Primary Antiphospholipid Antibody Syndrome Presenting as Unilateral Adrenal Hemorrhage

Sir,

We came across a 35-year-old apparently healthy woman presenting with an acute onset right hypochondriac pain. She was hemodynamically stable. Computed tomography scan showed the right adrenal mass which was moderately hypodense (HU: −50 to −55) on plain scan and did not show any contrast enhancement, suggestive of acute hemorrhage. The left adrenal was normal [Figure 1a and b]. There were no features suggestive of adrenal hypersecretion (Cushing's syndrome, pheochromocytoma, and primary aldosteronism). She was single and had normal menses and no medical history in the past. Twenty-four hours' urinary metanephrines, 8 am serum cortisol, and adrenocorticotropic hormone were normal. The patient remained stable. Pain subsided with supportive treatment. The first differential diagnosis was adrenal hemorrhage in a tumor which was probably not clearly delineated as it was obscured by the bleeding. In that case, there was a possibility that the tumor may resolve with the hemorrhage or may be well visualized with biochemical evidence of hypersecretion with resolution of hemorrhage. The second possibility was spontaneous hemorrhage in a normal adrenal gland. As the patient was recovering, the decision of watchful waiting was taken. She had severe thrombophlebitis at the site of intravenous line insertion. The venous Doppler revealed thrombophlebitis involving superficial veins of the dorsum of hand and along cephalic and basilic veins in the distal forearm.

Follow-up magnetic resonance imaging after 1 month revealed subacute right adrenal hemorrhage. It was hyperintense on T2 and centrally hypointense with peripherally hyperintense rim on T1 [Figure 1c and d]. Follow-up scan at 4 months revealed resolved right adrenal hemorrhage [Figure 1e and f]. The left adrenal gland was normal throughout.

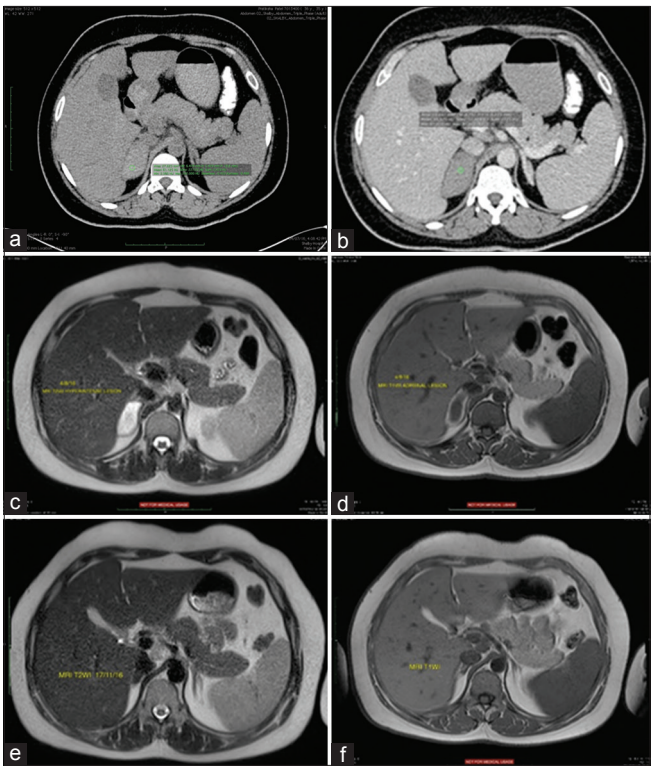

Figure 1: Serial imaging of the right adrenal mass is shown. The left adrenal is normal throughout computed tomography at baseline. (a) computed tomography (plain): Moderately hypodense right suprarenal lesion (HU: −50 to −55). (b) computed tomography (contrast): No enhancement (HU: Same as that of plain) magnetic resonance imaging (after 1 month from baseline). (c) T2: Well-capsulated hyperintense lesion in the right suprarenal. (d) T1: Well-capsulated peripherally hyperintense and centrally hypointense lesion. Magnetic resonance imaging (after 4 months from baseline). (e) T2 and (f) T1 images showing complete resolution of the right adrenal hemorrhage

To look for the cause of severe thrombophlebitis, hypercoagulation profile was advised. It revealed elevated IgM and IgG antiphospholipid antibody (APLA). Dilute Russell's viper venom test was consistent with the presence of lupus anticoagulant (LA), and it remained positive after 12 weeks as well. Thus, this patient had spontaneous right adrenal hemorrhage in the normal adrenal gland with superficial vein thrombosis with APLA positivity and persistent LA positivity 12 weeks apart confirming the diagnosis of APLA syndrome. The clinical and laboratory workup for coexisting rheumatological disorder (including antinuclear antibody) was negative. Hence, the diagnosis of primary APLA syndrome was confirmed. The patient was started on anticoagulation therapy.

