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Case Report

A case of recurrent pancreatitis with subclavain artery stenosis: a clinical dilemma (unrelated co-existence or IgG4 related disease?) and a clinical lesson

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ABSTRACT

We report a case of a 56-year-old woman who presented with clinical features suggestive of acute pancreatitis. She had a similar episode 5 years ago. A blood pressure (BP) recording of 80/50 mmHg in the presence of acute pancreatitis led to the diagnosis of circulatory shock and vasopressors were about to be commenced. However, her overall appearance was stable and measurement of BP in the lower limbs was normal. An angiogram revealed left subclavian artery stenosis which explained the low BP reading on left arm. The patient responded to conservative management of pancreatitis, however refused further evaluation of subclavian stenosis. This case is being reported to highlight a clinical dilemma and a clinical lesson. Dilemma arises if the pancreatitis and subclavian artery stenosis is just a coincidental occurrence or it is a case of IgG4 related disease consisting of autoimmune pancreatitis (AIP) type 1 with subclavian artery stenosis a part of extra-pancreatic manifestation of the IgG4 related disease spectrum. This distinction is important in management of pancreatitis and other organ involvement. It is important to examine a patient fully and especially the vascular system - wherein all pulses are to be felt and blood pressure recorded in both sides and both limbs - whenever there is a conflict between the condition/appearance of the patient and the signs that we elicit. In this case recognising that BP was normal in the lower limbs prevented unnecessary use of vasopressors.

Keywords: Acute pancreatitis, IgG4 related disease, Subclavian artery stenosis, Doppler examination

INTRODUCTION

IgG4 related disease is a clinical entity discovered as recently as in 2003 – which includes acute pancreatitis along with other extra pancreatic manifestations.[1] It started with description of autoimmune pancreatitis (AIP) as early as 1995.[2] Then, increased circulating levels of IgG4 in the serum of patients with pancreatitis was described , which correlated with the degree of disease activity.[3] Subsequently, other organ involvements came to light and IgG4 related disease was described as a separate clinic-pathological entity.[4] IgG4 related disease being an auto immune disease responds dramatically to steroid therapy.

Autoimmune pancreatitis (AIP) exists as two types – type 1 AIP associated with IgG4 related disease and type 2 AIP related to idiopathic duct centric chronic pancreatitis.[5] Extra pancreatic manifestations occur only in type 1 AIP. Vascular involvement by IgG4 disease has been described to involve thoracic and abdominal aorta, hepatic artery, inferior mesenteric artery and subclavian artery.[1,6]

We describe a patient with acute recurrent pancreatitis and subclavian artery stenosis. Hence, possibility of IgG4 related disease causing pancreatitis and subclavian artery stenosis was considered, and options discussed. While it might be an association by chance, this report is being

made to highlight the presence of IgG4 related disease and to consider this diagnosis whenever extra pancreatic lesions are discovered along with pancreatitis.

CASE REPORT

A 56-year-old diabetic woman presented to our institute with abdominal pain of 10-days duration. The pain was in the epigastrium radiating to back. It was associated with vomiting and had no relation to food intake. She denied urinary symptoms on admission however was treated for right renal colic 10 days prior to this admission. She had an episode of pancreatitis a few years ago for which she had endoscopic retrograde cholangiopancreatography (ERCP) with biliary stenting which was subsequently removed. She underwent appendicectomy 5 years ago and a laparotomy for reasons unknown to her twelve years ago. Despite her significant past medical history, she did not have any medical records of her previous treatments.

Abdominal examination revealed generalised tenderness maximal in the epigastrium. There was no rigidity and bowel sounds were heard normally. She was apyrexial and her saturations were 96% on room air. The pulse rate was 90/min and blood pressure (BP) was 80/50 mm Hg in the left upper arm.

Routine blood investigations revealed normal white cell count of 8900 cells/cu/mm, elevated amylase (600 IU) and lipase (1800 IU) levels. Computed tomographic (CT) scan (Figure 1 and 2) revealed acute interstitial pancreatitis. CT severity index for pancreatitis of 6. There were also 3 mm calculi in right kidney, 1.5×1 cm in right renal pelvis. The abdominal aorta was normal.

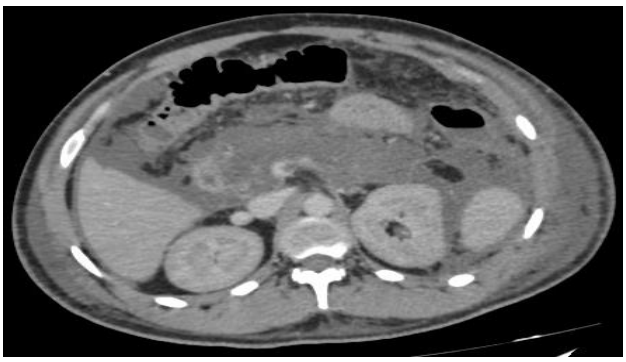

Figure 1: CT scan demonstrating acute pancreatitis with no enhancement.

Presence of hypotension in the setting of acute pancreatitis led to the calculation of a worse prognostic score. Fluid resuscitation was started with a wide bore cannula in the right antecubital vein. Fluid resuscitation did not improve the blood pressure recorded in the left arm. Presence of the wide bore cannula in the right elbow prevented measurement of the blood pressure in the right arm. Arrangements were being made to commence vasopressors to improve the mean arterial pressure. However, the appearance of the patient did not match the clinical signs elicited a low reading of blood pressure in the left upper arm. A careful palpation of lower limbs pulse and measurement of blood pressure were normal. The decision to use vasopressors was revoked. The patients responded to conservative treatment of pancreatitis.

