CASE REPORT

HEPATIC INVOLVEMENT IN EOSINOPHILIC GRANULOMATOSIS WITH POLYANGITIS, AN ATYPICAL MANIFESTATION

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ABSTRACT

Eosinophilic granulomatosis with polyangiitis (EGPA) is a systemic necrotizing vasculitis mainly affecting small-sized arteries. It usually presents with pulmonary and renal involvement with rare hepatic manifestation. We report the case of a 33-year-old young adult, known case of EGPA who presented with dry cough, hemoptysis, fever, right hypochondrial pain. His contrast enhanced CT chest and abdomen demonstrated hepatic aneurysm with hematoma in the right hepatic lobe. What makes this case worth reporting is the coexistence of liver aneurysms and hematoma with concomitant systemic disease affecting the lungs with keeping in mind that limited forms of GPA with atypical site involvement might occur. Until recently, little had been reported in radiology literature regarding polyarteritis nodosa.

Keywords: Granulomatosis polyangiitis, hepatic involvement.

Introduction

Eosinophilic granulomatosis with polyangiitis is an autoimmune small and medium vessel necrotizing vasculitis associated with both granulomatosis and polyangiitis. It is characterized by necrotizing granulomatous lesions of the respiratory tract, along with necrotizing glomerulonephritis and widespread disseminated vasculitis.[1] The primary abnormal changes of the involved vessel consist of fibrinoid or hyaline necrosis of the media with simultaneous or subsequent involvement of the intima and adventitia. Secondary changes include aneurysm formation, hemorrhage, and thrombosis. Lung parenchymal disease is the most frequent manifestation which produces multiple nodules and masses[2] It is followed by involvement of kidney. Other less frequently involved organ systems include the central and peripheral nervous system, skin, muscles, large joints, heart, and eyes. Rarely, numerous other sites can be affected. Virtually any organ can be attacked, and in some patients, GPA can manifest unusually.[3]

Case Summary

A known young EGPA patient visited to the emergency department in Liaquat national with dry cough, hemoptysis, fever, abrupt squeezing right hypochondrial pain. On baseline blood investigations, he showed drop in hemoglobin which raises the possibility of pulmonary hemorrhages. His contrast enhanced CT chest and abdomen showed varying sizes intraparenchymal and subpleural nodules in both lungs with surrounding ground glass haze. One of the nodules in right lower lobe shows cavitation. On abdominal sections, a large hematoma is demonstrated in liver occupying almost whole of the right lobe of liver. A slight tortuous vessel is seen extending from inferior vena cava in right lobe of liver. It shows nodular ending likely representing aneurysm.

Given the clinical situation,emergency abdominal angiography was then performed, celiac axis was cannulated and hepatogram was obtained. It shows multiple areas of narrowing and aneurysms in branches of both right and left hepatic arteries so

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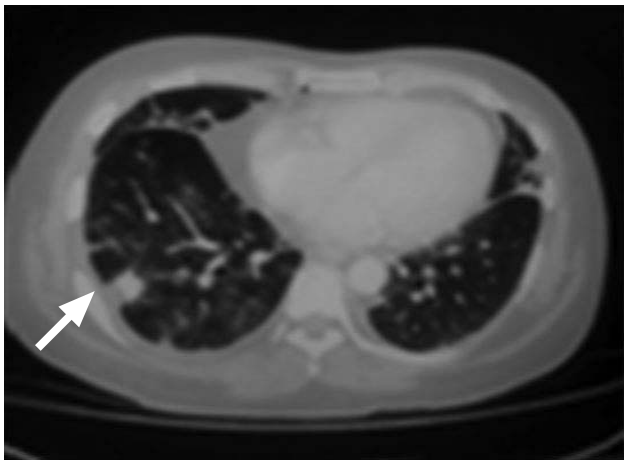

Figure 1a: A subplueral nodule with surrounding ground glass haze (arrow)

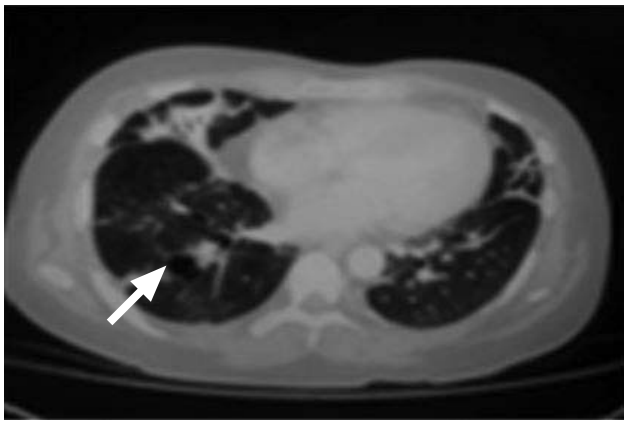

Figure 1b: A cavitatory nodule in right lower lobe (arrow)

