A Rare Case of Meckel's Diverticulum Neuroendocrine Tumour

Meckel's Diverticulum Neuroendocrine Tumour

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A sixty-six year old male presented to the emergency department, brought in by ambulance after experiencing severe, sudden onset, central abdominal pain. The onset of symptoms was noted two hours postprandial, was associated with nausea and multiple episodes of vomiting. Bowel motions in the days preceding were noted to be regular and the patient denied any urinary symptoms. Past medical history was significant for diet-controlled type two diabetes mellitus, atrial fibrillation (for which the patient was anticoagulated), hypertension, gout and obesity. The patient had no prior abdominal surgical history, however colonoscopy had been performed one year prior, which revealed a benign colonic polyp and no other abnormalities.

On examination, the patient was non-tender to palpation of the abdomen. Heart rate and blood pressure were within the normal expected range and the patient was afebrile. The abdominal pain persisted despite analgesia. The patient was admitted from the emergency department for surveillance, pain management and further investigation.

Routine blood examination was largely normal demonstrating inflammatory markers within normal limits and only mildly deranged intrahepatic function. In light of persisting abdominal pain requiring intravenous analgesia a computed tomography (CT) scan of the abdomen was requested, specifically to exclude ischaemic colitis in the setting of known atrial fibrillation. The CT images revealed a small focal soft tissue area of calcification in the mesentery of the right iliac fossa with some associated distortion of an adjacent small bowel loop (Figure 1). To further characterise the suspicious lesion a Fluorodeoxyglucose (FDG F 18) Positron emission tomography (PET) study and Gallium-68 Dotatate study were requested. Findings consisted of prominent somatostatin receptor (SSTR) expression with Krenning score 3, consistent with a neuroendocrine tumour metastasis. Tumour markers prostate-specific antigen (PSA), carcinoembryonic antigen (CEA), carbohydrate antigen 125 (CA 125),

carbohydrate antigen (CA 19-9), alpha-fetoprotein (AFP) and Chromograffin-A were not raised.

However, 24-hour urine 5-hydroxyindoleacetic acid (5-HIAA) was determined to be elevated.

With the suspicion of a neuroendocrine tumour, the patient was scheduled for surgery. Commencing laparoscopically the small bowel was examined from the ileocecal valve to the duodenojejunal flexure. A Meckel's diverticulum was identified and was found to be densely adherent to the underlying mesentery (Figure 2). At this point the decision was made to proceed with conversion to open midline laparotomy to facilitate small bowel and mesenteric resection via LigaSure sealing device and gastrointestinal anastomosis (GIA) stapler. Anastomosis was achieved with a double layer, continuous 3-0 PDS, side-to-side, isoperistaltic join. The operation was uncomplicated, and the patient made an expected postoperative recovery with complete resolution of symptoms.

Histopathological analysis of the specimen confirmed the suspicion of a well differentiated neuroendocrine tumour involving a Meckel's diverticulum with adjacent lymph node metastases (Figure 3). The patient was subsequently referred for medical oncology follow up.

Meckel's diverticulum is the most common congenital anomaly of the gastrointestinal tract and is caused by incomplete obliteration of the vitelline duct during intrauterine life.¹ It is localised approximately 45 to 60 cm proximal to the ileocaecal valve on the antimesenteric side, on the projection of the terminal branch of the superior mesenteric artery. This position represents the rotational axis of the foetal gut.² Neuroendocrine tumours are the most common malignancy arising in Meckel's diverticula.^{3,4} Even so, the finding of a neuroendocrine tumour inside a Meckel's diverticulum is uncommon, representing less than 1% of alimentary tract endocrine tumours.⁵ There are fewer than 200 cases described in the literature. Some patients display symptoms of a typical carcinoid syndrome, including flushing and diarrhoea, however many patients, as in this case do not, and the disease is often an incidental radiological or operative finding. Histopathological analysis of

surgically resected specimens, particularly specimens from the gastrointestinal tract, may also incidentally reveal carcinoid disease in the setting of other suspected pathology such as appendicitis. Although rare, when present, neuroendocrine tumours involving Meckel's diverticula are often associated with nodal metastases and liver metastases, even when the tumours are small. Carcinoid syndrome resulting from intestinal carcinoids usually manifests only when patients have metastases to the liver. Solitary intestinal carcinoids secrete mediators into the portal circulation where they are metabolised by the liver, hepatic metastases may impair mediator metabolism, or themselves secrete mediators directly into the hepatic vein and thus into the systemic circulation.⁶ Therefore, the optimal management of these neuroendocrine tumours is resection of the primary tumour with regional lymphadenectomy, and debulking of liver metastases where appropriate and feasible.⁷

This case highlights a rare but serious condition of neuroendocrine tumour involving a Meckel's diverticulum warranting surgical intervention with oncological resection. As diagnosis is made often incidentally in the absence of pathognomonic signs or symptoms, it is paramount that any suspicious lesion is further investigated and that in all laparoscopic and open abdominal surgical procedures where feasible the small bowel is checked for the presence of a Meckel's diverticulum, which may in fact be harbouring disease.

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Figure Legends:

Figure 1. Computed tomography (CT) abdominal imaging. a) Axial; and b) Coronal sections, revealed a small focal soft tissue area of calcification in the mesentery of the right iliac fossa (indicated by arrows) with some associated distortion of an adjacent small bowel loop.

Figure 2. Macroscopic, operative resection specimen. Small bowel opened longitudinally and surrounding peri colic fat displayed. A Meckel's diverticulum was evident with associated mass (sectioned), tethered to the underlying peri colic fat.

Figure 3. Histopathological analysis. Haematoxylin and eosin stain confirmed the suspicion of a well differentiated neuroendocrine tumour involving a Meckel's diverticulum with adjacent lymph node metastases: a) 10x magnification of tumour; b) 200x magnification of tumour; c) 100x magnification of nodal deposit; d) 10x magnification of adjacent large node.

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