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Case Report

An Unusual Case of Neck Swelling in a Patient with Intestinal Failure on Home Parenteral Nutrition

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Abstract

Home Parenteral Nutrition (HPN) is the standard management for patients with Intestinal failure. We present a 35-year-old Caucasian woman, on HPN for stricturing Crohn's disease, who was admitted to our hospital for further investigation and management of a large neck swelling. Differential diagnoses included possible line related complications which are frequently observed in patients on HPN. Our patient also had a recent history of acute pancreatitis and had required cyst-gastrostomy, raising mediastinal pseudocyst as a possible diagnosis for this admission. The diagnosis was confirmed with radiological imaging and biochemical investigations. We present her diagnostic workup, multi-disciplinary team discussion, and subsequent treatment in our hospital. As there are no guidelines available for the investigation and treatment of the unusual presentation of this condition, we highlight the importance of taking a detailed history and examination in order to make the diagnosis of this rare scenario of pancreatic pseudocyst presenting as a neck swelling.

Keywords: Mediastinal pseudocyst; Complication; Intestinal Failure; Home Parenteral Nutrition

Introduction

Home parenteral nutrition (HPN) is the standard management of patients with type 3 intestinal failure, commonly known as short bowel syndrome [1]. It has associated complications which could be line related, including thrombosis, phlebitis and occlusion;[2] or metabolic, such as liver disease, cholelithiasis and metabolic bone disease [3]. We present an interesting case of neck swelling in a patient receiving HPN for multifocal stricturing Crohn's disease. Diagnosis was made after reviewing the clinical history, examination findings, radiology and biochemical analysis of cyst fluid. Through this case report, we want to emphasize the importance of detailed history taking in making a diagnosis in a complex patient presenting with a very unusual symptom of neck swelling.

Case Presentation

A 35-year-old Caucasian woman on Home Parenteral Nutrition (HPN) for obstructing multifocal stricturing Crohn's disease presented with a left neck swelling of insidious onset with gradual progression over three weeks. She was admitted to her local district hospital. The initial working diagnosis was a neck abscess due to suspected parenteral nutrition fluid leak. Thick purulent fluid was aspirated and she was started on broad spectrum antibiotics. Blood and fluid cultures were sterile. She was transferred to our hospital, University Hospital of Wales, which is a tertiary care and referral centre for intestinal failure in South Wales. She had a complex medical history and had been on HPN for three years for medically refractory Crohn's disease. In addition, she was diagnosed with invasive pulmonary aspergillosis involving the left upper lobe and had been on long term prophylaxis. She had also been admitted the previous year with acute pancreatitis secondary to gallstones and developed peri-pancreatic pseudocysts which required endoscopic ultrasound guided cyst-gastrostomy.

On examination, she had a tender boggy swelling in the left side of her neck and generalised oedema which was more pre-

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*Table 1. Results from biochemical fluid analysis. CEA, carcinoembryonic antigen; CA 19-9, cancer antigen 19-9. *Please note that these assays are not CE marked or accredited for use in fluid analysis and therefore results may not be as accurate for the analytes as they would be in serum.*

Days from admission	Sample type	Analyte	Result	Units	Reference interval	Notes
0	Fluid	Amylase	469.0	U/L	N/A	
		Cholesterol	0.7	mmol/L	N/A	
		Triglycerides	1.0	mmol/L	N/A	
	Serum	Amylase	21	U/L	0 - 125 U/L	
4	Serum	Amylase	40.0	U/L	0 - 125 U/L	
5	Fluid	Amylase	1173.0	U/L	N/A	*Analysed locally*
		CEA*	89.0	ng/L	N/A	
		CA 19-9*	2018.0	U/ml	N/A	
	Serum	Amylase	38	U/L	0 - 125 U/L	
5	Fluid	Amylase	1238.0	U/L	N/A	*Analysed at referral lab*
		Pancreatic isoform*	1154.0	U/L	N/A	
		Salivary isoform*	84.0	U/L	N/A	
		Pancreatic:total ratio*	0.93			
13	Fluid	Amylase	25	U/L	N/A	
		CEA*	29	ng/L	N/A	
		CA 19-9*	40	U/ml	N/A	

dominantly left-sided in the face and upper limbs. She had a right internal jugular tunnelled central venous catheter with no sign of occlusion or infection. Ear, nose and throat examination was normal. Abdominal examination revealed a soft abdomen with no tenderness or palpable masses. Our list of differential diagnoses included: salivary gland abscess, ruptured aspergilloma of the left lung, chyle leak, vascular complication related to central venous catheter and a mediastinal pseudocyst.

Computed Tomography (CT) of the neck and thorax confirmed a massive multiloculated cystic collection which extended from the left retro-pharyngeal region to the diaphragm through the posterior mediastinum. This compressed the left brachiocephalic vein and displaced the carina and oesophagus. She was managed with surgical placement of three percutaneous catheters draining the collection.

