Metastatic Small Bowel Carcinoid Tumour: Long-Term Survival - A Case Report and Review of Literature

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Citation

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Abstract

Long-term patient survivability with a metastatic bowel carcinoid tumour is seldom heard of.We present a unique case of a patient in his seventh decade who was originally diagnosed with a primary carcinoid of the small bowel (the initial diagnosis being unknown to us) and who waited for more than 35 years before finally being diagnosed with a metastatic recurrence. Until his presentation to our department he remained totally asymptomatic. Interestingly the patient was totally oblivious of the findings of his surgery for a primary cancer many years ago until it was proven by his old medical records.

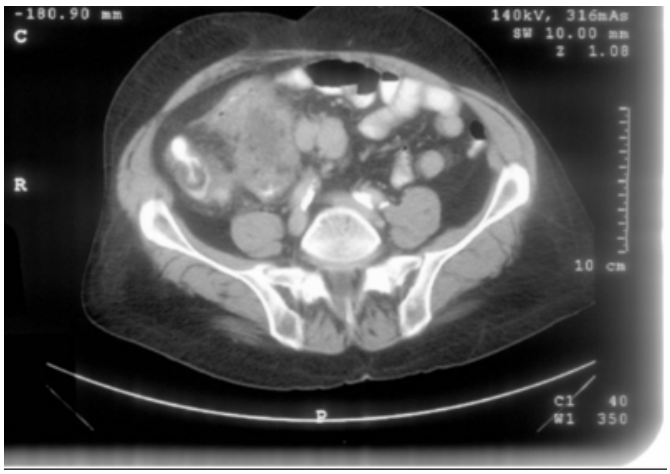
CLINICAL HISTORY

A 70-year-old retired soldier was admitted to the general surgical ward via emergency department in September 2007 at Toowoomba Hospital, Queensland, Australia. His complaints included worsening pain in his right iliac fossa for the previous few days. He denied any other associated symptoms. His past surgical history included a small bowel operation in 1971 for a cancer while still serving in the army but he did not have exact recollection of it.

Clinical examinations revealed a tender right iliac fossa with a soft abdomen. The white cell count, biochemistry and ultrasound findings were normal. CT scan of his abdomen (Figure 1) revealed a tiny collection in the right iliac fossa in relation to the distal ileum (considered most likely secondary to a small contained small-bowel perforation) and non-specific thickening at the ileocaecal region. It also showed numerous tiny mesenteric lymph nodes along with a noticeable 3.2 to 3.6cm sized solitary enlarged lymph node in the small bowel mesentery. The liver was free of any significant pathology.

Figure 1

Figure 1: Note the collection in the RIF along with an adjacent enlarged lymph node near the superior mesenteric artery



Conservative management was initiated. CT scan guided drainage of the collection was attempted with minimal yield. Lab tests of the fluid were unremarkable. Subsequent inpatient CT scan revealed reduction in the collection size and no evidence of bowel lesion was found. He was discharged after few days as his symptoms resolved.

Outpatient clinics follow-up with CT scan after few months still showed the enlarged mesenteric lymph node with smaller satellite nodules but no evidence of bowel or adjacent tumour. Colonoscopy was normal. With high suspicion of a missed lesion, a need for laparoscopy or laparotomy and lymph node biopsy was discussed with the

patient and was undertaken shortly.

He was explored in June 2008. Operative findings revealed widespread lymph nodes in the mesentery with the largest one being 4cm in size abutting the superior mesenteric artery. Evidence of previous small-bowel resection was noted but no tumour found in the small or large bowel. A segment of the small bowel, with its adjacent mesentery involving the enlarged lymph node, was resected (Figures 2, 3) and a primary ileoileal anastomosis fashioned. The superior mesenteric artery was preserved. Appendicectomy was carried out. The resected specimen was opened but no evidence of a luminal tumour was found including at the site of the previous anastomosis. Postoperative convalescence was uneventful and the patient was discharged after few days.

Figure 2

Figure 2: Note the enlarged single lymph node (50×50×50mm), with suture marks in the mesentery along with other tiny satellite nodules at the 10 to 11 o'clock position in the mesentery.