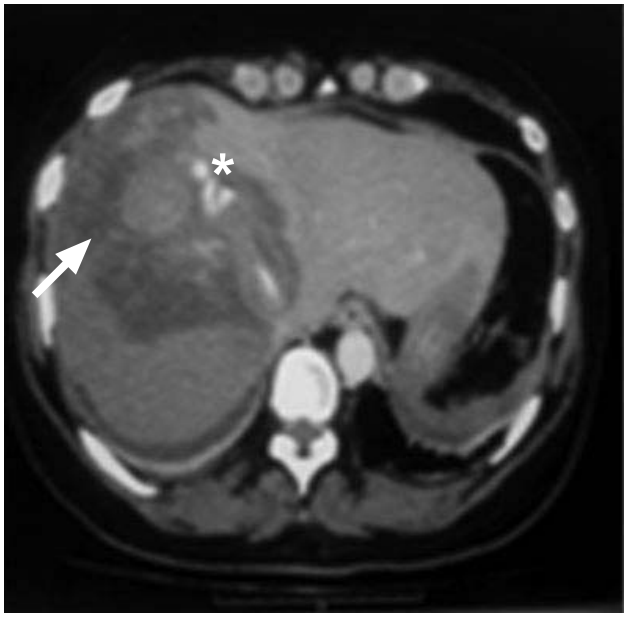

Figure 1c: Haematoma in right lobe of liver (arrow), and a dilated vessel with blind ending anuerysm (sterick)

selective embolization of segment IV arteries was done.

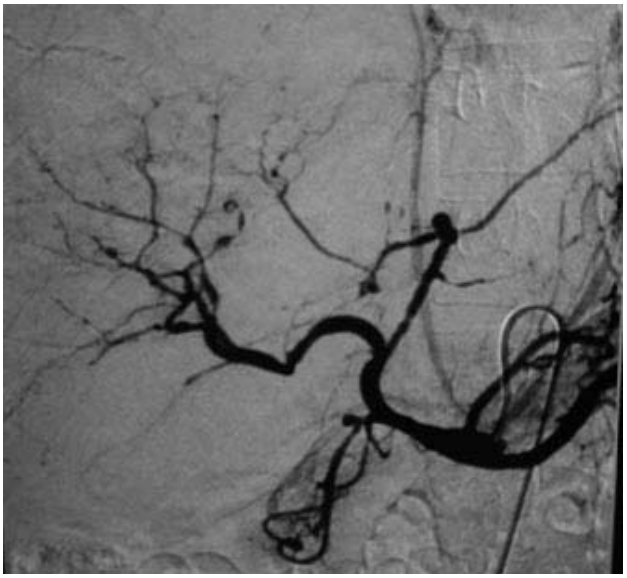

Figure 2a: Hepatogram shows multiple aneurysmal dilatations (arrows) involving branches of hepatic artery

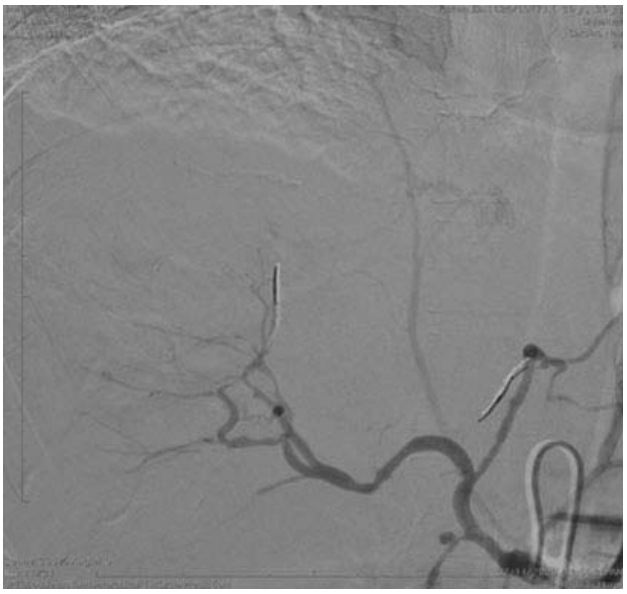

Figure 2b: Post embolization of segment IV artery .

Discussion

EGPA is a predominantly reno pulmonary disorder, rarely has gastrointestinal system manifestations. The involvement of this system usually occurs long after the onset of initial symptoms.[1,2] Among them, the pancreas, liver or colon involvement is exceedingly

rarely reported[3] Similarly, liver involvement in GPA is very rare with only few case reports in the literature, presenting patients with concomitant systemic disease affecting the lungs.[3] Many authors have agreed that angiography helps to confirm the suspected clinical diagnosis. Findings of multiple, small-sized aneurysms on angiography are pathognomonic for the disease. The aneurysm are usually multiple (most often 10 or more in any one visceral circulation) and 2-5 mm, commonly affecting the branch points of arteries.[1-3] In older studies, hepatic involvement was not a feature of GPA, however, isolated case reports have been published mentioning of hepatic involvement in GPA patients. The most common hepatic complications in patients with polyarteritis nodosa as reported in the literature are hepatic infarction or aneurysm formation and rupture, acute liver failure, ischemic cho-langitis and nodular regenerative hyperplasia.[1-4] The diagnosis is ideally made by means of biopsy of involved tissue in a patient with the appropriate clinical symptoms and laboratory data, but an angiogram provides the proof in some cases. Most patients with PAN have positive angiographic evidence of their disease, predominantly in the visceral arteries but also in arteries of the extremities and in small branches of the aorta.[4,5] The most well-known angiographic feature is the presence of so-called microaneurysms in medium or small arteries.[5] In our case, patient is having multisystem involvement, including pulmonary and hepatic manifestations. Though after successful embolization, patient maintained his hemoglobin, but due to fulminant sepsis, this resulted in fatal outcome.

Conclusion

There should be awareness of less frequent manifestations, seemingly not compatible with EGPA or even mimicking other diseases and to keep in mind that limited forms of EGPA with atypical site involvement might occur. Therefore, it should not be forgotten as a manifestation of EGPA.

Conflicts of interest: There are no conflicts of interest.

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