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Behçet's Disease Revealed by Budd-Chiari Syndrome, Neuro-Behçet's and Pulmonary Embolism

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Abstract **Case Report**

Behçet's disease is a vasculitis of the vessels affecting the arterial and venous territories whatever their calibers. It preferentially affects young subjects between 10 and 45 years old and also affects men and women. The history and physical examination are key steps in the diagnosis. We report the case of a 37-year-old patient with a history of repeated oral aphtosis for several months, chronic active smoker and former chronic alcoholic, in whom the revelation of Behçet's disease by the Budd-chiari syndrome, neuro-behçet and pulmonary embolism in the complication phase, this despite the multidisciplinary intervention, the vital prognosis was put into play. The revelation of Behçet's disease by multi-visceral complications justifies the diagnostic and management complexity, hence the importance of the contribution of imaging. These patients require early diagnosis of this polymorphic pathology and regular monitoring. Its manifestation by the Budd-Chiari syndrome is its seriousness due to its complications. Essentially all forms of liver disease are possible: fulminant, acute, subacute, chronic or an inaugural digestive hemorrhage by portal hypertension. The diagnostic delay in the complications phase, with multi-visceral involvement associating Budd-chiari syndrome, neuro-behçet and pulmonary embolism are the consequence of life-threatening.

Keywords: Behçet's disease, Diagnosis, polymorphic pathology, portal hypertension.

INTRODUCTION

Behçt's disease is a vasculitis of vessels of all calibers, affecting the arterial and venous territories. It preferentially affects subjects of young age, most often between 10 and 45 years old, and affects both men and women. A first flare after age 50 is rare. Behçet's disease is ubiquitous but more common in patients from the Mediterranean basin, the Middle East and Asia.(2) Behçet's disease being a protean disease, the history and physical examination are key steps in the diagnosis and must be exhaustive. Diagnosis and management are all the more difficult when the disease is revealed by several complications. Due to the extreme polymorphism of the disease, any doctor can be confronted with early Behçet's disease [1].

Budd-chiari syndrome results from obstruction of hepatic venous drainage from the hepatic venules to the terminal part of the inferior vena cava, regardless of the cause of the obstruction; it is primary when caused by thrombosis or its fibrous sequelae, secondary when the vein is invaded or compressed by a tumoral or parasitic obstruction [2].

Despite the progress of therapeutic means on the management of Behçet's disease, the association of Budd-chiari syndrome, neuro-Behçet's disease and pulmonary embolism is of potential severity and poor prognosis. These patients require special attention and multidisciplinary management because the prognosis is often compromised.

PATIENT AND OBSERVATION

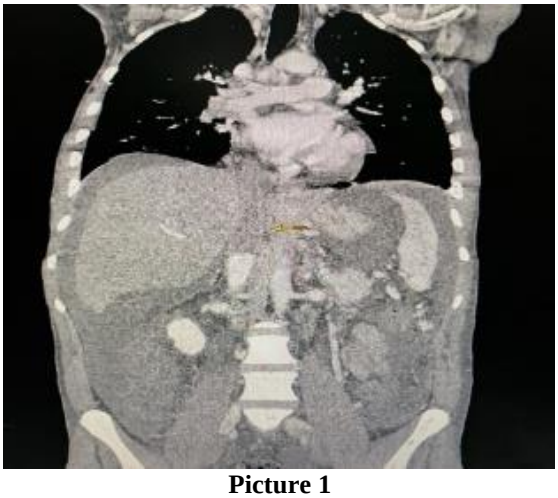
This is Mr A.J., 37 years old, with a history of repeated oral aphtosis for several months, chronic active smoker and former chronic alcoholic not quantified weaned for 5 years, who has presented for five months of intermittent abdominal pain beginning in the right hypochondrium, of medium intensity, then becoming progressive and diffuse. This symptomatology was associated with abdominal distension, moderate diarrhea made of three to four stools / day without vomiting, all evolving in a context of apyrexia, deterioration of general condition (anorexia, asthenia, weight loss 19 kg) and lower limb edema.