Figures 2: CT scan demonstrating acute pancreatitis involving the whole of the pancreas.

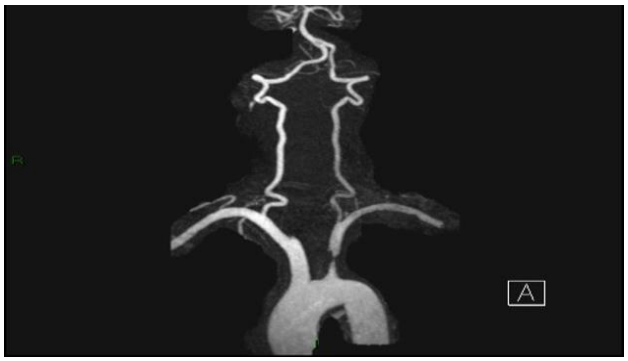

Figure 3: Angiogram showing tight proximal stenosis of the proximal left subclavian artery.

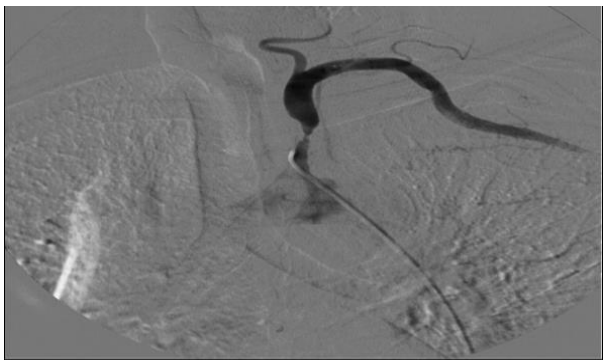

Figure 4: Another view demonstrating tight narrowing of the proximal left subclavian artery.

Doppler examination of the entire vascular system revealed normal venous system in all limbs. The arterial Doppler of the lower limb vessels were normal with triphasic waveforms. The arterial Doppler showed only monophasic waves in both the upper limb arteries. Angiogram revealed stenosis of the left subclavian artery. She had no symptoms in her upper limbs. The patient

refused further investigations such as ERCP and measurement of serum IgG 4 levels.

DISCUSSION

This patient could have possibly had acute pancreatitis with a coincidental and unrelated asymptomatic subclavian artery stenosis. Apart from diabetes mellitus, she had no risk factors for atherosclerosis. Her significant past medical history with two ERCP examinations and biliary stent deployment and removal with the presence of subclavian artery stenosis makes it possible that pancreatitis could be due to systemic cause such as IgG4 related disease.

This patient's refusal for further investigations once her acute abdominal pain settled prevented our assessment of her possible IgG4 related disease.[7]

IgG4 related disease is now understood to be caused by immune mediated process involving many organs with common pathological features. It came to light when extra-pancreatic lesions were seen in patients with type 1 AIP. Involvement of every organ has been described.[8] Commonly after pancreas, the biliary system, kidneys, salivary gland, lymph nodes, thyroid, prostate gland, breast, lung and pituitary gland are involved.

Patients have symptoms related to the involved organ which can become enlarged. Neoplastic and other inflammatory conditions must be ruled out. Diagnosis is made histologically by the presence of IgG4 positive plasma cells, fibrosis in the specimen. Serum levels of IgG4 are often elevated.

Next to type 1 AIP, biliary involvement is common present in about 88% with AIP. Thickening, stenosis and irregularities of the biliary tree can occur. It is commonly described in the intra-pancreatic part of the common bile duct, but can also occur elsewhere. The similar need for 2 ERCPs with biliary stents in our patient – makes this condition a possibility in our patient.

Vlachou et al report one case of subclavian artery stenosis in IgG4 related disease out of 7 patients with vascular involvement in a series of 57 patients with IgG4 related disease.[6] This was similar to the finding we observed in our patient.

Subclavian artery stenosis has a prevalence of about 1.9% in the general population and 7.1% in patients with cardiovascular diseases who have risk factors for atherosclerosis.[9] While it is commonly due to atherosclerosis, other causes include congenital abnormalities such as aberrant origin of subclavian artery causing vascular slings, Takayasu arteritis, IgG4 related disease, trauma and hypercoagulable states.[6,10] The development of collaterals especially around the scapula delay presentation and commonly patients deny symptoms and is identified during differential recordings of blood pressure in the upper limbs.[11,12]

Treatment for IgG4 related disease consists of immunosuppression with steroids first. There is a very good response to steroids in this condition.

CONCLUSION

The response to steroids is dramatic in patients with IgG4 related disease. Hence this disease, which is increasingly being recognised, needs to be considered in the differential diagnosis of patients with pancreatitis and extra-pancreatic manifestations – where patients can anticipate good recovery with steroid therapy.

Meticulous bedside clinical examination is very important and with modern technological advancements in medicine, there is a real risk that periodic clinical examination may be given lesser priority. This case illustrates how a simple measurement of lower limb blood pressure avoided an erroneous risk score allocation and unnecessary inotrope therapy.

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