Case report

Unusual cause of dysphagia

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SUMMARY

In this case, we describe a unique case of large renal hydronephrosis in a 79-year-old Indian male patient who had initially presented with 3 months of progressive dysphagia and loss of weight. His dysphagia was initially thought to be related to the atypical diagnosis of achalasia and was being considered for an elective laparoscopic Heller myotomy. On performing CT of the abdomen, a large renal mass was discovered. However, predicament remained regarding the exact aetiology of this renal mass. This case highlights a tremendously intriguing case of dysphagia with an underlying aetiology that has not been reported elsewhere previously.

BACKGROUND

We chose to present this patient, as this is an unusual cause of dysphagia that mimicked achalasia. The patient presented to our hospital with symptoms of dysphagia, vomiting, and loss of weight. Prior to his admission, he had a recent admission at another metropolitan hospital for aspiration pneumonia. During that admission, he was investigated and worked up for dysphagia. The investigations performed included barium swallow test, which showed 'birds beak' appearance, inconclusive oesophageal manometry and gastroscopy. Initial differential diagnoses were achalasia, gastro-oesophageal tumours, gastro-oesophageal reflux disease (GORD) and hiatal hernia.

This patient presented as a diagnostic dilemma with an atypical clinical history that only became clear after further imaging. As there are no other case reports with similar presentations, this would serve as a good eye opener for other clinicians—in terms of when to consider CT abdomen and pelvis in patients presenting with vague gastrointestinal symptoms.

CASE PRESENTATION

A 79-year-old Indian male patient presented to a tertiary centre in Melbourne with 3 months of progressive dysphagia, which significantly worsened over the 3 weeks prior to admission. He was admitted under the upper gastrointestinal surgical unit for further evaluation and consideration of an elective Heller myotomy.

The patient presented with the primary complaint of dysphagia; unable to tolerate any solids or liquids without regurgitating it back up. The vomit contained undigested food with no apparent features of haematemesis such as coffee ground appearance. The patient did however complain of mild epigastric pain while vomiting. This dysphagia was accompanied by substantial weight loss of 5 kg over the preceding 2 weeks. He denied any associated fever, cough or other coryzal symptoms. He also denied symptoms of bowel obstruction such as new constipation or significant abdominal distension.

The patient had a recent presentation to another large metropolitan hospital due to aspiration pneumonia in October 2017. During that admission, he was investigated for dysphagia. A barium swallow test revealed the following features: 'bird beak' appearance, marked distal oesophageal dilation with abnormal peristalsis and tertiary peristaltic waves; however, subsequent oesophageal manometry (lower oesophageal sphincter basal pressure: 8.9 mm Hg, residual pressure: 9.3 mm Hg, upper oesophageal sphincter basal pressure: 48.9 mm Hg, residual pressure: 7.5 mm Hg) was not typical of achalasia. He consequently underwent a gastroscopy, which showed oesophageal dilation and gastritis. The patient was given the diagnosis of achalasia and managed conservatively at that stage and was referred to the upper gastrointestinal surgical unit at Monash Medical Centre for surgical management of achalasia.

His medical history includes a previous Heller myotomy at the age of 17 and neurofibromatosis. The indication for the Heller myotomy remains unclear as it was completed overseas with no records available in regard to this. He has no known drug allergies and does not take any regular medications.

Clinical examination was unremarkable. His abdomen was soft and slightly distended and no organomegaly was appreciated. Abdominal pain was not elicited on deep palpation.

INVESTIGATIONS

As mentioned above, the patient had barium swallow study and oesophageal manometry done during his recent admission in another hospital. The barium swallow study had shown 'bird beak' appearance, which is a key radiological feature of achalasia. He then underwent oesophageal manometry and the following test pressures were recorded:

- Lower oesophageal sphincter basal pressure: 8.9 mm Hg, residual pressure: 9.3 mm Hg.
- Upper oesophageal sphincter basal pressure: 48.9 mm Hg, residual pressure: 7.5 mm Hg.