Initial fluid analysis showed presence of white blood cells with negative cultures, including for tuberculosis. Serum beta-glucan and aspergillus polymerase chain reaction tests were subsequently found to be negative. Fluid amylase was 469 U/L and the serum amylase was 21 U/L. The location of the cyst in relation to the patient's central line in combination with the biochemical characteristics of the fluid minimised the possibility of a parenteral nutrition fluid leak. Cytology and tumour markers gave no evidence to suggest malignancy. Fluid triglyceride concentration of 1.0 mmol/L alongside with the clinical history made chyle leak a less likely diagnosis (Table 1).

After initial intra-operative drainage of a milky fluid, drain output became more serious and browner in colour. Biochemical fluid analysis of serial samples revealed a peak elevated amylase of 1173 U/L. Serum amylase concentrations remained normal. Surgical and radiological opinion was that there was no evidence of damage to the oropharynx to explain a salivary fluid leak, nor evidence of communication of the cyst with the diaphragm. For clarity the fluid was sent to a specialist laboratory for amylase isoenzymes. This confirmed the raised total amylase concentration and found predominant pancreatic isoforms with a pancreatic: total amylase ratio 0.93. This is consistent with pancreatic fluid and thus a pancreatic pseudocyst.

The patient was managed with a course of broad-spectrum antibiotics under the guidance of the microbiology team. Drainage catheters were removed after two weeks and the patient had no recurrence of collection in the four-week period whilst she was an inpatient. Fluid biochemistry obtained on day 13 of admission show declining amylase and tumour marker concentrations which also suggest closure of cyst communication with the pancreas. A three month follow up CT scan shows no recurrence. A detailed magnetic resonance imaging (MRI) scan of the pancreas suggests that there may have been a causative dilated branch of the pancreatic duct that was deemed not amenable to a stenting procedure by the hepatopancreatobiliary (HPB) surgical team. Increased TPN provision has improved her nutritional status and the gastroenterology team are optimising her treatment to reduce her symptoms and improve bowel function. Written patient consent and waiver from the research governance were obtained.

Discussion

Peripancreatic pseudocyst is a well-known complication of acute or chronic pancreatitis [4]. Mediastinal pseudocysts are very rare and only described in case reports. They are formed due to posterior disruption of the pancreatic duct and pancreatic fluid tracking to posterior mediastinum through a route of least resistance via the aortic or oesophageal hiatus [5]. They are most commonly detected incidentally on imaging performed for a peripancreatic collection. They are very difficult to diagnose if there is no concomitant pancreatic pathology. In such a scenario, the physician's awareness and knowledge of this rare complication is required particularly in the context of a past history of acute pancreatitis such as in this patient. Diagnosis was only made after awareness of the fact that the patient had sustained acute pancreatitis in the past year complicated by peripancreatic pseudocysts.

Despite finding a single case of mediastinal pseudocyst that presented as a neck swelling in the literature [6], to our knowledge this is the first case report where there has been no clear radiological evidence of pseudocyst communication across the

diaphragm. Biochemical analysis of pancreatic pseudocysts should reveal a markedly raised fluid amylase although confusion may arise when its location is atypical and various cut-offs exist [7] . Fluid lipase can be tested although in this case would not help differentiate between a possible salivary or pancreatic source. The utilisation of amylase isoenzyme analysis at a specialist laboratory gave definitive evidence to reach the final diagnosis. Biochemical assays of fluid can give clues to the nature of an unexplained collection and must be interpreted alongside other clinical and diagnostic findings. Close discussion with biochemistry colleagues can be informative, as in our case.

Management involves appropriate imaging and adequate drainage. CT scan is the first investigation of choice to delineate anatomy of the mediastinal pseudocyst [8]. MRI with MRCP (Magnetic Resonance Cholangiopancreatography) is the best modality to identify ductal disruption and ductal communication with the pseudocyst. Endoscopic Ultrasound (EUS) has been a recent advance with role in both diagnostic and therapeutics for pancreatic pseudocyst [9].

Treatment options include conservative management, percutaneous drainage with or without EUS guidance and surgery [10]. Conservative management is only suitable for small and asymptomatic mediastinal pseudocysts. Symptomatic pseudocysts (e.g dysphagia, respiratory distress or neck swelling) and infected collections are more appropriately managed with definitive drainage. Endoscopic transpapillary treatment of pseudocysts involves insertion of a pancreatic duct stent or a nasopancreatic drain. Patients with partial duct disruption tend to respond better to endoscopic therapy whereas patients with complete duct disruption often require surgery for a continuous active pancreatic fistula [11].

Conclusion

Mediastinal pancreatic pseudocyst is rare but a well-recognised complication of acute and chronic pancreatitis. Diagnosis is often difficult, exemplified in this case where there was no clear source for the fluid collection nor communication with the pancreas. Reaching a diagnosis relies on the clinical skills of the physician and involvement of the multi-disciplinary team, including diagnostic specialties. This is particularly important in patients on HPN where the list of differential diagnoses is long and includes central venous catheter related complications.

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Conflict of Interest

None declared.

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