Splenectomy and unilateral nephrectomy complicating granulomatosis with polyangitis

Sir,

A 20-year-old, female presented in September 2005 with chronic sinusitis, hypertension (150/100 mmHg), and elevated serum creatinine of 9.58 mg/dl. Her further workup was showed an elevated anti-proteinase 3 (PR3) titer at 69 IU/ml and a renal biopsy showed pauci-immune crescentic glomerulonephritis. A diagnosis of granulomatosis with polyangitis (GPA) was made, and she was treated by plasma exchange, oral prednisolone, mycophenolate mofetil, and antihypertensive drugs. The patient was lost to follow-up for 5 years until she presented currently with purulent ear discharge, macroscopic hematuria, and abnormal renal functions. Her investigations showed high C-reactive protein at 20.8 mg/dl, anti-PR3 >100 IU/ml, and urine protein creatinine ratio of 4.8. Her family refused any sort of active intervention apart from treating her infections. She presented 1 week later with significant pallor, abdominal pain, developed hypotension, and anemia. The patient was resuscitated and an abdominal ultrasonography showed perinephric hematoma. Computed tomography angiography it showed multiple small aneurysms in kidneys, spleen, and liver. After stabilizing, she was started on methylprednisolone 500 mg/day. One week later, she presented in hemorrhagic shock requiring exploration which resulted in splenectomy and left nephrectomy. Pathological examination of the kidney and spleen showed multiple hemorrhagic areas [Figure 1a and b], ischemic infarcts, and foci of necrotizing granulomatous vasculitis [Figure 1d and e]. In addition, the kidney showed diffuse pauci-immune crescentic and necrotizing glomerulonephritis [Figure 1c].

GPA is a necrotizing granulomatous vasculitis with clinical predilection for the upper airways, lungs, and kidneys, preferentially involving venules, capillaries, and arterioles, but may also affect arteries and veins. Arterial aneurysms are characteristic of medium-size vessel vasculitis occurring in 50-60% of polyarteritis nodosa, but are quite unusual of GPA. Approximately 16 cases of GPA complicated by arterial aneurysms of large and medium-size vessels are documented in the literature.[1-4]

Literature review showed that the incidence of these aneurysms was more in men and they occurred at the onset than during the course of the disease.[1] Involvement of vessels such as renal, hepatic, aorta, gastric, splanchnic, and cerebral arteries has been reported.[1-4] Chronic granulomatous inflammation with fibrinoid necrosis results in aneurysmal dilatation. Abdominal pain is the most common warning sign of intraabdominal arterial aneurysm. Rupture of an aneurysm in GPA is life threatening as this can lead to death from fatal intraabdominal bleeding.

Treatment of aneurysmal vasculitis should combine aneurysm embolization, when possible, with prompt high-dose steroids and immunosuppressive agents to prevent life-threatening rupture. Watchful waiting for

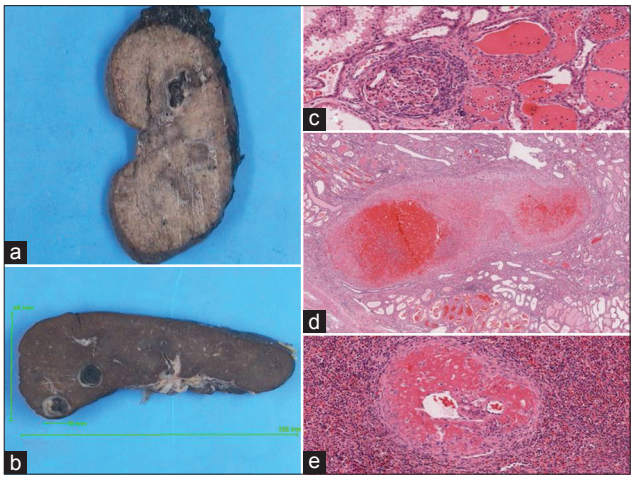

Figure 1: Kidney (a) and spleen (b) with hemorrhagic areas. Kidney with crescentic glomerulonephritis (c) (H and E, ×400). Kidney (d) (H and E, ×400) and spleen with necrotizing granulomatous vasculitis (e) (H and E, ×400)

any alarming signs, evaluation by imaging modalities complemented by angiographic studies, and timely intervention, either medical or surgical, can be life saving in this clinical scenario.

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References

1. Arlet JB, Le ThiHuong D, Marinho A, Cluzel P, Wechsler B, Piette JC. Arterial aneurysms in Wegener's granulomatosis: Case report and literature review. Semin Arthritis Rheum 2008;37:265-8.
2. Ohta N, Waki T, Fukase S, Suzuki Y, Kurakami K, Aoyagi M, *et al*. Aortic aneurysm rupture as a rare complication of granulomatosis with poyangiitis: A case report. J Med Case Rep 2013;7:202.
3. Senf R, Jürgensen JS, Teichgräber U, Kampf D, Schindler R. Ruptured arterial aneurysm of the kidney in a patient with Wegener's granulomatosis. Nephrol Dial Transplant 2003;18:2671-3.
4. Durai R, Agrawal R, Piper K, Brohi K. Wegener's granulomatosis presenting as an abdominal aortic aneurysm: A case report. Cases J 2009;2:9346.

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Quick Response Code:	Website: www.indianjnephrol.org
	DOI: 10.4103/0971-4065.127923