Such pressures are not typical of achalasia. The patient was further investigated with gastroscopy, which showed oesophageal dilation and gastritis.

During his admission at our hospital, initial blood investigations on presentation were significant for elevated white cell count (16.2×10^9/L) and neutrophilia (13.5×10^9/L). Other blood investigations

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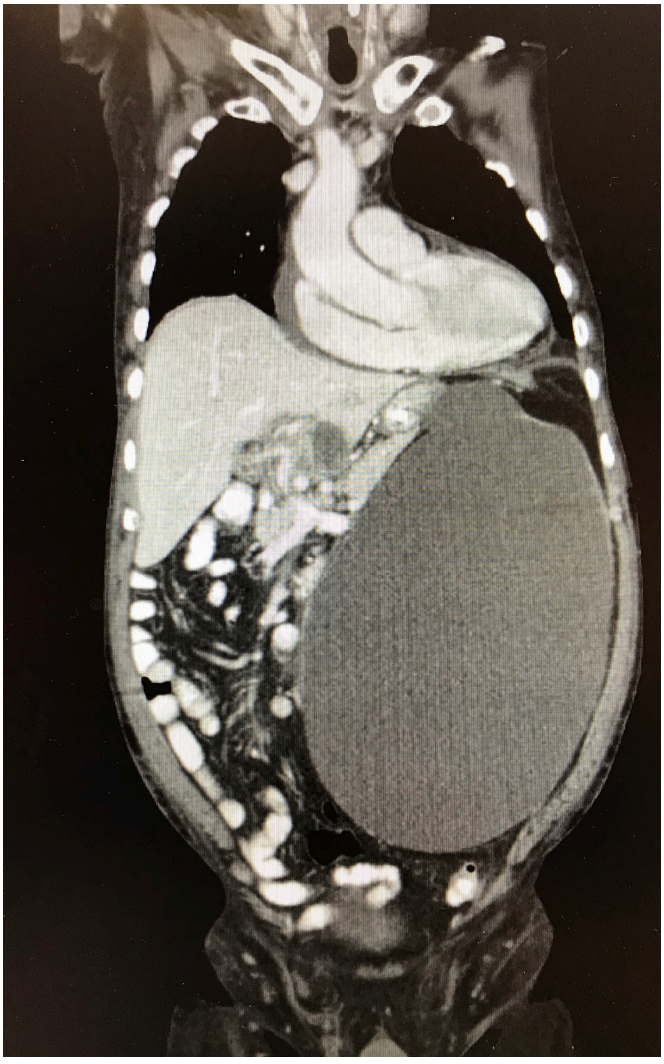

Figure 1 CT abdomen and pelvis showing a very large left hydronephrosis.