On clinical examination, the patient was conscious, presented with conjunctival subicteritis,

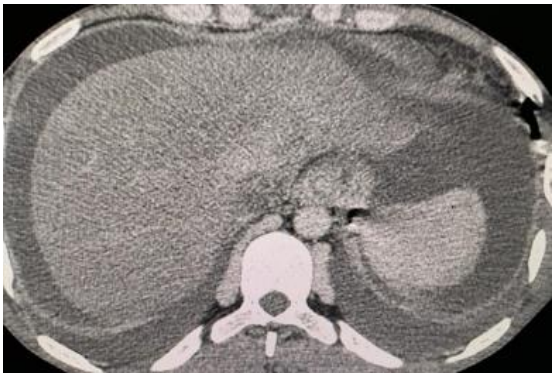
hemodynamically and respiratory stable (blood pressure 10/6 CmHg, heart rate 87 beats/minute, respiratory rate 18 cycles/minute), impaired general condition classified as WHO II, fever at 37.8°C, distended abdomen with edema-ascites syndrome (edema of the lower limbs and abundant ascites), collateral venous circulation, abdominal tenderness, mouth and genital ulcers. Crackles in both lung fields and the rest of the physical examination were normal.

Biologically, there is hepatocellular insufficiency (prothrombin level 35% INR 2.13, hypo-albuminemia at 23 g/l), an inflammatory syndrome with the C-reactive protein CRP at 75mg/l, impaired liver function with mild cytolysis alanine amino-transferase at 71 IU/L, aspartate amino-transferase (ASAT) at 119 IU/L, cholestasis alkaline phosphatase (ALP) at 309 IU/L, gamma glutamyl transferase (ϒGT) at 168 IU/L Total bilirubin 27.6 mg/L and conjugated bilirubin at 21.8mg/L), renal balance normal, biochemistry of ascitic fluid finds low protein ascites (transudate) at 12g/l and sterile culture. The complete blood count shows a slight normochromic microcytic anemia (hemoglobin at 12 g/dl, mean corpuscular volume at 70.7 fl, mean corpuscular concentration of hemoglobin at 34.5 g/dl) normal leukocytes at 6000/mm3, thrombocytopenia (platelets at 117,000/mm3). Vitamin B12 was twice the upper limit, hypokalaemia at 3.2 mmol/L, viral serology for hepatitis B and C was negative, blood culture and cytobacteriological examination of urine were sterile.

On imaging, the chest X-ray found a small pleural effusion at the right lung base. Abdominal ultrasound revealed thrombosis of the hepatic veins, incomplete obstruction of the inferior vena cava and abundant ascites. Then we completed with a thoraco-abdomino-pelvic scanner showing thrombosis of the inferior vena cava (IVC) and suprahepatic veins (VSH), bilateral massive proximal pulmonary embolism, bilateral pleural effusion and pericardial low abundance and abundant peritoneal effusion, Picture 1, 2 and 3.



Picture 1



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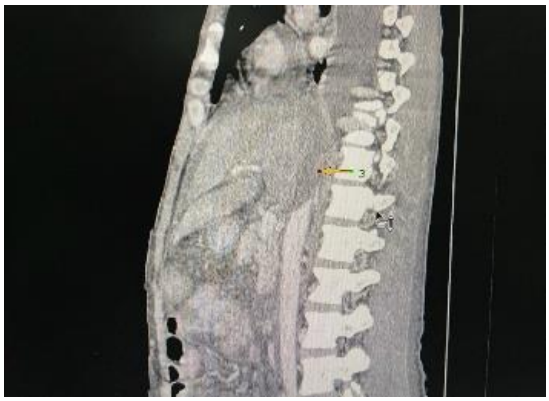


Picture 3

An ophthalmological examination carried out found neither uveitis nor retinal vascular anomaly compatible with Behçet's disease (normal fundus).

The esophagogastroduodenal fibroscopy performed found 2 stage I esophageal varices.

Then we complete the biological assessment by protein C and S tests without abnormality, anti-phospholipid IgM antibodies 1.70 UMPL/ml (negative); anti-phospholipid IgG antibodies 2.90 UMPL/ml (negative); the factor V mutation Leiden p. Arg 506Gln (real-time PCR) with absence of mutation, the search for G20210A mutation of the prothrombin gene (real-time PCR) notes an absence of mutation, the search and quantification of the V617F JAK2 mutation notes the undetected mutation. Finally, the search for BK and the covid19 PCR were negative.

In the presence of these clinical, biological and imaging arguments, the diagnosis of Budd-Chiari syndrome complicated by Behçet's disease was made.

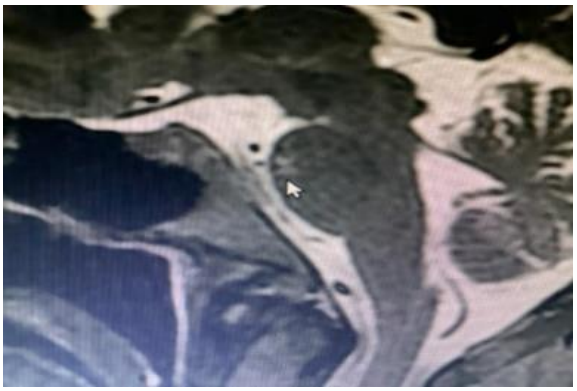
Faced with this picture, the patient is put on anticoagulant treatment based on LMWH 100 IU (1mg)/kg/12h, potassium syrup, corticosteroids, azathioprine, then spironolactone on the 5th day. The evolution was good with the improvement of clinical signs (pain, disappearance of oedemas, a decrease in ascites) during the first week.

