

Case and Review

# Intussuscepting Ampullary Adenoma: An Unusual Cause of Gastric Outlet Obstruction Leading to Cavitating Lung Lesions

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## Keywords

Intussusception · Ampullary adenoma · Gastric outlet obstruction

## Abstract

Ampullary adenomas are a rare clinical entity, occurring at a rate of 0.04–0.12% in the general population. They are premalignant lesions which have the capability to progress to malignancy, and they should be excised if they are causing immediate symptoms and/or are likely to degenerate to carcinoma. Intestinal intussusception in adults is rare and, unlike in children, is often due to a structural pathology. Intussuscepting duodenal/ampullary adenomas have been reported in the literature on 13 previous occasions, however never before with this presentation. We report the case of a woman who presented with a 1-year history of recurrent chest infections. She was treated with numerous antibiotics, whilst intermittent symptoms of recurrent vomiting and weight loss were initially attributed to her lung infections. A chest CT demonstrated multiple cavitating lung lesions, whilst an obstructing polypoid mass was noted at D2 on dedicated abdominal imaging. Due to ongoing nutritional problems, she had a semi-urgent pancreaticoduodenectomy. Intraoperative findings demonstrated a large mass at D2 with a duodeno-duodenal intussusception. Histological analysis

reported a duodenal, ampullary, low-grade tubular adenoma, 75 × 28 × 30 mm in size, with intussusception and complete resection margins. The patient recovered well and was discharged on postoperative day 10, with no complications to date. Ampullary adenomas may present with obstruction of the main gastrointestinal tract and/or biliary/pancreatic ducts. Common presentations include gastric outlet obstruction, gastrointestinal bleeding or acute pancreatitis. This unique presentation should remind clinicians of the need to investigate recurrent chest infections for a possible gastrointestinal cause.

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## Introduction

Ampullary adenomas arise from the ampulla of Vater, the confluence of the pancreatic and common bile ducts, located at the major duodenal papilla in the second part of the duodenum. Based upon autopsy studies, ampullary adenomas are thought to occur at a frequency of 1:1,000–2,000. However, the vast majority of these patients will remain asymptomatic [1–4]. As with colonic adenocarcinoma, ampullary adenomas also follow the adenoma-carcinoma sequence and may thus evolve from localised premalignant lesions into widespread carcinoma [5]. Such lesions may be removed endoscopically or via surgical excision, with either a local resection or an extended procedure, for example, a pancreaticoduodenectomy.

Intussusception occurs when a section of the intestine invaginates into another segment. This may occur as an idiopathic process, as is more common in the paediatric population, or it may be due to a structural pathology, such as colonic malignancy.

As other gastrointestinal adenomas, ampullary adenomas assume tubular, tubulovillous or villous forms. They can occur either sporadically or in patients with a genetic predisposition, such as familial adenomatous polyposis or Gardner's syndrome, and such lesions may therefore be identified via endoscopic surveillance programmes [5]. Indeed, previous studies have suggested a further genetic link to other disorders, such as adult polycystic kidney disease [6], whilst no specific environmental factors such as diet have been identified in their development [7]. Clinically, they are usually asymptomatic and found incidentally at endoscopy; however, some patients may present with symptoms caused by obstruction of the main gastrointestinal tract and/or the distal biliary/pancreatic ducts. The most common presenting symptoms are gastric outlet obstruction or gastrointestinal bleeding [8, 9]. However, rarer presenting complaints may include acute pancreatitis, biliary obstruction or, as in this case, intussusception.

Indications for the excision of such lesions are essentially two-fold: to attempt resolution of immediate symptoms and to prevent malignant progression. The management of such lesions may be endoscopic surveillance, endoscopic excision or surgical resection. Surgical resection has the advantage of lower recurrence rates as opposed to endoscopic resection; however, with the obvious caveat of a much higher risk of morbidity and mortality [7].

In this article, we present a case where a female patient developed recurrent chest infections and gastric outlet obstruction secondary to an intussuscepting ampullary adenoma. Due to the size and location of the mass, it was surgically removed by pancreaticoduodenectomy. To the best of our knowledge, this is the first such presentation worldwide, and we hope that it may be of educational value to both physicians and surgeons alike.

