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Catastrophic antiphospholipid syndrome presented with sudden renal failure and history of long-lasting psychosis and hypertension in a 42 years old women

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The catastrophic variant of the antiphospholipid syndrome (CASP) is defined as a potential life-threatening variant of antiphospholipid syndrome (APS), which is characterized by multiple small-vessel thrombosis that can lead to organ failure especially renal deterioration. This vaso-occlusive nephropathy mainly affects arterioles, interlobular arteries and glomerular tufts. However, interstitial area and tubules maybe subsequently involved.

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Brief Report

1. Introduction

The catastrophic variant of the antiphospholipid syndrome (CAPS), which is also named as Asherson's syndrome (1-4), is defined as a potential life-threatening variant of this syndrome, which is characterized by multiple small-vessel thrombosis that can lead to multi-organ failure, particularly renal deterioration (2-4). A renal small-vessel nephropathy, defined as antiphospholipid syndrome nephropathy (APS nephropathy; APSN), has been described (3,4). This vaso-occlusive disease mainly affects arterioles, interlobular arteries and glomerular tufts. However, interstitial area and tubules may subsequently be involved (2-7).

2. Case

A 42-year-old woman was admitted in the emergency ward because of confusion which started five days ago. She had also suffered from nausea and vomiting. Patient was a passenger, who came from a neighboring province. On examination, a 160/90 mmHg of arterial blood pressure, asterixis and fever was found. Other physical examinations were normal. In the past medical history, a history of hypertension (under

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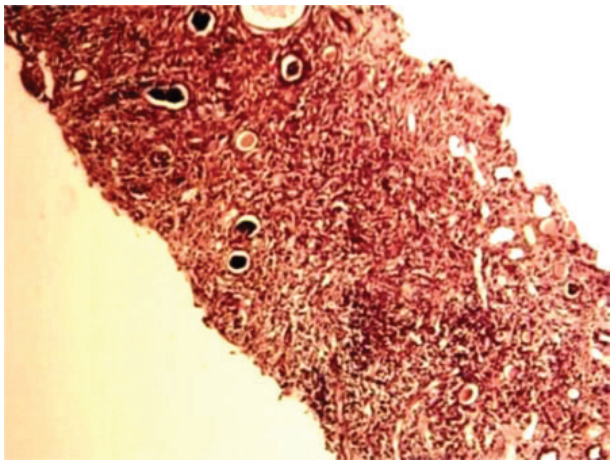
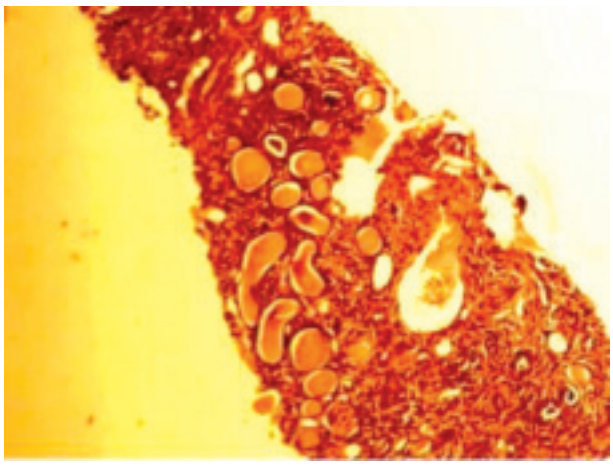
treatment of atenolol; 100 mg/day) for 10 years and psychiatric disturbance, bipolar type (which was under treatment of Lithium carbonate, Carbamazepine and Amitriptyline) from 5 years ago were identified. Laboratory tests revealed anemia, elevated ESR (102 mm/1hr) and raised serum creatinine (10 mg/dL). Urinalysis showed mild proteinuria and pyuria. Ultrasonography showed normal size kidneys with increased parenchymal echogenicity. Urine protein excretion during 24 hours was 280 mg. With the primary diagnosis of acute renal failure (ARF), hemodialysis was started. Patient was also treated by appropriate antibiotic and other supportive therapies. Because, there was not any improvement after 18 days of admission and with the primary impression of acute tubular necrosis or interstitial nephritis due to chronic Lithium therapy, patient underwent kidney biopsy. Light microscopic examination disclosed global sclerosis in four out of eight glomeruli and four a tuft glomeruli (glomerular ballooning). In glomeruli, no evidence of hypercellularity, inflammation, necrosis and/or thrombosis was visualized. Severe chronic interstitial inflammation accompanied by fibrosis was also observed. Foci of tubular thyroidization were also identified (figure.1A, B). Interlobular arteries revealed a constellation of findings including fibrinoid necrosis, thrombosis with partial to complete luminal closure, onion skin appearance and vascular wall edema (figure.2A, B &C). Immunofluorescence study revealed positive immunoreactivity for IgM and C3 in mesangial area. The morphologic diagnosis was thrombotic microangiopathy. Further history taking revealed two episodes of miscarriage (after six months of pregnancy). Laboratory evaluations revealed a positive anti-cardiolipin antibody (IgG, >40 U/mL). Other autoantibodies consisting of p-ANCA, c-ANCA, anti-nuclear antibody, anti-ds-DNA were negative. Serum complement levels were also within normal limits. According to morphological, clinical and laboratory data, diagnosis of APS nephropathy was made. Anti-cardiolipin antibody (IgG) was retested after 14 weeks and continued to be positive, confirming catastrophic variant of the APSN. Patient was treated with intravenous heparin followed by oral Warfarin and also other supportive treatment, while hemodialysis and corticosteroids were continued due to persistent renal failure. On further follow up, the patient remained dependent on dialysis in her province.

