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CASE REPORT

Churg-Strauss Syndrome Presented With Abdominal Pain And Foot Drop

ABBAS MT*, ERRAYES M, MALALLAH H***, KUMAR R****, KHUDAIR RF*****

ABSTRACT

Abdominal complications are occasionally reported in the Churg-Strauss syndrome (CSS), cholecystitis, pancreatitis, and diffuse vasculitis. We report a case of a patient with CSS, with all these complications. A 33 year old man with an eight year history of asthma and recurrent sinusitis was admitted for abdominal pain. Later on, a vasculitic skin rash developed on the left foot drop.. An increasing peripheral oeosinophilia rising from 25% to 49% in the past month was detected. His pancreatic and liver enzymes increased, and abdominal ultrasonagraphy performed, disclosed a partially distended thick-walled gall bladder with diagnosis of acalculous cholecystitis. Computed tomography (CT) scan of his abdomen showed normal thickness of the bowel wall with oedema in the mesentery, suggestive of an inflammatory process, with patent major mesenteric vessels. Upper endoscopy done, showed diffuse gastritis with multiple mucosal petechiae and positive perinuclear antineutropil cytoplasmic antibody (p-ANCA) of 164 IU/Ml. Nerve conduction study showed severe prominent axonal damage of the sural and tibial nerves in the left leg, and normal peroneal nerve parameters, suggestive of Mononeuritis multiple. The diagnosis of Churg-strauss syndrome was made, with small vessel abdominal vasculitic process.Intravenous pulse methylprednisolone 250 mg/day for three days was given, and a pulse dose of cyclophosphamide was given. His condition showed dramatic improvement and was discharged on oral steroids.

Key-Words: Asthma, abdominal pain, oeosinophilia, Churg-Strauss.

*Senior Specialist/ Department of Medicine/Hamad general Hospital/Doha-Qatar.
**Consultant/ Department of Medicine/Hamad general Hospital/Doha-Qatar.
***Consultant Rheumatology/ Department of Medicine/Hamad general Hospital/Doha-Qatar
****Senior Specialist/ Department of Radiology/Hamad general Hospital/Doha-Qatar.
*****Medical resident/ Department of Medicine/Hamad general Hospital/Doha-Qatar

Corresponding Author:
Dr. Mushtak Talib Abbas (MD) (MRCP)
Senior Specialist/ Department of Medicine
Hamad general Hospital/Doha-Qatar
E-mail: amushtak@hotmail.com.
PO Box 3050. Tel: 009745220486
Fax 009744392273.

Introduction

Churg and Straus first described this syndrome at 1951 in 13 patients who had bronchial asthma, oeosinophilia, granulomatous inflammation, necrotizing systemic vasculitis, and necrotizing glomerulonephritis, and this was called Churg–Strauss syndrome (CSS)[1]. It is a serious but rare pauci-immune vasculitis of small- and medium-sized blood vessels, commonly seen in association with bronchial asthma and/or allergic disorders[2], characterized by the presence of asthma, hyperoeosinophilia, and vasculitis in any part of the body. Vasculitis is often associated with significant distortion of normal functions of the organ involved.

Case History

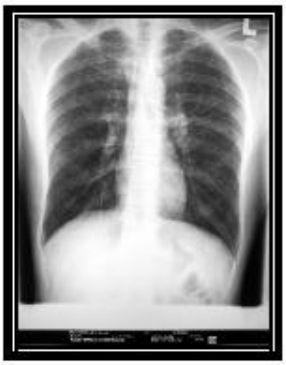
A 33 year old male patient, a known case of Asthma for the past 8 years which was controlled by steroid and B2 agonist combined inhaler and courses of oral steroid during acute exacerbations, presented with a one month history of diffuse and constant abdominal pain, with maximum intensity in the epigastric region which worsened after any oral intake, associated with episodes of vomiting, with no change in bowel habits, and without any haematemsis or melena. There was no history of fever. The condition was associated with mild shortness of breath and cough with wheezing, but without orthoponea or paroxysmal nocturnal dysponea. The condition was associated with a one week history of left ankle pain with swelling with red skin rash over it, generalized fatigue, anorexia, and loss of weight of more than 15 kg in 2 months. No monteulokast therapy was given before. Four days after admission, the patient started to feel numbness in his left foot and toes, with an inability to move his big toe and ankle movements at full range. He had a past medical history of allergic rhinitis and repeated attacks of sinusitis, with an average monthly acute attack of asthma responding to short courses of intravenous and oral steroid.