## Case Presentation

A 49-year-old Nigerian female presented to her local hospital with a 1-year history of recurrent chest infections. She had non-insulin dependent diabetes but no other significant past medical history. She took metformin twice daily and had no known drug allergies. She had no past surgical history and was a lifelong non-smoker. She had no relevant past family history of any genetic disorders, in addition to no recent foreign travel or contact exposure to tuberculosis. When she initially presented, plain chest radiographs were performed and demonstrated multiple lung lesions suggestive of an infective/inflammatory process. She grew pseudomonas on sputum culture, but despite treatment for atypical pneumonia with multiple courses of antibiotics, she failed to adequately improve. Chest CT was consequently performed and demonstrated multiple cavitating lung lesions (fig. 1). During this time, she had intermittent vomiting with an associated 15-kg weight loss over a 12-month period. These symptoms were initially attributed to her recurrent chest infections and were not further investigated.

The lung lesions were sampled and were found to be negative for tuberculosis. A working diagnosis of aspergillus infection was established, and the patient remained under the care of the respiratory physicians. At the time of chest CT, however, an incidental finding was noted in the second to the fourth part of the duodenum. Concurrently, she developed subacute bowel obstruction with persistent vomiting and was unable to tolerate oral intake. A dedicated CT scan of the abdomen and pelvis was therefore performed, demonstrating an obstructing polypoid mass at D2 (fig. 2).

This lesion was confirmed on upper gastrointestinal endoscopy, and a biopsy demonstrated a tubulovillous adenoma with high-grade dysplasia. Nasojejunal tube insertion was attempted at her local hospital; however, this failed to bypass the obstructing lesion. Her case was discussed at the regional hepatopancreaticobiliary multidisciplinary team meeting at the Royal London Hospital. As her biopsy demonstrated high-grade dysplasia, in addition to her symptomatic presentation, it was felt that a resection was required. Based upon her cross-sectional and endoscopic imaging, it was deemed that this would not be amenable to an endoscopic excision, and she was likely to require surgical resection and should be transferred to the HPB unit at The Royal London Hospital. Interestingly, despite this large lesion, she never became jaundiced and maintained a normal bilirubin level.

On arrival at the hospital, she was admitted to the surgical ward, and although she had persistent vomiting and was cachectic, she was haemodynamically stable. She was nutritionally deplete with a moderate hypokalaemia at 2.7 mmol/l (normal range 3.5–5.0) and an albumin level of 30 g/l (normal range 35–50) due to the failed nasojejunal tube insertion. Consequently, a PICC line was inserted by interventional radiology, and in an attempt to optimise her for major abdominal surgery, total parenteral nutrition was administered. However, due to these ongoing nutritional problems and a failure to adequately improve, she underwent a semi-urgent conventional pancreaticoduodenectomy. The procedure was successful without any intraoperative complications.

Intraoperatively, a large mass at D2 with a duodeno-duodenal intussusception and distal duodenal ischaemia was identified (fig. 3). Histological analysis reported a polypoid tumour at the Ampulla of Vater, 75 × 28 × 30 mm in size, with a reduced intussusception and an ischaemic distal duodenum (fig. 3). These findings were in keeping with a duodenal ampullary low-grade tubular adenoma, with intussusception and complete resection margins. There was no evidence of any high-grade dysplasia, in contrast to her previous diagnostic biopsy, or invasive malignancy. The patient was admitted to the high-dependency unit post-

operatively and was stepped down to the surgical ward on postoperative day 2. She progressed well with early mobilisation, and her nutritional status greatly improved during this time, with due care not to induce a refeeding syndrome. She was medically stable and deemed fit for discharge on postoperative day 10. She resumed work 4 weeks postoperatively, with no evidence of recurrent chest infections. She did not develop any exocrine pancreatic insufficiency or any derangement from her baseline glycaemic control. To date, the patient has not suffered any long-term complications, and remains clinically well on follow-up.