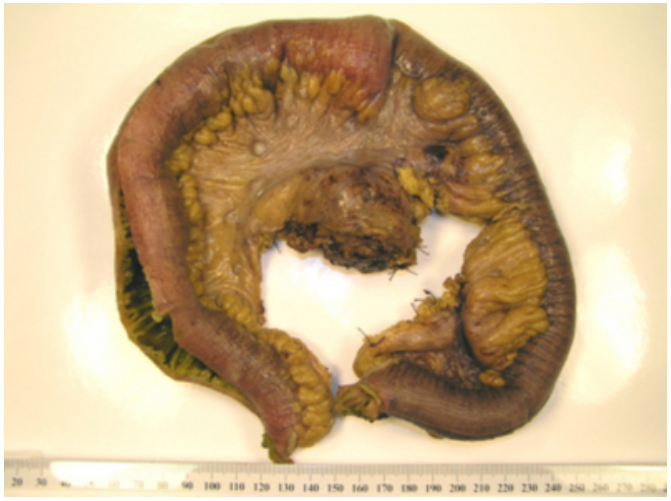
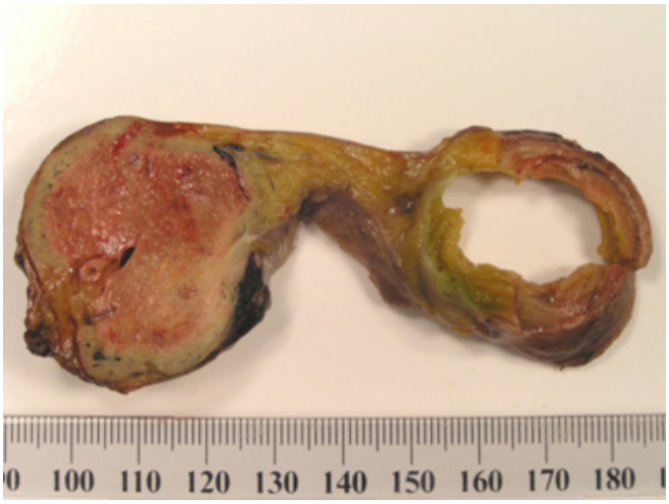


Figure 3

Figure 3: Cross section of the enlarged lymph node and small bowel with the intervening bridge of mesentery. Note the isolation of the lymph node and the small bowel lumen.



Histopathology (Figures 4, 5) revealed extensive multiple carcinoid deposits in all the specimens including the small bowel, lymph nodes (largest deposit of 50×50×50mm), mesentery, mesenteric resection margins and appendix. Of the fifteen mesenteric lymph nodes isolated, all were involved along with their lymphatics. Despite the entire tumour mass, a clear-cut primary site was not detected.

Figure 4

Figure 4: Low Resolution (10×10): H&E stain showing the carcinoid deposits in the lymph node.

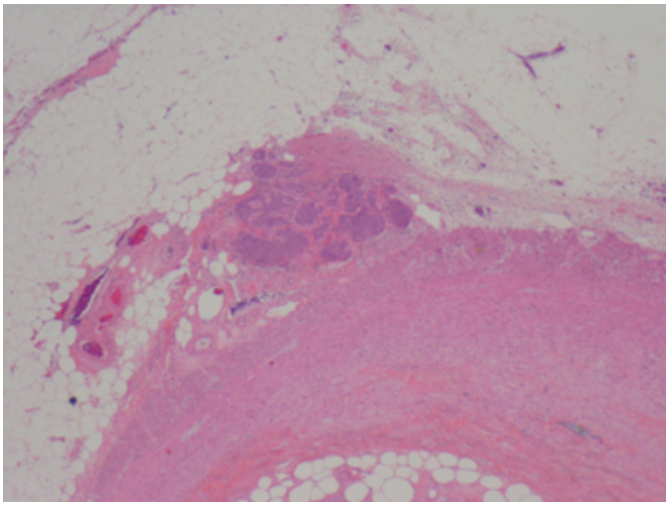
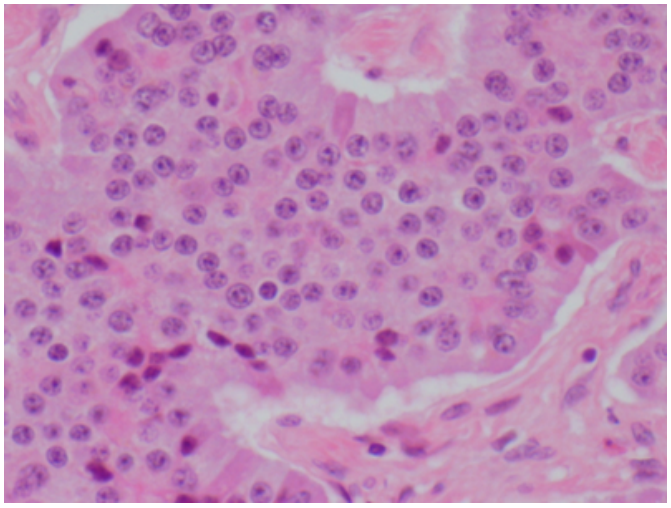