On examination, the patient was fully conscious, oriented, looked unwell, cachectic, in pain, and not dyspneic, with a temperature of 36.8 C, Blood pressure 125/60, Pulse rate 94/minute, RR 18/minute, Body Weight: 44kg, Height: 171cm, Body mass index of 15, and oxygen saturation 95-99% on 1 L/min oxygen. He had scattered expiratory rhonchi heard all over his chest, with normal first and second heart sound and no heart murmur. His abdomen was soft, not distended with tenderness all over, abdomen was mainly epigastric and periumblical region; [d1]no organomegaly or ascitis, bowel sound was normal. Musculoskeletal examination showed purpuric palpable non blanching skin rash with some lesions which were haemorrhagic over the dorsum of the left foot. The left ankle joint was warm, swollen and tender, with limitation of its movement. There was no involvement of other joints.
Investigations in the form of haematological, serological and biochemical tests with urine microscopic examination are shown in [Table/Fig 1]. Chest X-ray showed accentuated hilar and medial bronchovascular marking and hyper inflated chest [Table/Fig 2]. Ultrasound abdomen showed that the gall bladder (GB) was partially distended with thick wall, with no GB stones seen, and no evidence of biliary obstruction with normal size of spleen and liver. [Table/Fig 3] Computed tomography of paranasal sinuses showed pan sinusitis [Table/Fig 4], and CT of abdomen with angiography of the mesenteric vessels showed normal mesenteric blood vessels and normal thickness of the bowel wall with oedema in the mesentery, suggestive of an inflammatory process, and with no evidence of mesenteric ischaemia [Table/Fig 5]. The patient was adviced to have skin biopsy from the skin lesion but he refused that strongly, because that was not his main concern. Instead, he was concerned about his inability to intake food orally and his increasing abdominal pain. Nerve conduction study was done, which showed severe prominent axonal damage of the sural and tibial nerves in the left leg, and normal peroneal nerve parameters, suggestive of Mononeuritis multiplex. Upper endoscopy

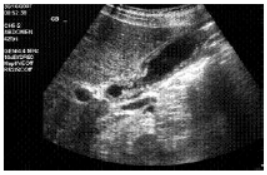
was done, which showed diffuse gastritis with multiple mucosal petechiae. No biopsy was taken from these lesions.

Table/Fig-1: Hematological, serological and biochemistry with urine exam.

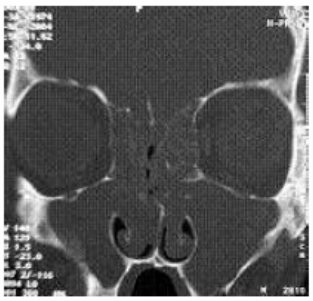
Hemoglobin	16.1 g/dl	Glucose	5 mmol/l
Hematocrite	40%	Urea	5.4 mmol/L
MCV	81fl	Creatinine	80 umol/l
WBC	17.2 u/l	Protein	63 gm/l
Neutrophil	55.0%	Albumin	40 gm/L
Lymphocyte:	3.5 %	Calcium	2.4 mmol/l
Eosinophils	35.8%	Potassium	4.4 mmol/l
Monocyte	2.5%	Sodium	136 mmol/l
Platelet	349	Chloride	99 mmol/l
ESR	49	HCO3	29 mmol
C reactive protein	164	Cholesterol	3.7 mmol/l
Peripheral smear: not anemic, moderate leukocytosis with eosinophilia		Triglyceride	1.17 mmol/l
Prothrombin time	13 sec	Total bilirubin	17/umol/l
Activated partial thromboblastin time	36 sec	Alkaline phosphates	326 U/L
ECG→ sinus tachycardia		ALT	147 U/L
		AST	56 U/L
Urine R/M		Uric acid	246 umol/L\
Protein	+	Amylase	161
RBC	+++	Lipase	267
Urine microscopy	WBC 12-15 /ul	**IMMUNOLOGY**	
RBC 21-25 /uL		Anti Nuclear Antibody	negative
no cast seen		C3	89.5 mg/dl (normal)
24 hr urine for protein was	0.25 gm/d	C4	16.3 mg/dl (normal)
Blood and urine culture	negative	Hepatitis serology	negative
		Rheumatoid factor	>128 u /Ml
		ANCA	287 Eu/ml
		CANCA	less than 20 EU/Ml
		PANCA	164 EU/Ml
		Immunoglobulin E	4777 KU/L



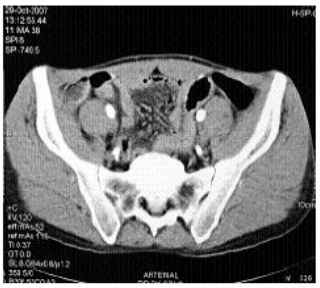
Table/Fig2: showed accentuated hilar and medial brochvascular marking and hyperinflated chest.



