# Small Bowel Intussusception Due To Malignant Melanoma Of Unknown Primary With Adrenal Metastasis At Presentation

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**Citation**

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**Abstract**

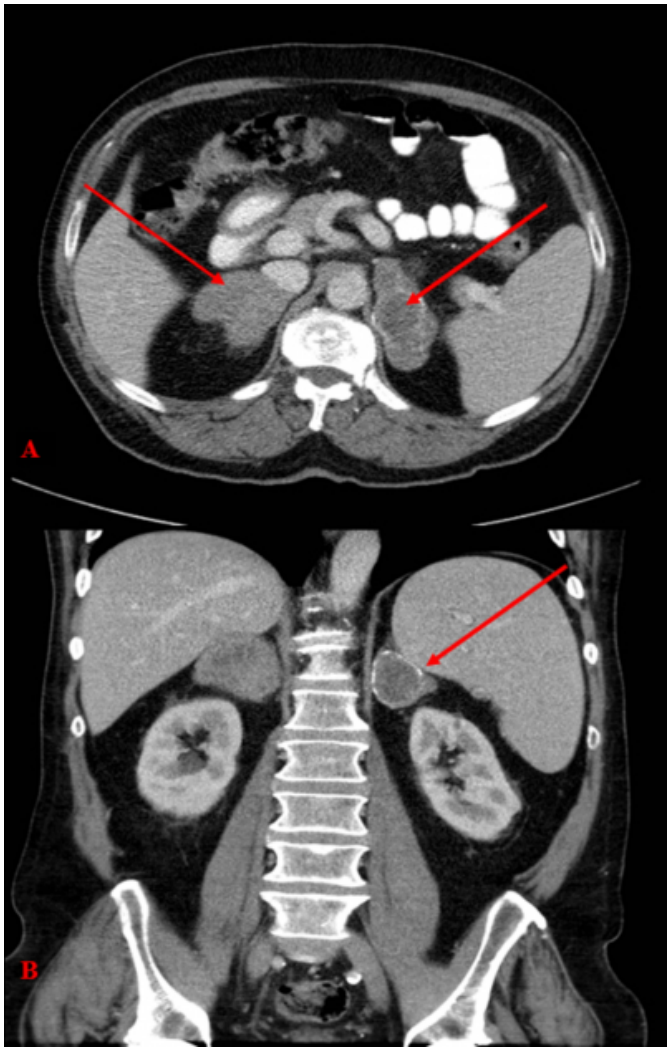
Reported cases of small bowel melanoma are becoming increasingly more common and this is consistent with the understanding that melanoma is the most common extra-abdominal source of small bowel metastases[1]. In fact, reported incidences ranging from 35% - 50% have been documented for metastatic spread of melanoma to the GI tract[2]. Generally, these individuals either present with symptoms of obstruction, particularly intussusception, or bleeding. There have been multiple sites of small bowel spread reported; however, of highest incidence published in the literature, both jejunal and ileal metastasis feature most prominently[1,2,3,4,5]. Of greatest controversy surrounding small bowel melanoma are those cases in which no primary cutaneous lesion has been identified and the validity of classifying these as primary malignant melanoma[6]. There are very few cases reported in the literature in this regard and the debate still wages as to the appropriateness of defining these as primary. We report below a rare case of small bowel melanoma causing ileo-ileal intussusception on presentation with bilateral adrenal metastasis. No primary malignant melanoma was found.

## CASE REPORT

A 70-year-old man presented to the emergency department of Toowoomba Hospital (Queensland, Australia) on the 4 th of August 2009, with a two-week history of constant, colicky right-sided abdominal pain. He described associated nausea and daily episodes of vomiting. He reported constipation for one week, however was still able to pass flatus. He had presented to his General Practitioner earlier that day who requested a computed tomography (CT) scan of his abdomen which revealed moderate dilatation of small bowel loops with ileo-ileal intussusception with no apparent cause. Furthermore, bilateral adrenal masses measuring 6.4 centimetres on the right and 7.8 centimetres on the left were evident on CT (Figure 1). The overall impression was of a subacute small bowel obstruction secondary to the intussusception and he was operated on shortly after presentation.

**Figure 1**

Figure 1: Axial abdominal CT slice showing bilateral adrenal masses as indicated by arrows (A). Note the lobular nature of the adrenal masses and ring of calcification on the left as indicated by arrow on this coronal abdominal CT slice. These features are consistent with malignancy (B).



