Etiological Diagnosis of BUDD-CHIARI Syndrome in Internal Medicine

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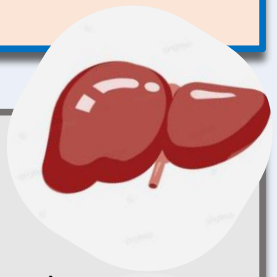
Introduction:

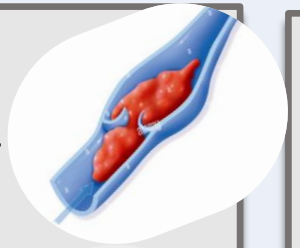
BUDD-CHIARI Syndrome (BCS) is a rare condition in young people that results from obstruction of hepatic venous drainage, from the suprahepatic veins to the terminal part of the inferior vena cava. The objective of our study was to report the epidemiological, clinical, and etiological profile of this condition.



Methods:

This is a retrospective, single-center, descriptive study collecting patients with BCS hospitalized in an internal medicine department over 12 years (2009-2022). All cases were confirmed by abdominal CT angiography.

Results:



There are 7 patients with a sex ratio (M/F) of 1/6.

The average age was 45 years [25-76].

Clinically, all patients were symptomatic (n=7).

The symptoms consisted of signs of portal hypertension in 7 cases, abdominal pain in 5 cases, and fever in three.

In 5 cases, BCS was complicated by cirrhosis.

The BCS was primary in all cases.

The thrombophilia assessment was carried out in all patients, returning normal in 4 cases and revealing a deficiency in protein C, protein S, and antithrombin III in one case.

It revealed a Factor V Leiden mutation in one case and hyperhomocysteinemia in one case.

Buerger's disease, being an exceptional etiology during SBC, was noted in one case.

The search for jack2 mutation was negative in 4 patients in whom myeloproliferative syndrome was suspected. Testing for antiphospholipid antibodies and antinuclear antibodies was negative in all patients and no patient had diagnostic criteria for Behçet's disease. Anticoagulant treatment was started in all patients.

Conclusion

In our series, the epidemiological and clinical findings were similar to those described in the literature. Several etiologies are at the origin of BCS, however idiopathic forms were also reported. Cirrhosis is a frequent complication of this condition and remains the most feared.