On the 11th day, he presented with a sudden drop in weight of 2 kg in 24 hours associated with a diuresis of 2.5 liters, dyspnea, agitation, psychomotor retardation, moderate headaches and a stato-kinetic cerebellar syndrome (hypotonia, ataxia, trouble standing and walking).

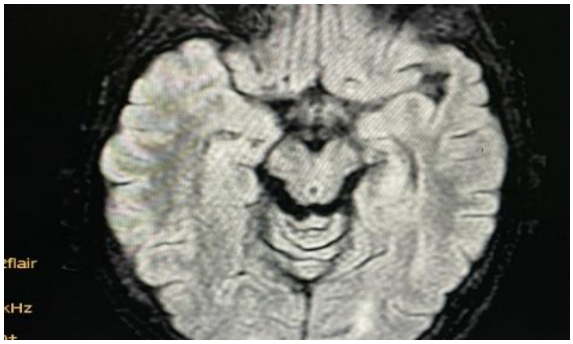
The biological check-up including the ionogram and blood count without abnormality, hypo-albuminemia at 19 g/l, worsening of cholestasis (gamma glutamyl transferase at 242 IU/L, alkaline phosphatase at 330 IU/ L, total bilirubin at 33.3 mg/L, conjugated bilirubin at 25.6 mg/L), cytolysis little modified alanine amino-transferase at 90 IU/L, aspartate amino-transferase at 87 IU/L, the inflammatory assessment is improved with CRP at 23.05 mg/L, normal renal function, hypoproteinemia at 57g/L, prothrombin level TP=31%.

A cerebral angio-MRI objectified the ponto-mesencephalic lesions falling within the framework of a neuro-behçet and the active right anterior pansinusitis Figure 4 and 5.

The diagnosis of Budd-Chiari syndrome associated with ponto-mesencephalic lesions and a bilateral massive proximal pulmonary embolism complicated by Behçet's disease is retained and the patient was referred to the resuscitation and intensive care unit after a neurological and pneumological opinion where he died on the tenth day of intensive care in a confusional picture.



Picture 4



Picture 5

DISCUSSION

We report a case of revelation of Behçet's disease from its complications of Budd-chiari syndrome, neuro-Behçet's syndrome and pulmonary embolism in a 37-year-old young man, this is a rare situation in the literature. According to Dominique-Charles VALLA “Budd-chiari syndrome is a rare disease in young adults” [5] and remains an infrequent complication in Behçet's disease. In developed countries, its incidence has been estimated at no more than 1 case per million per year, and its prevalence at 2.5 per million inhabitants [5]. However, in very disadvantaged populations, it has been the main reason for admission for liver disease [5]. Budd-Chiari syndrome is asymptomatic in 15 to 20% of cases and discovered incidentally [8]. In other cases, all forms of liver disease are possible: fulminant (exceptional), acute, subacute or chronic. The clinical manifestations are abdominal pain, ascites often, but not always, rich in protein unlike in the case of our cohort where the ascites was poor in protein, hepato-splenomegaly which was absent in our patient. According to the literature, jaundice or inaugural gastrointestinal bleeding due to portal hypertension are rare.

The diagnosis of Behçet's disease is essentially clinical, due to the absence of specific biological criteria. The average age at diagnosis is around 30 years old and the vast majority of diagnoses are made between 15 and 45 years old. Conversely, it is exceptional to make a new diagnosis before the age of 15 and after the age of 50 [1].

In our cohort, the transaminases are moderately elevated, less than twice the upper limit of normal, comparable to those described in the literature, they are discreetly elevated or even normal, on the other hand they can be very elevated in the acute forms then returning quickly. to near normal values in Budd-Chiari syndrome. Cholestasis was also noted, including elevated alkaline phosphatase and gammaglutamyl-transferase with moderately elevated bilirubin justifying our patient's subicterus. Hepatocellular insufficiency was well marked in our cohort with a very low prothrombin level, severe hypoalbuminemia and mild thrombocytopenia. For Dominique-Charles VALLA, the moderate increase in bilirubinemia is usual. Hypo-albuminemia, a decrease in the Quick rate <50% are observed in severe forms. Moderate functional renal insufficiency is common in forms with ascites [5]. On the other hand, renal function was normal in our case.

