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Case Report
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Bilateral Spontaneous Adrenal Hemorrhage in Antiphospholipid Syndrome

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Abstract

Antiphospholipid Syndrome (APS) is characterized by arterial and/or venous thrombosis independent of the size of the vessels. Deep veinthrombosis, pulmonary embolism and cerebrovascular thromboembolism represent the most frequent clinical manifestations while adrenal vessels are very rarely involved.

Here we describe the case of a young male with primary APS who developed acute adrenal failure due to bilateral hemorrhage and we underline that full anticoagulation is the therapy of choice in patients with bleeding and high thrombophilic profile.

Case Presentation

A 23 year old man (Kg 80) with primary antiphospholipid syndrome and a past history of pulmonary embolism was admitted to the Emergency Department due to severe chest and flank pain in the right side. He had been treated with warfarin for five years, and then warfarin was interrupted and replaced with enoxaparin 120 mg die for a dental extraction. After the extraction the patient stopped enoxaparin and the day after he should have resumed warfarin, but in the afternoon he felt ill and went to the Hospital. He didn't understand that he should have taken enoxaparin and warfarin together for some days.

At admission a thorax CT scan was negative for pulmonary embolism and an abdomen ultrasound showed a mild edema of the perirenal right fat. Due to the severe pain, a CT scan of the abdomen was performed and the enlargement of the right adrenal gland with signs of recent bleeding was evident (Figure 1).

Enoxaparin was stopped and pain relief therapy with opiates was started.

Four days later hypotension (100/70 mmHg), hypoglycemia (70 mmHg) and severe pain in the left side appeared and a drop in the hemoglobin levels was noted (from 14 g/dl to 12.5 g/dl). A second CT scan showed a new onset adrenal hemorrhage in the left side (Figure 2), so enoxaparin therapy- 160 mg die- was immediately restarted. Laboratory investigations confirmed the presence of lupus anticoagulant and high titres of IgG anticardiolipin antibodies (1375 U/ml) and IgG anti beta 2 glycoprotein I (6100 U/ml). C3 and C4 levels were normal and test for antinuclear antibodies was negative. Blood cultures and procalcitonin resulted negative as well. After three days from resumption of anticoagulants, CT scan was repeated and the bilateral adrenal gland enlargement appeared unchanged.

After the first hemorrhage cortisol level was 7.4 microgram/dl (n.v. 4.3 µg/dl to 22.4 µg/dl), but dropped to 1 microgram/dl after the second bleeding despite the beginning of cortone acetate therapy. In the following days abdominal pain disappeared and the patient felt progressively better without other signs of adrenal failure. He was discharge with cortone acetate and warfarin and he was advised to keep INR between 2.5 and 3.5.

Discussion

The most common causes of primary adrenal failure is the autoimmune destruction of the glands, infections, especially tuberculosis in the past, metastatic cancer, adrenal hemorrhage, infarct and medications. Conditions associated with hypercoagulation rarely cause adrenal failure

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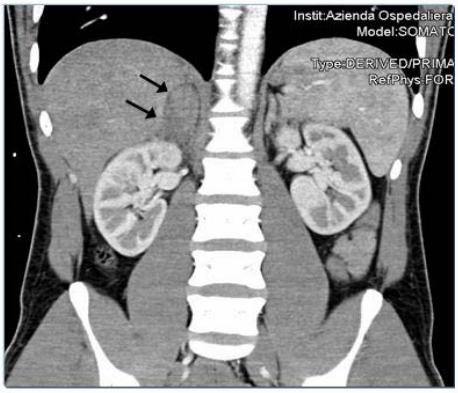

Figure 1: Angio-Multilayer CT. Coronal reconstruction.Gland enlargement and increase in right adrenal density as in blood suffusion. Left adrenal gland was normal.

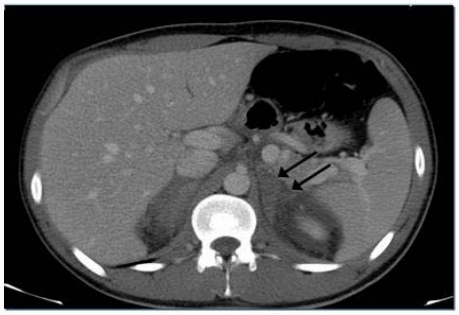

Figure 2: Multilayer CT scan performed in another phase. Post-mdc axialscan. Slight increase in findings on the right and volume increase and slight fluids of fusion in the left gland.

and, in particular, the APS accounts for <0.5% of Addison disease [1]. On the other hand adrenal failure represents the most common endocrinologic manifestation of APS, is more common in males (55%) and appears as the first manifestation of APS in 36% of cases [2]. APS is considered if at least one clinical criteria, arterial and/or veinthrombosis and pregnancy morbidity, and at least one of the laboratory criteria, such as the presence of antiphospholipid antibodies (aPL) on two or more occasions at least 12 weeks apart, are fulfilled [3]. aPL are directed against phospholipid-binding proteins and those included in the classification criteria of APS are Lupus Anticoagulant (LAC), anticardiolipin antibodies (IgG or IgM) and anti-beta 2-glycoprotein I antibodies(IgG or IgM).

APS can be an isolated disorder (primary APS) or can be accompanied by other autoimmune diseases, especially systemic lupus erythematosus (secondary APS) [4].

The majority of post-mortem studies indicate that the most frequent pathogenetic mechanism of bleeding is a thrombosis-mediated adrenal venous occlusion with subsequent edema of the gland, secondary obstruction of the arterial supply and hemorrhagic infarction [5]. Adrenal gland has a particular vascular anatomy with a high flow arterial network and a single vein for drainage, so this feature may predispone to venous thrombosis and secondary bleeding.

More rarely histopathology reports demonstrated spontaneous adrenal hemorrhage without vessel thrombosis, especially in patients undergoing surgery or are receiving anticoagulation [2].

In the case described here the first bleeding occurred the day after the patient had stopped enoxaparin. Even if not in full dosage, we initially suspected a spontaneous bleeding, maybe due to a minor trauma, and we stopped enoxaparin also taking into account that there weren't thrombi in other sites.

But when the bleeding of the contralateral adrenal gland was evident in the second TC scan, we thought that the underlying mechanism of bleeding was another, thrombosis-related, and were started enoxaparin at full dosage.

Pain improvement and a CT scan after few days confirmed that therapy was right.

There wasn't enough time to organize an arteriography or a biopsy to confirm the suspect of thrombosis-related bleeding because the patient was ill and the other gland had been already damaged. So, despite some doubts raised from the CT images were started enoxaparin 160 mg die. We didn't think to an immune-mediated adrenalitis because ANA was negative and the patients didn't present signs and/or symptoms of autoimmune diseases different from APS.

In conclusion, even if very rarely, APS can cause adrenal failure due to bilateral thrombosis-related hemorrhage and, despite the evidence of bleeding, just clinical suspicion makes anticoagulants the therapy of choice in patients with so high thrombophilic profile.

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