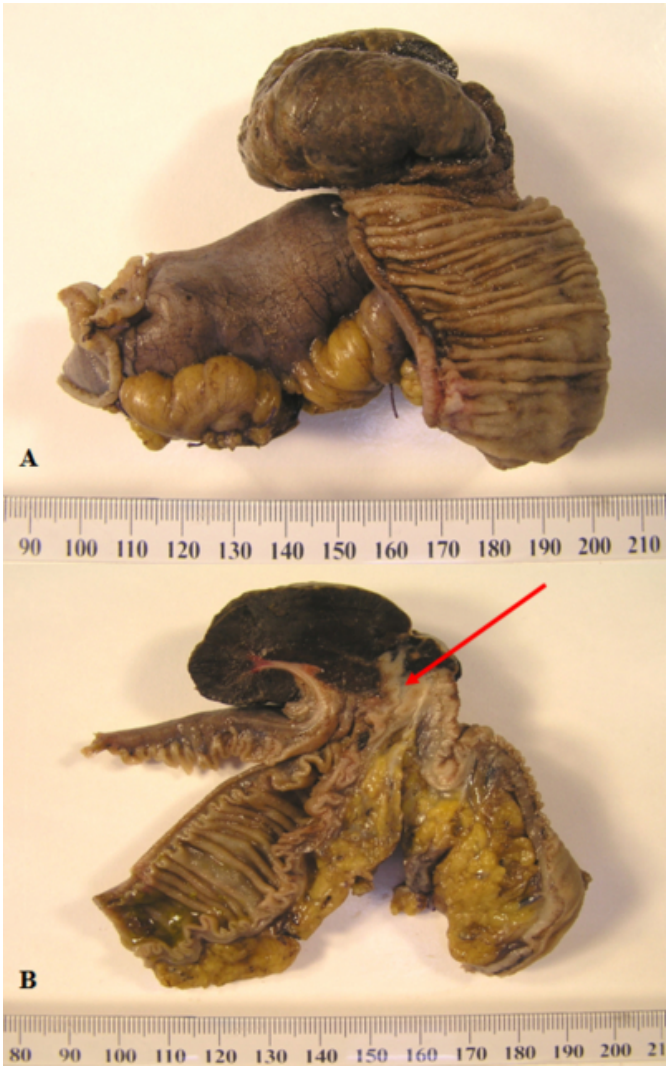

His past medical history included hypertension, dyslipidaemia, previous open appendicectomy and laparoscopic cholecystectomy.

The patient underwent an initial laparoscopic procedure which was converted to a mini-laparotomy after identifying the obstruction point. Small bowel containing the intussusception and tumour was resected and an end-to-end anastomosis was performed with an endo-luminal stapler.

Macroscopically, the tumour mass appeared as a nodular dark brown mass measuring 55x40x32 millimetres and had formed the intussusceptum. The tumour was found to ulcerate through the mucosal surface of the bowel and protrude into the bowel lumen (Figure 2). The tumour involved the submucosa but did not obviously infiltrate the muscularis nor adjacent mesenteric fat. Microscopically, the polypoid tumour showed features consistent with melanoma. The tumour formed a polypoid mass within the mucosa and submucosa and surface ulceration was present. Tumour cells had both an epithelioid and spindle cell appearance with abundant cytoplasmic pigmentation and prominent mitotic activity.

**Figure 2**

Figure 2: Mucosal and serosal surface of a section of ileum showing an intussusception with a nodular dark brown tumour mass (A). Note the ulceration of the tumour mass through the bowel wall into the bowel lumen as indicated by arrow in a transverse section through the section of ileum (B).




Post-operative recovery was complicated by basal atelectasis and consolidation and the patient spent two days to recover

in the high dependency unit. He was subsequently treated on the ward for a further four days and discharge was enabled following resolution of a post-operative ileus. Prior to discharge the diagnosis of melanoma was explained to the patient and a thorough skin examination was conducted without discovery of a primary melanotic lesion. A repeat CT of abdomen and head were also performed prior to discharge. The abdominal CT again revealed the adrenal masses without increase in size and the CT of the head revealed no focal intracranial pathology. He was subsequently referred to the melanoma clinic at the Princess Alexandra Hospital (PAH) (Brisbane, Australia) for further management. On review by the surgeons at PAH he was determined to have stage IV disease and no further surgical intervention was offered and referral to medical oncology was made. He was observed by medical oncology and repeat staging CTs were performed over a period of six months without significant progression in the disease.

On the 11 th of April 2010, he represented to Toowoomba base hospital (Queensland, Australia) with right leg weakness which had progressed gradually over two days. Subsequent CT scan of his head revealed a rounded predominantly hyperdense lesion with perilesional oedema in the left high parietal region. The appearance was in favour of a haemorrhagic metastatic lesion. Following a sixteen day admission under the medical team, and through discussions with the patient and family he was pronounced palliative and was discharged to a hospice on the 27 th of April 2010. He deceased shortly after.

