in patients who are older than 50 years, except the appendiceal carcinoids which can occur in younger patients in the 2nd to 4th decades of life.

A majority of the carcinoids are asymptomatic or they may present with pain in the abdomen, intestinal obstruction, weight loss, palpable masses, intussusceptions, perforations or gastrointestinal haemorrhage. Asymptomatic carcinoids are diagnosed either during endoscopy, intraoperatively or during an autopsy, where a definitive diagnosis is reached by histopathological examination. The caecal carcinoids present with similar features as that of the ileal carcinoids and are many times clubbed together as ileocaecal carcinoids. Only about 2/1,00,000 of the carcinoid tumours are malignant [5]. Metastasis occur in approximately 30-50% of the patients with carcinoid tumours, and they can occur at any time from before the diagnosis of the primary tumour to 20 years after the initial diagnosis. Carcinoids have a tendency to metastasize to the lymph nodes and the liver, but soft tissue or bone metastasis have rarely been reported [6]. Hepatic metastasis are seen in 10-60% of the cases, depending on the site of the primary tumour [7].

The size of the primary tumour is also cited as the most important determinant of the malignant behaviour; the risk of the metastasis increases if the original tumour size is larger than 10 mm. The most common sites of the metastasis are the mesenteric lymph nodes, the liver, the lung, the peritoneum and the pancreas. Soft tissue or osseous metastasis are exceedingly uncommon. Only 2 cases have been reported, which have described metastasis to the orbit and the soft tissues and only a single case with bony metastasis has been reported. The present case however, describes a caecal carcinoid with bony and soft tissue metastasis in an otherwise asymptomatic patient.

The metastasis are frequently formed from a midgut primary tumour: tumours which are < 1cm in size metastasize in 2% of the cases, tumours which are 1-2cm in size metastasize in 50% of the cases and tumours which are > 2cm in size are clinically silent but may be present with the carcinoid syndrome [2]. Bone and soft tissue metastasis are generally osteoblastic, rarely lytic or mixed lesions [8]. The chest radiograph, in the present case, showed a soft tissue mass along with an osteolytic lesion in the medial end of the right clavicle. The treatment of carcinoids is mainly aimed at the control of the symptoms and can be closely monitored by evaluating serum serotonin and its urinary metabolite. The

somatostatin analogue, octreotide, is frequently used, with benefit. For metastatic carcinoids, chemotherapy with Cisplatin and 5 FU based regimens have been used. Radiotherapy has no role in a curative intent. However, palliative radiotherapy can be used for relief from the pain and swelling.

CONCLUSION

Carcinoids are slow growing, indolent tumours and the patients may remain asymptomatic for years. Gastrointestinal carcinoids may metastasize to the liver and the lymph nodes, but osseous and soft tissue metastasis in an asymptomatic patient is exceedingly rare. Besides the serum levels of serotonin and tryptamine, a definitive histopathological examination can reveal the diagnosis. The present report adds to the literature, an interesting case of caecal carcinoid with osseous and soft tissue metastasis, besides highlighting the importance of histopathology in its diagnosis, as well as the role of chemotherapy and palliative radiotherapy in the control of the symptoms.

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DECLARATION ON COMPETING INTERESTS:

No conflicting Interests.

Date of Submission: **May 09, 2011**

Date of peer review: **May 09, 2011**

Date of acceptance: **Aug 13, 2011**

Online first: **Aug 25, 2011**

Date of Publishing: **Oct 05, 2011**