According to the literature, there is no relationship between the progressive form (fulminant, acute, subacute or chronic) and the age of the venous or hepatic lesions [5, 9].

In view of the clinical criteria, the diagnosis of Budd-Chiari syndrome was confirmed by Doppler

ultrasound and angio-scanner in our cohort with the presence of thrombosis of the supra-hepatic veins and the inferior vena cava complicated with portal hypertension. In the absence of the most frequent causes, the cause of the Budd-Chiari syndrome retained in our cohort is Behçet's disease by the presence of bipolar aphtosis and the presence of pulmonary and ponto-cerebral lesions on MRI- brain in favor of Behçet's disease. Budd-chiari syndrome in Behçet's disease is rare but remains a severe complication with a poor prognosis [4]. In the literature, Behçet's disease represents 1-5% of the etiologies of the primitive Budd-Chiari syndrome, on the other hand several etiologies are identified in particular the primitive myelo-proliferative syndrome present in 50% of the patients; factor V Leiden 25%; the G20210A mutation of the prothrombin gene in 5-10%; paroxysmal nocturnal hemoglobinuria in 1-5% [5, 6]. The diagnostic delay remains greater than one month for 50% of patients. Its manifestations are variable and range from the absence of symptoms to fulminant hepatic failure or cirrhosis [3]. In a patient already followed for thrombogenic disease, the diagnosis remains easier. However, the inaugural Behçet's disease with multi-visceral involvement (hepatic, cerebral, pulmonary) is responsible for diagnostic difficulties, as in the case of our cohort.

The so-called “angio-Behçet” or “vasculo-Behçet” vascular involvement is particular in certain aspects because it is mainly observed in young men without thrombotic or cardiovascular risk factors [7]. Which is comparable to the case of our patient. All vessels, whatever their type (arterial or venous), their caliber or their location, can be affected, often with multifocal vascular manifestations [7].

Neurological manifestations occur in approximately 25-30% of patients. Parenchymal damage is frequent (70-80% of patients with neurological damage) than extra-parenchymal damage. The coexistence of the two forms is exceptional in the same patient. The particularity of these attacks is that they can be inaugural. The existence of damage to the peripheral nervous system has rarely been described, but its real link with Behçet's disease remains debated [2].

Pulmonary involvement is rare and essentially consists of nodular consolidation lesions post pulmonary infarction which subsequently tend to excavate. One can sometimes observe ground glass infiltrates reflecting intra-alveolar hemorrhage. In a few cases, vasculitis has been demonstrated. CT angiography and ventilation/perfusion scintigraphy can detect them. Vasculitis mainly results in aneurysms of the pulmonary arteries, the rupture of which can be fatal. Parenchymal lesions such as nodules, pleural effusions, or mediastinal lymphadenopathy are also part of pulmonary Behçet's disease. It is necessary to eliminate an infection in this context [1]. Vascular damage, particularly arterial, is serious and remains the main cause of death in patients with Behçet's disease [1]. Specialized multidisciplinary care in an expert center is necessary for this rare disease with very polymorphic expression and requiring treatment and prolonged follow-up.

Drug treatments for Behçet's disease essentially depend on the clinical manifestations. More serious ocular, vascular or neurological damage requires immunomodulatory treatment, usually combining systemic corticosteroid therapy with immunosuppressants or biotherapy (anti-TNFα) as indicated. The place of anticoagulant treatment in the management of venous vascular damage is debated but remains recommended, the thrombotic mechanism being essentially an inflammation of the vascular wall. The prescription of aspirin at an antiaggregating dose is considered in stenosing arterial disorders [1]. The therapeutic management of Budd-Chiari syndrome associated with Behçet's disease is based on anticoagulation, corticosteroid therapy, cyclophosphamide or azathioprine [4].

Despite the multidisciplinary management between hepato-gastroenterologists, internists, radiologists, resuscitators, neurologists and pulmonologists as in our cohort, the diagnostic delay with the presence of several complications leads to a poor prognosis in these cases. A. Allaoui1 et al., also reported cases of death during their study, including four out of fifteen cases of death [4].

CONCLUSION

The diagnosis of Behçet's disease is clinical, but the disease can be revealed by its complications, hence the importance of the contribution of imaging. The prognosis of this rare disease varies from one subject to another, sometimes potentially serious. The revelation of Behçet's disease by multi-visceral complications justifies the diagnostic and management complexity. These patients require specialized multidisciplinary care in an expert center for this rare disease with very polymorphic expression as soon as it is diagnosed.

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