Unilateral adrenal hemorrhage has been associated with trauma, sepsis, anticoagulation, hematologic disorders, pregnancy, and adrenal masses (metastases, pheochromocytomas, adrenocortical cancers, or hematomas masquerading as neoplasms).[1]

APLA syndrome may occur with coexistent autoimmune disease (secondary APLA) or without coexisting autoimmune disease (primary APLA).[2] Criteria to confirm the diagnosis of APLA include at least one clinical sign, such as vascular thrombosis or pregnancy complications, and one biochemical criterion such as APLA (IgG/IgM) and LA (IgG/IgM) detected in two occasions at least 6–12 weeks apart.[2]

Presotto *et al*. identified twenty cases of primary adrenal insufficiency as the heralding symptom of primary APLA. In 17 patients where imaging was available, 14 had bilateral whereas 2 had unilateral hemorrhage and 1 had bilateral normal adrenal glands.[3] Espinosa *et al*. reviewed 86 patients with APS and primary adrenal insufficiency. In 69 patients where imaging was available, adrenal hemorrhage was seen in 40 patients.[4]

Patients with APLA syndrome have hypercoagulable state. Adrenal hemorrhage in these patients is nothing but hemorrhagic infarction of adrenal gland likely to be related to the adrenal vein thrombosis. The unique vascular structure of adrenal glands in the form of rich arterial supply but a limited venous drainage by a single vein predisposes this gland for the secondary hemorrhagic infarction after venous thrombosis. The capillaries branching from the arterial system form a vascular plexus around the zona reticularis. Due to the abrupt transition from artery to capillary plexus, a "vascular dam" is formed. Walls of adrenal veins have eccentric longitudinal muscular arrangement. When these muscle bundles contract, pockets of turbulence and local stasis are formed where platelet thrombi are formed.[3,5]

Thus, in a patient presenting with unilateral spontaneous adrenal hemorrhage, it is worthwhile to look for possibility of APLA syndrome. In such case with APLA positivity, adrenal surgery can be avoided with watchful waiting. Moreover, with diagnosis of APLA syndrome, further workup for associated rheumatological disorder can be initiated. The patient can be started on anticoagulation to prevent further thrombotic episodes.

Acknowledgment

We would like to thank Dr. Manjunath Goroshi for his valuable inputs in managing this patient.

Financial support and sponsorship

Nil.

Conflicts of interest

There are no conflicts of interest.

Shruti Khare, Hardik Patel[1], Gargey Sutaria[2], Reena Sharma[3]

Departments of Endocrinology, [1]Gastrosurgery, [2]Radiology and [3]Rheumatology, Shalby Hospital, Ahmedabad, Gujarat, India

Address for correspondence:
Dr. Shruti Khare,
Shalby Hospital, Ahmedabad, Gujarat, India.
E-mail: shrutigkhare@yahoo.co.in

References

1. Marti JL, Millet J, Sosa JA, Roman SA, Carling T, Udelsman R, *et al*. Spontaneous adrenal hemorrhage with associated masses: Etiology and management in 6 cases and a review of 133 reported cases. World J Surg 2012;36:75-82.
2. Negrini S, Pappalardo F, Murdaca G, Indiveri F, Puppo F. The antiphospholipid syndrome: From pathophysiology to treatment. Clin Exp Med 2017;17:257-67.
3. Presotto F, Fornasini F, Betterle C, Federspil G, Rossato M. Acute adrenal failure as the heralding symptom of primary antiphospholipid syndrome: Report of a case and review of the literature. Eur J Endocrinol 2005;153:507-14.
4. Espinosa G, Santos E, Cervera R, Piette JC, de la Red G, Gil V, *et al*. Adrenal involvement in the antiphospholipid syndrome: Clinical and immunologic characteristics of 86 patients. Medicine (Baltimore) 2003;82:106-18.
5. Fox B. Venous infarction of the adrenal glands. Journal of Pathology 1976;119:65-89.

Access this article online

Quick Response Code:

Website: www.ijem.in

DOI: 10.4103/ijem.IJEM_474_17

How to cite this article: Khare S, Patel H, Sutaria G, Sharma R. Primary antiphospholipid antibody syndrome presenting as unilateral adrenal hemorrhage. Indian J Endocr Metab 2017;21:932-3.