Nontraumatic hepatic hematoma caused by Wegener’s granulomatosis: an unusual cause of abdominal pain

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Abstract

Wegener’s granulomatosis (WG) is a vasculitis of unknown origin characterised by prominent involvement of upper and lower respiratory tract and kidney. There are only a handful of reported cases in the literature about hepatic involvement of WG. This report shows a patient with WG whose main complaint was severe abdominal pain due to nontraumatic subcapsular hepatic hematoma. To our knowledge, this is the first reported case of WG with hepatic hematoma depicted by US and CT in the English literature.

Wegener’s granulomatosis (WG) is a multisystem disorder of unknown aetiology first described in the 1930s. Prevalence in the US is estimated at 3 per 100,000. The common manifestations of the disease include the classic triad of upper airway, lung, and kidney, in 87%, 69%, and 48% of the patients. Gastrointestinal tract and spleen involvement are less common.

There are only a handful of reported cases in the literature about hepatic involvement of WG. Patients present with symptoms that are similar to common diseases. Many patients usually present with chronic fatigue, upper respiratory infection, sinusitis, hearing loss, otitis media, acute interstitial nephritis, or pulmonary haemorrhage.

Malaise, fever, weight loss, arthralgias, myalgias, and rashes are common but rarely dominate the clinical picture. The symptoms of WG may manifest individually and progress slowly.

We describe a patient with WG whose main complaint was severe abdominal pain due to nontraumatic subcapsular hepatic haematoma. To our knowledge, this is the first reported case of WG with hepatic subcapsular hematoma depicted by US and CT in English literature.

Case report

A 27-year-old man presented to our hospital with abdominal pain, weight loss, and multiple purpuric lesions on the lower legs and feet bilaterally which had been gradually worsening over the previous 1 month.

Laboratory values were significant for elevated white blood cell count of 13,310 cells/mm³ with 78% neutrophils, mild anaemia with haematocrit 33%, an erythrocyte sedimentation rate of 72 mm/hr, cytoplasmic antineutrophil cytoplasmic antibody (c-ANCA) titre positive at 1:32. Urinalysis was normal.

Chest X-ray showed a cavitary lesion in the right lung. On the thoracic computed tomography (CT) there was a 2.5cm diameter thin-walled cavitary lesion (Fig. 1).
Fig. 1. Axial CT image shows a cavitary lesion in the right lung (arrow)

Fig. 2A. Ultrasonography images show multiple hypoechoic well defined lesions (arrows) in the liver and spleen

Fig. 2B. Axial CT scan shows multiple hypodense cystic lesions (arrows) in the liver and spleen
Abdominal ultrasonography showed multiple hypoechoic well-defined lesions in the liver and spleen (Fig. 2A).

A contrast-enhanced CT scan of the abdomen showed multiple non-enhancing cysts in the liver and spleen (Fig. 2B). A peripheral angiogram of the lower extremities revealed the decrease of calibrations in the peripheral arteries of the feet bilaterally.

Multiple liver and spleen biopsy specimens showed inflammatory granulomatous tissue which contains necrosis and suppuration. Based on the histopathologic results, clinical picture, radiological findings and laboratory results (c-ANCA titres) the probable diagnosis was WG with multiple organ involvement. The surgeons suggested splenectomy which the patient refused. So the patient was treated with prednisone, 1mg/kg/day, and cyclophosphamide, 2mg/kg/day.

Less than 2 years later, the patient returned to our hospital with a severe right upper abdominal pain. His blood pressure was 80/50mmHg. Laboratory examination showed elevated white blood cell count of 24,742 cells/mm³ with 68% neutrophils, severe anaemia with haematocrit 21%, an erythrocyte sedimentation rate of 76 mm/hr, c-ANCA titre positive at 1:32. His Hb was 6.9g/dl.

An urgent US revealed an enlarged liver with a giant subcapsular cystic mass consistent with hematoma (Fig. 3A). On Doppler ultrasonography, the mass was avascular (Fig. 3B). Abdominal CT scan confirmed a 15x12x7cm diameter subcapsular hepatic hematoma (Fig. 4A) and also showed small cystic lesions in the liver (Fig. 4B).

A blood transfusion was given and patient was treated and followed-up conservatively. Three weeks later the patient was discharged when he was asymptomatic and the size of hematoma had decreased.

Fig. 3A. Ultrasonography image shows a giant hepatic subcapsular cystic mass (arrows)
Fig. 3B. Doppler ultrasonography shows no arterial or venous flow in the cystic mass (arrows)

Fig. 4A. Axial CT scan shows a giant subacute subcapsular hepatic hematoma (arrows)
Fig. 4B. Axial CT image shows a giant subcapsular hematoma (big arrow) and cystic lesions (small arrows) due to WG

Discussion

Wegener’s granulomatosis is a vasculitis of unknown origin characterised by prominent involvement of upper and lower respiratory tract and kidney. Histological pattern consists of the triad of giant cell granuloma, necrosis, vasculitis involving capillaries and small and middle-sized arteries. The case here described is characterised by hepatic nontraumatic hematoma with WG an unusual cause of severe abdominal pain. Spontaneous hepatic bleeding is a rare condition. The most common causes of nontraumatic hepatic hemorrhage are hepatocellular carcinoma and hepatocellular adenoma. Nontumoral causes of hepatic hematoma include HELLP syndrome, amyloidosis, miscellaneous, and intrahepatic aneurysm.

Intrahepatic aneurysm is a very rare condition. Chronic inflammation can lead to arterial aneurysm formation, a characteristic of medium-sized vessel vasculitis, but a very unusual feature of WG. Thirteen cases of WG complicated by arterial aneurysms are reported in the English literature. Only four cases of aneurysmatic dilatation of the hepatic artery due to WG has been described in these literature. In our case the probable cause of the nontraumatic hepatic subcapsular hematoma with WG could be the aneurysm of the hepatic artery. But we could not depict any aneurysms with Doppler US or abdominal CT.
On the other hand a review\textsuperscript{11} described a case of splenic rupture in WG and reported that splenic vasculitis and vessel destruction with hemorrhage, infarct, and resultant necrosis may lead to rupture. In our case perhaps the same mechanism may lead to the hepatic subcapsular hematoma.

Hepatic hematoma can be easily diagnosed by US and CT. CT can be useful in defining the extent of the hematoma and showing density changes related to the age of the hematoma and can often indicate the underlying cause.\textsuperscript{8,12}

During the acute stage (the first 24–72 hours), the hematoma is hyperattenuating, but it decreases in attenuation and develops a pseudocapsule by 10–30 days.\textsuperscript{8} In our case the hematoma was at a subacute stage and we used these imaging methods to follow the diameter of the hepatic hematoma.

In conclusion, hepatic involvement and hepatic hematoma of WG in a patient whose main complaint is severe abdominal pain is very rare. CT and US can be useful to diagnose and follow-up.

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