## DISCUSSION

Malignant melanoma is primarily a cutaneous disorder which has the ability to metastasis locally either to neighbouring areas of subcutaneous tissue but also local lymph nodes[7]. Of greatest concern, however, is the impact of distant metastasis which has been reported in the liver, ovaries, uterus and meninges to name a few sites[7]. Small bowel metastasis more importantly is quite common in malignant melanoma and rates of 2-5% for patients with known cutaneous melanoma have been reported for small bowel metastasis[3]. Furthermore, in regard to gastrointestinal metastasis, small bowel metastasis is by far the most common with rates reported of up to 50% and colonic rates of up to 32%[5]. In fact, malignant melanoma is responsible for the most frequent cause of gastrointestinal metastasis.

Although it appears that small bowel metastasis of melanoma is quite common, only recently have the numbers of case reports in the literature increased significantly to mirror this observation. A well established reason for this is that often small bowel metastasis is an asymptomatic condition and only 1-4% of patients are diagnosed before death[8]. There still exists a broad interval between diagnosis of primary malignant melanoma and identification of gastrointestinal metastasis, anywhere between 2 to 180 months[5]. If symptoms exist, these individuals commonly complain of nausea and vomiting, abdominal pain, weight loss or present with intestinal obstruction or bleeding[5,8]. Obstruction typically presents as intussception both jejunal or ileal; however, ileo-ileal intussusception is a rare entity and few case reports currently exist. Despite cases of small bowel malignant melanoma presenting with fulminant obstruction, there have been case reports published whereby patients have presented with intermittent symptoms[8]. Thus the caveat exists in individuals with a previous history of cutaneous excision of melanoma and abdominal symptoms of potential malignancy. The emphasis in these individuals is to further investigate their symptoms through imaging and subsequent exploratory laparotomy or laparoscopy.

Although primarily a cutaneous lesion, up to 10% of malignant melanomas can arise from a noncutaneous source[8]. These include the mucosa of the nasal cavity, turbinates and sinuses. These are collectively referred to as sinonasal melanomas and account for only 1% of all cases of malignant melanoma[8]. The eyes, particularly retina, are another important noncutaneous source reported in the literature[8]. Knowledge of these noncutaneous sources is important when attempting to delineate a primary site when obvious cutaneous lesions have not been identified. Much controversy still exists as to the classification of primary metastatic melanoma of the small intestine. Case reports have been published showing primary small bowel melanoma; however, whether these cases should be defined as such is a mooted subject. The case presented here did not reveal a primary cutaneous source on further investigation. The likelihood is, however, that the melanoma excised was truly metastatic. Although no universal diagnostic guidelines for primary malignant melanoma of the intestine exist, several criteria have been proposed. Blecker et al. recently proposed a set of criteria for primary malignant melanoma of the intestine. The criteria suggest that this diagnosis can be made when there is lack of concurrent or previous removal of a melanoma or atypical melanocytic lesion from the skin, lack of other organ involvement, and in-situ change in overlying of adjacent GI epithelium[9]. Given the presence of

adrenal metastasis on presentation, our case would not fulfil the given criteria. Current sentiments suggest that primary malignant melanoma of the intestine may exist; however, diagnosis as such is rare. Case reports presented as unknown primary still likely represent malignant disease as spontaneous regression of pre/coexisting cutaneous malignant melanoma or a history of depigmentation of a lesion may have occurred[4]. Furthermore, hidden primary lesions such as retinal lesions may also exist unidentified as the primary source.

Adrenal metastasis of melanoma as evident in the case reported is extremely common. Up to 50% of cases of malignant melanoma show evidence of adrenal metastasis[10]. This rate may actually be even higher due to undiagnosed cases which is a reflection of the silent nature in which this metastasis presents. The majority of adrenal metastasis are both biochemically and clinically silent and only identified incidentally on CT, often many years following initial treatment of the primary lesion. Although the adrenal lesions on CT presented here are not classical in the sense that no evidence of the presence of central or irregular areas of necrosis and bleeding was apparent, the lobular nature and bilateral involvement as well as areas of calcification are highly suggestive of malignancy[10].

In conclusion, malignant melanoma of the small intestine presented as ileo-ileal intussusception is a rare entity; however, increasingly more case reports are surfacing in this regard. We presented here a case of ileo-ileal intussusception due to malignant melanoma with unknown primary and bilateral adrenal metastasis on diagnosis. Although proponents exist suggesting primary malignant melanoma of the small intestine is a real entity, there currently exists no universal guidelines for its diagnosis and to suggest a primary intestinal source is thwart with controversy. Furthermore, imaging modalities used to establish the diagnosis are still not perfect with sensitivities in regard to CT not exceeding 70%[4]. With the advent of magnetic resonance enteroclysis (MRE) it is hoped that the sensitivity of primary malignant melanoma of the small bowel detection can be improved.

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