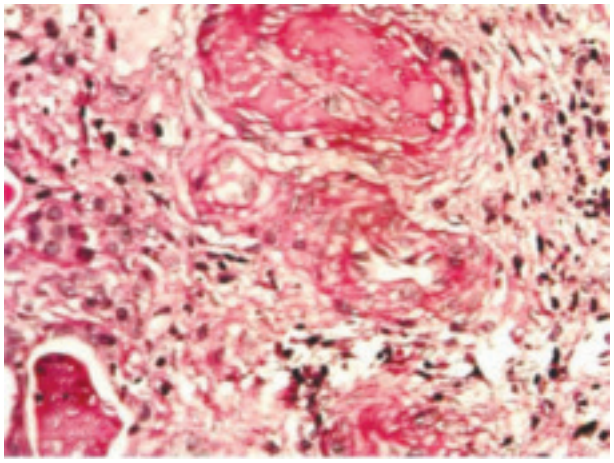


A



B

Figure 1. A: Sever interstitial inflammation and fibrosis .B: tubular thyroidization (H&E ×400).



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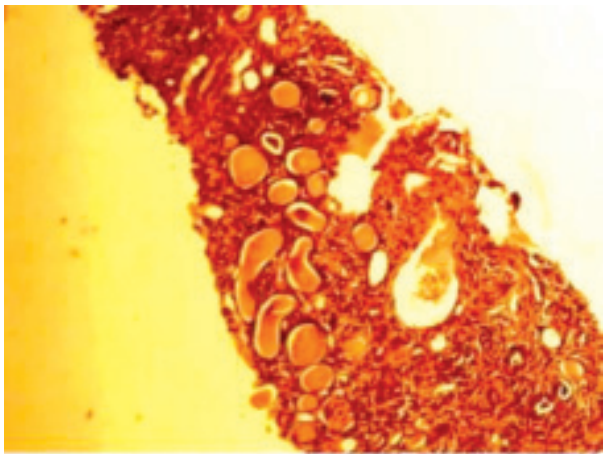

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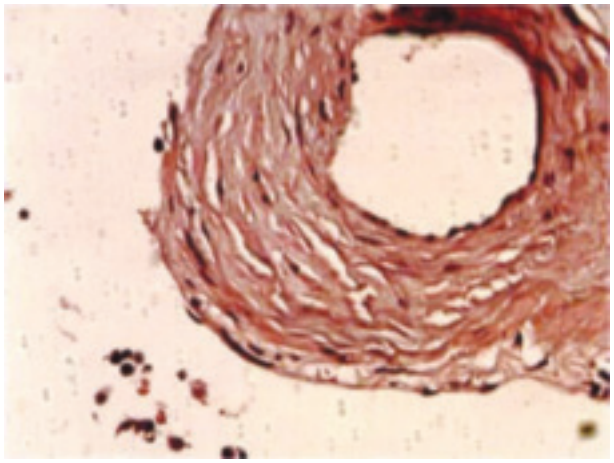

C

Figure 2 (A, B& C). A:Fibrinoid necrosis and thrombosis of interlobular artery. B: Vascular wall edema. C: Fibrous intimal hyperplasia of an interlobular artery, made aspect of onion skin pattern (H&E and PAS ×400).

3. Discussion

APS is as an autoimmune disorder characterized by recurrent thrombosis and/or obstetrical morbidity (1-3). CAPS is the most severe form of APS with acute multiple organ involvement and small vessel thrombosis (4-9). CAPS is an accelerated variant of APS, resulting in multi-organ failure due to extensive thrombosis formation (2-4).The diagnosis of this syndrome requires the presence of clinical evidence of involvement of three or more organs in a period of less than a week, histopathological evidence of small vessel occlusion in at least one organ, and finally laboratory confirmation of the presence of antiphospholipid antibodies (APAs), usually in high titer (2-6). CAPS represent less than 1% of all patients with APS. However, they are usually in a life-threatening situation and its potentially lethal outcome emphasizes its importance in clinical medicine today. The rarity of this variant of APSN makes it extraordinarily difficult to study in any systematic way (4-9). Early recognition of APS is crucial, while aggressive management can result in a favorable outcome (2-5). The recognized renal manifestations of this syndrome defined as APS nephropathy, are the renal artery thrombosis/stenosis, renal vein thrombosis, renal infarction, end-stage renal disease, increased allograft vascular thrombosis, some types of glomerular disease, hypertension and thrombotic microangiopathy (TMA) (1,2,10-12). TMA is characterized by acute thrombotic lesions in glomeruli and/or arterioles and chronic vascular lesions such as fibrous intimal hyperplasia of arterioles and interlobular arteries, organized thrombi with or without recanalization, and fibrous arterial and arteriolar occlusions or focal cortical atrophy (3-9). TMA is histological diagnosis and clinical differential diagnosis includes malignant hyper-

tension, hemolytic uremic syndrome, thrombotic thrombocytopenic purpura, disseminated intravascular coagulation, scleroderma, toxemia of pregnancy and APS. The final diagnosis of APS can be made only if patient have TMA plus clinical and laboratory data (anti-cardiolipin antibody). The most frequent clinical and laboratory characteristics of APSN are hypertension (often severe), proteinuria (ranging from mild to nephrotic range), hematuria, and acute or chronic renal insufficiency (12-14).

4. Conclusions

The CAPS is a very rare complication encountered in a subset of patients with APS. This rare syndrome is defined by the development of multiple blood clots that block small blood vessels in several organs in the body. One of the organs most commonly affected by these small blood clots includes kidneys so-called APS nephropathy.

Author's contributions

SM performed renal biopsy and handled the patient and also wrote some parts of the manuscript. HN reviewed the patient's biopsy and prepared the final draft. All authors approved the final manuscript.

Conflict of interest

The author declared no competing interests.

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