Figure 5

Figure 5: High resolution (40×10) showing typical membrane-bound secretory granules with dense-core granules in the cytoplasm



Doubts surfaced whether his previous bowel cancer in 1971 was the primary source until it was proven by his old army histopathology report as "Small Intestine Argentaffinoma - Carcinoid Tumour." No lymph nodes were reportedly found in the specimen back then. The tumour size was 1.4cm and it was seen to infiltrate extensively through the full thickness of the bowel wall. It, however, did not penetrate into the adjacent mesentery. Unfortunately the old report did not comment on the bowel resection margins.

Retrospectively he did not have any symptoms of carcinoid syndrome like cutaneous flushing, abdominal pain (except during admission), diarrhoea, nausea, vomiting, or cardiorespiratory issues prior to his latest operation.

He started developing diarrhoea and losing weight only postoperatively but has been managed well with octreotide injections commenced by the medical oncologists who are regularly following him up simultaneously. No chemotherapy was initiated due to its ineffectiveness. He was also started on vitamin B12 and folate supplements.

Subsequent 6- to 12-monthly follow-ups so far have been uneventful. The patient remains asymptomatic despite his recent Indium-labelled octreotide scan revealing four areas of metastatic deposits in the abdominal cavity. His latest Platelet Serotonin level is 6240 (<6000 - normal) and serum Chromogranin A 13 (0-21.8 - normal).

Unfortunately he has been recently diagnosed with laryngeal cancer and is under the care of the ENT surgeons.

DISCUSSION

Carcinoid tumours of bowel are not uncommon. They pose a diagnostic dilemma and therefore command a low threshold for clinical suspicion. Long-term survival with metastasis is rarely heard of as the 5-year survivability for carcinoid tumours can range from 30% to 100% depending on the tumour site.

Our patient has provided a unique case by remaining symptom-free for nearly forty years with an unknown metastasis and still continues to do so, now that he is known with one.

Studies state that the majority (55%) of carcinoids are found within the gastrointestinal tract[1]; 45% involve the small bowel, followed by rectum (20%), appendix (17%), colon (11%) and stomach (7%). Of the small bowel carcinoids, the Moertel series[2] reported distal ileum involvement in 60% of the cases, followed by proximal ileum in 32%, jejunum in 5% and duodenum in 3% of carcinoids. In our patient the distal ileum was the site of involvement.

Carcinoids remain asymptomatic for prolonged periods due to their slow rate of growth and indolent course. Around 25 to 30% of patients will already have metastasis at the time of the diagnosis and periods as long as 20 years will have already elapsed before it is diagnosed. In our case, the patient remained asymptomatic with the metastasis for thirty-seven years. To our knowledge none but one case has been reported in the literature (Zacharias et al.[3] - thirty years) which had more than 20 years gap till the diagnosis of metastatic disease.

More than 30% of these tumours metastasise to the local lymph nodes and/or liver prior to their surgical discovery. The tumour size is a good predictor of its metastatic potential. The Moertel series[2] reported that tumours bigger than 2cm showed evidence of metastasis in 95% of cases. There were no metastases in tumours less than 0.5 cm. Only 15% spread in tumours of 0.5 to 0.9cm and 72% in those between 1.0 to1.9cm. For our patient, the size of his primary tumour was 1.4cm.

The median age of discovery is 60 to 70 years and males are involved in 50 to 60% of the patients. Carcinoid syndrome is rare and manifests only in 6-7% of patients. Younger patients are more likely to develop the syndrome and have worse prognosis. The overall 5-year survival rate is around 65% for loco-regional metastasis and 35% for the distant ones.

Multidisciplinary approach is the effective way of treating carcinoid tumours and their metastasis. It includes surgical resection of the primary tumour and debulking, hepatic chemoembolisation and medical/oncological treatment. The preferred initial treatments are surgical resection of the tumour and reduction of the tumour burden as much as possible even though specific treatment decisions are complex and related to the primary carcinoid disease. For patients with metastatic disease no therapy to date has been shown in any trials to prolong the survival of patients and thus, therapy remains palliative.

In conclusion, bowel carcinoids are not uncommon. Their diagnosis can be challenging. Asymptomatic long-term survivability with metastasis is rare but possible and even more, surprising. Multidisciplinary approach is the most effective means of treatment. To date, no therapy so far has prolonged life of patients with metastasis and therefore treatment has always remained palliative. Our patient is one such unique case in a nutshell.

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