## Discussion

Intussusception in adults is rare and is responsible for approximately 1–5% of all adult intestinal obstructions. These can be classified by location into enteroenteric, colocolonic, ileocolic and ileocaecal, respectively [10]. The presenting symptoms are usually non-specific and chronic, with pain and symptoms of intermittent intestinal obstruction, such as vomiting, bloating and/or diarrhoea or constipation being the major complaints. The classic paediatric triad of cramping abdominal pain, bloody diarrhoea and a tender abdominal mass is rare in adults. As opposed to the paediatric population, where the aetiology is normally idiopathic, in adults, the cause is usually a structural pathology, with two-thirds of colonic cases being a malignant neoplasm, as opposed to one-third of those occurring in the small intestine [11–13].

Abdominal CT is now regarded as the modality of choice for investigating such lesions. Due to the preponderance for a structural aetiology, the management of such cases is almost always via laparotomy, with no role for the use of air insufflation as utilised in the paediatric population [12]. Intraoperative reduction of the intussuscepted bowel is possible; however, there are associated risks such as tumour seeding and anastomotic complications due to the pathological bowel tissue [14]. It may, however, be carried out if the lesion has been confirmed as benign in the preoperative period, and can be performed by milking the lesion in a distal to proximal direction, followed by a limited resection [15, 16].

A literature search revealed 13 cases of duodenal/ampullary adenomas causing duodeno-duodenal intussusception, and to the best of our knowledge, no case of an ampullary adenoma causing symptomatic intussusception with recurrent chest infections was previously reported. Incidental duodenal polyps are reported in the literature at a rate of 0.3–4.6% and have a variety of pathological origins, including adenomas, submucosal tumours and hamartomas amongst others [17]. Duodenal tubular adenomas are extremely rare, representing approximately 0.5–1% of all duodenal lesions [18].

Ampullary adenomas are thought to be precursors to adenocarcinomas, as such tissue is present in 90% of the resected ampullary adenocarcinoma specimens, whilst based on autopsy studies, they may occur at a rate of 0.04–0.12% in the general population [1–3].

Due to their location, ampullary adenomas may clinically manifest via obstruction of the main gastrointestinal tract or the biliary and/or pancreatic ducts [19]. However, they are usually asymptomatic and found incidentally at endoscopy, whilst some patients may present with symptoms, most commonly gastric outlet obstruction or gastrointestinal bleeding [8, 9]. Rarer presenting complaints may include acute pancreatitis, biliary obstruction or, as in this case, intussusception.

Depending on the clinical urgency of definitive treatment, such lesions may be further investigated using a wide range of modalities, including ultrasonography, CT, endoscopic ultrasonography (EUS), magnetic resonance cholangiopancreatography and endoscopic

retrograde cholangiopancreatography. EUS has proven to be a vital technique in the local assessment of periampullary tumours and has a higher sensitivity to both US and CT when detecting such lesions and assessing muscular (suggesting invasion, and therefore malignancy) and vascular involvement (to assess resectability) [20]. However, ERCP is also extremely useful and has a similar sensitivity to EUS in detecting ampullary tumours [21].

There are three broad management strategies for ampullary adenomas: surveillance, endoscopic resection and surgery. Endoscopic resection may be possible in adenomas with low-grade dysplasia, which are <1 cm in size and demonstrate benign characteristics at endoscopy. High-grade dysplasia is a source of controversy. However, Chini and Draganov [19] suggest that they believe endoscopic resection to be an appropriate course of action even in the presence of high-grade dysplasia if the two above criteria are met. The presence of an ampullary adenocarcinoma warrants surgical resection if the candidate is fit. However, there have been sporadic case reports where such malignancies have been removed endoscopically [22–26]. However, debate surrounding their management exists, as some groups speculate that the progression from adenoma to carcinoma is a low percentage, whilst others argue that the quality of forceps biopsy via endoscopy may miss occult foci of adenocarcinoma within the adenoma [7].

Asymptomatic patients may be treated with endoscopic management with endoscopic surveillance as follow up; however, there is no current consensus as to the ideal surveillance post endoscopic resection [19]. If endoscopic removal fails or is unsuitable, surgical intervention may be required. Surgical options include polypectomy, pancreas-sparing duodenectomy or pancreaticoduodenectomy [27].

