

Trombosis venosa esplácnica y de venas cerebrales

Splanchnic vein thrombosis and cerebral vein thrombosis

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Venous thrombosis occurring in unusual sites, which may virtually affect any venous segment, is often challenging. Given the large variety of clinical conditions, this presentation will focus on two of the most relevant sites of venous thromboembolism: the splanchnic and the cerebral veins.

Splanchnic vein thrombosis encompasses portal vein thrombosis, mesenteric vein thrombosis, splenic vein thrombosis and the Budd-Chiari syndrome. Portal vein thrombosis is the most frequent manifestation of splanchnic vein thrombosis, whereas the Budd Chiari syndrome is the least common disease, with incidence rates of about 2.0 cases per million inhabitants⁽¹⁾. The incidence of objectively

diagnosed splanchnic vein thrombosis is possibly increasing due to the large use of routine imaging tests in some patient populations⁽²⁾.

The clinical presentation of splanchnic vein thrombosis can be quite heterogeneous. On the one hand, a non-negligible proportion of thrombi are detected incidentally, in particular in patients undergoing regular staging of solid cancer or liver cirrhosis. On the other hand, splanchnic vein thrombosis can present with acute abdomen or massive gastrointestinal bleeding requiring urgent invasive procedures⁽³⁾. Abdominal pain is the most commonly reported symptom, other symptoms include nausea and anorexia. Signs that may suggest the presence of splanchnic vein thrombosis include increased ab-

dominal girth (due to sudden onset or worsening of ascites), jaundice and fever⁽³⁾. Ultrasound Doppler is the imaging of choice in particular for the diagnosis of portal vein thrombosis, whereas CT angiography remains the standard diagnostic imaging test for the diagnosis of mesenteric vein thrombosis⁽⁴⁾.

Solid abdominal cancer and liver cirrhosis are responsible of up to 50% of all splanchnic vein thrombosis, the remaining are associated with JAK2 V617F mutation with or without overt myeloproliferative neoplasms, inflammatory diseases or infections, abdominal surgery or trauma. Other very rare predisposing factors include paroxysmal nocturnal haemoglobinuria or Behcet disease, to name a few⁽³⁾. Patients who develop splanchnic vein thrombosis have a considerable risk of short and long-term consequences. These include bleeding and recurrent thrombosis, but also liver cirrhosis in patients with Budd Chiari syndrome and portal hypertension in patients with Budd Chiari syndrome and portal vein thrombosis. The risk of bleeding is driven by the use of thrombolytic or antithrombotic drugs, by the presence of underlying risk factors such as gastrointestinal varices or low platelet count, but also by increased portal venous pressure induced by residual venous obstruction^(5,6). Information on bleeding rates is hampered by the lack of a standardized definition of major bleeding events in this setting. Establishing the optimal treatment for splanchnic vein thrombosis patients is therefore challenging, due to severity of presentation in a non-negligible proportion of cases (25% present with gastrointestinal bleeding, others with acute abdomen requiring surgery), the difficult balance between the high risk of recurrent thrombosis and the short and long-term risk of hemorrhagic complications. The identification of underlying provoking factors is crucial to define the prognosis of splanchnic vein thrombosis and to drive appropriate treatment strategies. Additional prognostic factors to be taken into account include the site and extension of splanchnic vein thrombosis and the severity of the clinical presentation. Observational studies suggest that up to one third of patients with portal vein thrombosis are left untreated in routine clinical practice, whereas low molecular weight heparin (LMWH) is the most commonly prescribed treatment. The long-term risk of venous thromboembolism recurrence is highest in patients with cirrhosis, followed by patients with cancer and myeloprolif-

erative neoplasms. Major bleeding complications are also highest in patients with cirrhosis⁽⁶⁾. LMWH is recommended as a standalone treatment during the first weeks for patients with cirrhosis, cancer, or thrombocytopenia. For all other patients LMWH and vitamin K antagonists represent the treatment of choice⁽³⁾. No studies have assessed the efficacy and safety of the direct oral anticoagulants in this setting, but a number of case series have been recently published.

Cerebral vein thrombosis has an estimated annual incidence of 3-4 cases per million adults and most commonly affects young women with gender related risk factors⁽⁷⁾. It represents a rare type of cerebrovascular disease accounting for about 0.5% of all strokes. In adult patients, the mean age at onset is about 40 years, and the ratio between women and men is approximately 3 to 1. The use of oral contraceptives is significantly associated with cerebral vein thrombosis, and during pregnancy the risk of cerebral vein thrombosis appears to be particularly increased during the last trimester and after delivery. Other risk factors include brain tumors, local infections (otitis, mastoiditis, meningitis, in particular in children), head injury, or surgery⁽⁸⁾.

Cerebral vein thrombosis has a wide spectrum of signs and symptoms, which may evolve suddenly or over the weeks. The most common clinical presentations include headache, which is reported in up to 90% of patients, seizures, focal neurological deficits, altered consciousness and papilloedema. According to the grouping of symptoms and signs, four main patterns have been identified: isolated intracranial hypertension, focal syndrome, cavernous sinus syndrome and subacute encephalopathy. These different clinical pictures depend on the site and the extension of the thrombus, the more severe clinical pictures are usually associated with occlusion of the deep cerebral venous system and with the presence of parenchymal lesions⁽⁸⁾. Of interest, up to 40% of cerebral vein thrombosis patients have radiologic signs of intracranial haemorrhage at the time of diagnosis and 10% develop a new haemorrhage during hospitalization. The first line of investigation in patients with suspected cerebral vein thrombosis is CT angiography, although MR angiography remains the gold standard for the diagnosis⁽⁸⁾.

The clinical outcome of cerebral vein thrombosis

appears to be more favourable than with thrombosis of the cerebral arteries in terms of mortality or disability. Factors associated with death or disability were related to the presence of intracerebral bleeding at the time of diagnosis, to the clinical presentation with a coma, seizures, or mental status disorders; to older age or male sex; or to underlying disorders such as cerebral nervous system infection or cancer⁽⁹⁾. Also the estimated recurrence rate of cerebral vein thrombosis was found to be lower than in other sites of venous thromboembolism, in particular in young women with gender related risk factors^(10,11). Anticoagulant treatment has a crucial role with the aim to avoid thrombus extension and to favor local resolution. There are only two small randomized controlled trials that evaluated the efficacy and safety of heparin, either unfractionated heparin or LMWH for the acute treatment of cerebral vein thrombosis⁽¹²⁾. Taken together, these studies show a trend towards reduced risk of death or dependence as compared with placebo. A challenging aspect of cerebral vein thrombosis is related to the concomitant presence of intracranial bleeding at the time of diagnosis. Based on observational evidence, guidelines are concordant that this should not be an absolute contraindication to anticoagulant treatment. Thrombolytic therapy should be considered only for patients with clinical deterioration despite adequate anticoagulant treatment and without any alternative cause of deterioration. The risk of recurrent thrombosis after cerebral vein thrombosis was reported to be low, in particular in young women with gender specific risk factors. Risk factors for thrombosis recurrence include male sex, severe thrombophilia, and previous venous thromboembolism. Studies assessing the role of the direct oral anticoagulants in this setting are ongoing.

Declaración de conflictos de interés:

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