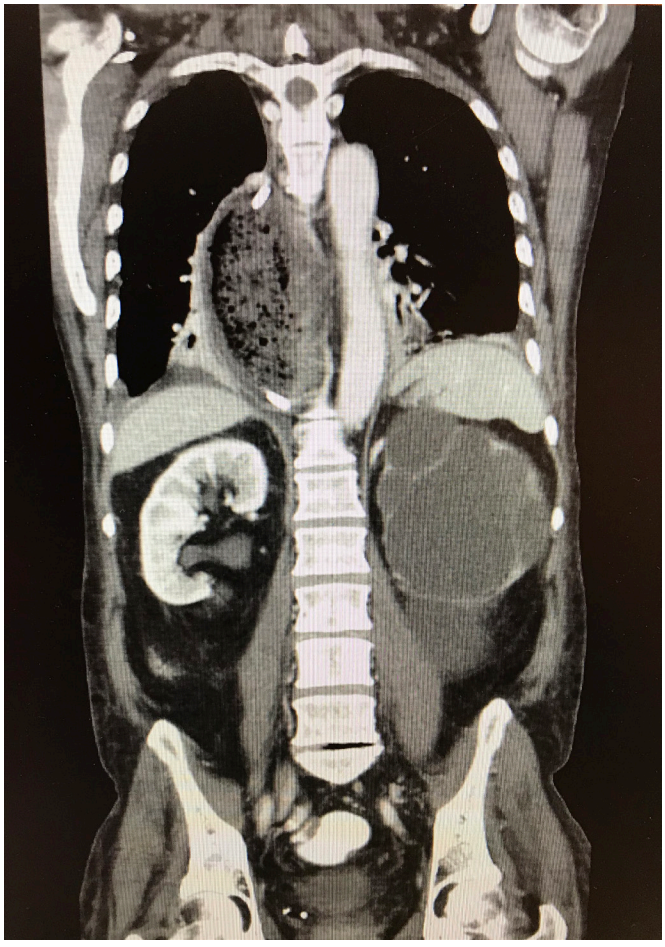

Figure 2 CT abdomen and pelvis showing a dilated oesophagus secondary to compression of the stomach by the large left hydronephrosis.

were unremarkable on admission. Additionally, an erect chest X-ray performed initially was also normal with no evidence of hiatal hernia.

A CT abdomen and pelvis (CTAP) revealed a large left intrarenal multiloculated lesion of simple fluid density measuring 110×201×212 mm which appeared to be arising from the renal pelvis, was compressing and displacing the stomach and other abdominal organs (figure 1). The left distal ureter was not dilated and had no evidence of calcification or mass. There was marked dilation of the oesophagus with air fluid level (figure 2). It was suggested that the large cystic renal mass was most likely secondary to a chronic pelviureteric junction obstruction. Small right-sided renal cysts were also observed. There were no masses external to the ureter seen to suggest external obstruction.

An ultrasound-guided interventional cystic drainage was subsequently organised to drain the left renal cystic mass as recommended by the consulting urology team. A total of 4 L of fluid was drained. The initial 200 mL was sent for microscopy, culture and sensitivity, acid-fast bacilli culture, as well as fluid cytology. Microscopy of the fluid showed debris, inflammatory cells, macrophages and small clumps of epithelial cells with no features of malignancy. There was no microorganism growth after a period of incubation. There was also no evidence of acid-fast bacilli detected on the stain and the culture after 6 weeks. A nephrostomy was left in situ to allow any further drainage.

To evaluate the nature of the mass, a left nephrostogram was requested which showed contrast pooling in the previous mass, with no flow entering the left ureter or renal pelvis. Furthermore, a renal nuclear scan with 99m-Tc MAG-3 concluded that there was likely long standing left pelviureteric junction obstruction given the absence of left kidney perfusion and excretion of radioisotope into the left kidney collecting system. The right kidney contributed to 100% of the total renal function. Due to the inconclusive investigations, the patient also underwent a CT intravenous pyelogram, which showed a collapsed left-sided non-functional multicystic dysplastic kidney. A repeated oesophageal manometry was not considered postdrainage of the cystic mass as the patient's symptoms had improved soon after. On a later admission under the general medical team, a gastroscopy and endoscopic ultrasound was performed to biopsy a retroperitoneal lymph node which had suspicious features on a new CTAP that was organised to investigate for fevers of unknown origin and weight loss. The gastroscopy had showed dilated distal oesophagus without features of achalasia. The endoscope was able to progress into the stomach without difficulties.

DIFFERENTIAL DIAGNOSIS

When the patient first presented, the differential diagnoses considered were achalasia, gastro-oesophageal tumours, GORD and hiatus hernia. Achalasia was considered due to radiological evidence of 'bird beak' appearance. Furthermore, with a significant history of Heller myotomy (for assumed achalasia), late recurrence of achalasia or postoperative complications such as GORD and hiatus hernia can develop. Lastly, given his age and symptom (loss of weight), malignancy was vital to exclude.