## Conclusion

Clinicians must ensure that patients who are previously well and present recurrently with chest infections are fully investigated and that they consider the possibility of a lesion causing gastric outlet obstruction. Clinicians should consider dedicated abdominal imaging, and if a mass is identified, undergo endoscopy to further characterise the lesion and develop an on-going management plan with appropriate specialist input. The index of suspicion should of course be higher if the patient goes on to further develop signs of an obstructing lesion, such as persistent vomiting.

## Statement of Ethics

The authors have no ethical conflicts to disclose.

## Disclosure Statement

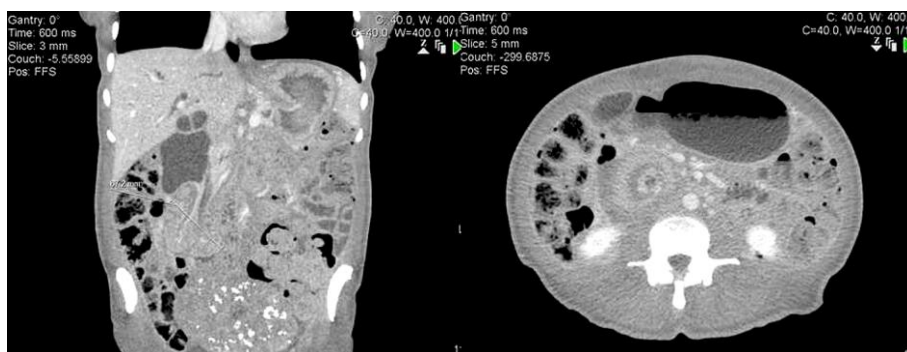
The authors have no conflicts of interest to declare.

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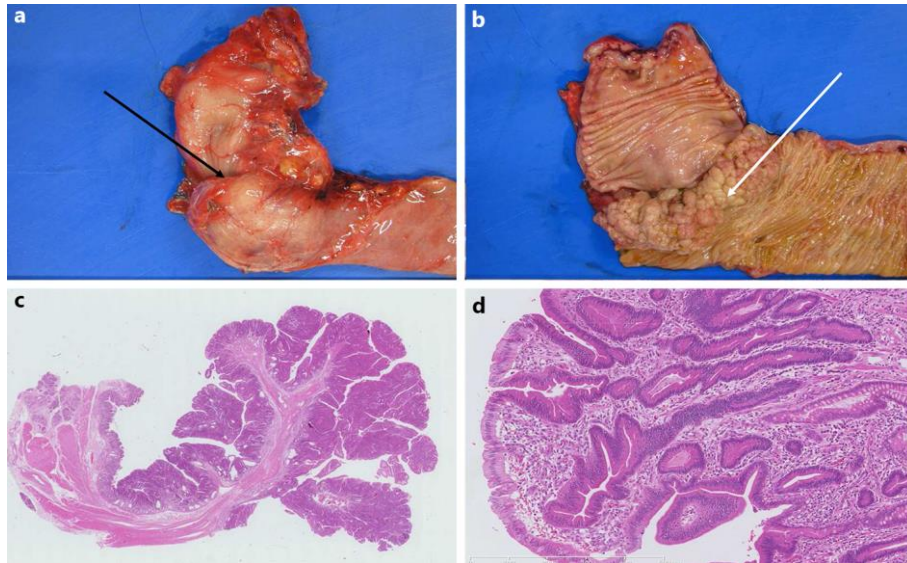
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**Fig. 1.** Coronal slice of the chest CT scan, demonstrating multiple cavitating lung lesions.



**Fig. 2.** Coronal and axial images from the CT scan of the abdomen, demonstrating an obstructing, 6.6-cm polypoid mass at D2, with a 'target sign' shown in the axial image.



**Fig. 3.** **a** External surface demonstrating intussuscepting duodenal ampullary tubular adenoma. The black arrow indicates the site of invagination. **b** Luminal surface. The white arrow indicates the circumferential duodenal ampullary tubular adenoma. **c** HE. ×5 magnification. Tubular adenoma. **d** HE. ×100 magnification. Tubular adenoma with low-grade dysplasia.