Table/Figure 3: Ultrasound abdomen showed gall bladder partially distended with thick wall, no GB stones seen, no evidence of biliary obstruction with normal size of spleen &liver.



Table/Fig 4: Computed tomography of para nasal sinuses showed pan sinusitis



Table/Fig 5: Computed tomography of abdomen with angiography showed normal mesenteric blood vessels and normal thickness of the bowel wall with edema in themesentery suggestive of inflammatory process, no evidence of mesenteric ischemia.

In the presence of history of Asthma, allergic rhinitis, pan sinusitis, oeosinophilia, evidence of vasculitic skin rash, positive p-ANCA and Mononeuritis multiplex, the diagnosis of Churg-Strauss syndrome was made, with diffuse abdominal small vessel vasculitis. He was started on intra venous methylprednislone 250 mg/day for 3 days, and was then shifted to 40 mg of oral prednisolone, daily. In addition, a pulse dose of cyclophosphamide was given. Within 10 days, the patient improved dramatically, with no more abdominal pain and was able to intake food orally, with disappearance of oeosinophilia, and skin lesions. Prednisone was planned to be given, and to be gradually tapered over 2 years. After one month from discharge, the patient gained a weight of 5Kg.

He was discharged on oral prednisolone 60mg once daily, to be tapered over 2 years, and on pulse cyclophosphamide per month. With regular follow up in clinic, he is in good general condition, and back to his normal life.

Discussion

CSS is a rare systemic vasculitis with an annual incidence of approximately 1.8 to 4.0 per 1 million people.[3],[4] The prevalence of CSS in asthmatic subjects is greater, 64.4 per 1 million subject.[4] The mean age at diagnosis is 50 years, but the systemic vasculitic phase is frequently apparent in patients who are in their late 30s[5]. Our patient age is 33 years old.

The gastrointestinal (GI) tract may be involved in 20% to 50% of patients, mainly with abdominal pain, diarrhoea, GI bleeding, and gastroduodenal and colorectal ulcers.[6] Indeed, severe GI involvement has been identified as an independent factor associated with poor outcome.[7] Considering the high risk of perforation of the GI tract, including the small intestine and the poor prognosis, any abdominal pain or discomfort must be carefully investigated.
The Churg-Strauss syndrome (CSS), is a multisystem disorder characterized by allergic rhinitis, asthma, and prominent peripheral blood oeosinophilia[3]. Blood oeosinophilia (more than 10,000/mm3) is present in 95% of cases. Myeloperoxidase-specific antineutrophilic cytoplasmic antibodies (MPO-ANCA) are detected in 70% of patients with active Churg-Strauss syndrome, and may play an important role in the pathogenesis of the disease.[8]
Our patient presented with a long history of asthma and allergic disorders (allergic rhinitis and sinusitis); other features were hyperoeosinophilia, vasculitic mononeuritis multiplex and vasculitic gastroenteritis, which were also a part of the medical problems in the patient. These features satisfied the American College of Rheumatology (ACR)[9] criteria for the diagnosis of CSS. His earlier exposure to corticosteroids could also have been suppressing a preexisting CSS.
Therapeutically, the use of high-dose prednisone is the most common and recommended treatment for intestinal vasculitis[10]. Even so, our case demonstrates that not all patients respond sufficiently to corticosteroid treatment alone, and some develop a relapse when steroid therapy is tapered.

The prognosis of patients with CSS is unclear; treatment appears to be associated with significantly decreased mortality. Prior to the use of glucocorticoids, for example, the disease was uniformly fatal, with 50 percent of untreated patients dying within three months of the onset of vasculitis. In comparison, most modern clinical series suggest a survival rate of greater than 70 percent at five years[7],[9] The presence of significant cardiac or gastrointestinal disease appears to be the strongest indicator of poor prognosis[11].

In conclusion, CSS has broad spectrum clinical presentation. In any patient who present with abdominal pain and neurological symptoms, CSS should be considered.

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