Based on the findings from the CTAP, it was ascertained that the symptoms of dysphagia were attributed to external compression of the stomach by the large renal mass. However, there was uncertainty as to whether the mass was cystic in nature or hydronephrosis secondary to chronic obstruction of the left pelviureteric junction. The fluid cytology from drainage of the left renal mass was inconclusive. Furthermore, the fluid was investigated for infective causes and the microscopy, culture and sensitivity did not yield any microorganisms including tuberculosis. Consequently, the mass was further investigated with a left nephrostogram and renal nuclear scan, which concluded that the lesion was likely hydronephrosis secondary to a chronic obstruction in the pelviureteric junction.

TREATMENT

Once it was established that there was large left-sided hydronephrosis, the urology team was consulted on its management. An ultrasound-guided interventional cystic drainage was performed, which drained up to 4 L of fluid. A nephrostomy drain tube was left in situ and was kept on free drainage. This was removed 1 week later.

OUTCOME AND FOLLOW-UP

About 4 L of fluid was drained during the ultrasound-guided interventional cystic drainage procedure. After the procedure, the patient reported that his symptoms pertaining to dysphagia and associated nausea had improved within 1–2 days. A weight chart indicated that he had lost about 4 kg of weight after the procedure (58.7 kg on admission to 54.5 kg). His feeds were upgraded over the course of 2 days after the nephrostomy tube was inserted. However, due to malnourishment and low oral intake, the nasogastric tube was left in place to supplement him with additional nourishment. On patient review, he reported that the low oral intake was due to loss of appetite and early satiety. He denies any nausea, dysphagia or regurgitation of food. The dietician was consulted throughout the process of reintroducing and titrating his feeds to optimise his nutrition as best as possible. The nephrostomy drain tube was subsequently removed 1 week after insertion. With the resolution of his symptoms and a clear cause of pathology, a repeated oesophageal manometry was not considered. He was discharged to the care of the geriatric team 2 weeks later for ongoing rehabilitation due to significant deconditioning.

On following up the patient's past medical records retrospectively, the patient was subsequently transferred back to acute care under the medical team after 1 week at the rehabilitation facility. He had become ill with fevers likely from an infection of unclear origin. A CTAP done during this period of deterioration revealed that a previous complex cystic liver lesion had increased in size, retroperitoneal lymphadenopathy and reaccumulation of fluid in the left kidney. On this readmission, he was found to have abdominal tuberculosis, which was positive on lymph node biopsy obtained via endoscopic ultrasound. Gastroscopy was performed at the same time of the endoscopic ultrasound biopsy and showed dilated distal oesophagus without features of achalasia. He was commenced on long-term antibiotic treatment for tuberculosis (rifampicin and isoniazid) and was looked after by the infectious disease team. The discovery of abdominal tuberculosis was unrelated to when he first presented with dysphagia given that this was investigated on his readmission to the general medical team and that earlier acid-fast bacilli cultures were negative.

The patient saw the urology team in the outpatient setting for management of the left hydronephrotic non-functioning kidney. An elective laparoscopic left nephrectomy was concluded and scheduled. Unfortunately, on the day of his elective procedure, the patient had a cardiac event after anaesthetic induction, and the procedure was aborted. He was discharged for further cardiology workup and will be relisted for surgery after preoperative optimisation.

DISCUSSION

Achalasia is a primary oesophageal motility disorder resulting from insufficient relaxation of the lower oesophageal sphincter (LOS) and is a cause of dysphagia.[1] It is typically defined by specific manometric criteria in the setting of dysphagia.[1] Classical features of achalasia are dysphagia to both solids and liquids associated with regurgitation of undigested food and saliva, radiological evidence of 'bird beak' appearance and poor emptying of barium and oesophageal dilation seen on gastroscopy.[1 2] The patient presented initially with symptoms and radiological evidence that are the same as the classical features of achalasia. In another study, other symptoms of achalasia include weight loss and aspiration, both of which were symptoms that the patient also had.[3] This diagnosis was further supported by his unclear surgical history, which included a previous Heller myotomy many years ago. At this point, the only factor ruling out the diagnosis was the manometry, in which the pressures were not representative of achalasia. In achalasia, typical manometric features of impaired LOS relaxation are increased LOS pressures, absent distal oesophageal peristalsis and impaired LOS relaxation with deglutition.[2 4] However, manometric results of the patient reveal normal LOS pressure (18.8 mm Hg) and absent peristalsis. The gold standard for diagnosing achalasia is by high-resolution manometry.[3] The CTAP was imperative in reaching the final diagnosis of the renal hydronephrosis and without it the patient would have potentially been advised to undergo a Heller myotomy instead.

There have been no similar published case reports on large renal hydronephrosis causing dysphagia. However, one case report published in the Sultan Qaboos University Medical Journal had similar pathophysiology in that a large left renal cyst, measuring 122×67×88 mm, was compressing on the stomach.[5] In this case report, the presenting symptoms of the patient was early satiety and ongoing left flank pain. This pain was attributed to the stretching of the renal capsule. Patients with giant hydronephrosis commonly report symptoms of abdominal pain (maybe intermittent) and fullness (or distension).[6] These symptoms were also consistent in other case reports.[6–9] Other symptoms include constipation, loss of appetite, urinary tract infection and haematuria.[7] In rare cases, symptoms of bowel obstruction, respiratory distress, hypertension, pedal oedema, obstructive jaundice and contralateral uteropelvic junction (UPJ) obstruction were identified.[6] Interestingly, Yang reported two cases of giant hydronephrosis that presented clinically as acute cholecystitis and right inguinal hernia.[9] When comparing the volumes of the renal masses between the two patients, our patient had a significantly

larger renal mass (4.6 L) than the other (0.72 L). Given the significantly larger size, the compression and displacement of the stomach is understandably more likely to cause dysphagia. On the CTAP, the patient's large hydronephrosis fits the radiological definition of the term 'giant hydronephrosis', presence of hydronephrosis occupying a hemiabdomen and extending at least fiver or six vertebral bodies in length.[7] The most common cause of giant hydronephrosis is congenital UPJ obstruction.[9] The case reports of giant hydronephrosis (adult patients only) that we found on our search, demonstrated two main causes: urolithiasis and large renal cysts.[6–9] After various radiological investigations, it was concluded that our patient had hydronephrosis secondary to chronic left UPJ obstruction. In this case, we could assume that the patient potentially may have congenital stenosis of the left ureter at the UPJ leading to chronic hydronephrosis. This would explain why the patient never had symptoms for a long time until recently when the volume of hydronephrosis was large enough to externally compress on the upper gastrointestinal structures resulting in dysphagia.

The diagnosis of dysphagia secondary to extrinsic compression by the large left giant hydronephrosis was clear based on atypical oesophageal manometry readings, CTAP findings of giant hydronephrosis with compression and of the stomach and distal oesophagus and swift resolution of symptoms (1 to 2 days) postdecompression of the renal pathology. A repeated oesophageal manometry or other investigations of dysphagia were not considered as the patient's symptoms had improved after cystic drainage of the left hydronephrosis. The diagnosis of achalasia was also subsequently ruled out by gastroscopy (which was done in conjunction with the endoscopic ultrasound for lymph node biopsy) that did not show features of it. Gastroscopy should be considered as part of investigations for dysphagia. It was not initially considered for our patient due the CTAP findings. Gastroscopy is indeed useful to rule out other causes of dysphagia such as oesophagitis, oesophageal stricture, Schatzki's ring or even oesophageal tumours.[10] Furthermore, it can be used to biopsy pathologies for a conclusive aetiology and has low perforation complication rates (2.6%).[11]

Learning points

- Diagnosis of achalasia can only be made with manometric results indicating aperistalsis and incomplete lower oesophageal sphincter relaxation with supportive symptoms and radiological evidence.
- Patients with unclear or undefined cause of dysphagia should have abdominal CT scan to complete their investigations.
- Renal hydronephrosis may have an atypical presentation; including dysphagia in extreme circumstances such as this.

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