CytoSorb®

Case of the week 10/2021

Use of CytoSorb in a patient with therapy-resistant pruritus due to primary biliary cholangitis

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This case reports on a 36-year-old female patient (relevant previous diseases: hereditary sensorimotor polyneuropathy with initial diagnosis in the '90s), who presented for a liver consultation.

Case presentation

- She presented for the first time in 2011 with elevated transaminases, positive antimitochondrial antibody titer and a cholestatic pattern (□GT/AP). Acute and chronic viral hepatitis infection (A, B, C, E), hemochromatosis, Wilson's disease and □1-antitrypsin deficiency were ruled out
- In 2013, she underwent a biopsy suggestive of low grade and low florid portal hepatitis and signs of cholangitis with slowly increasing fibrosis without signs of liver remodeling, resulting in the diagnosis of primary biliary cholangitis (PBC) and subsequent initiation of ursodeoxycholic acid therapy with significant improvement over time
- In 2015, and despite the good response to therapy, transaminases again increased resulting in suspected autoimmune hepatitis overlap syndrome with a positive antinuclear antibody titer. Without performing a new biopsy, immunosuppressive therapy with budenoside and azathioprine was started. This was followed by a further deterioration in the clinical situation with bilirubin levels being elevated for the first time
- In 2017, immunosuppressive therapy with budenoside was stopped and therapy with azathioprine continued. In addition, relapse therapy with 100 mg prednisolone was initiated, however, this had no apparent effect on the level of transaminases
- The leading clinical symptom was massive pruritus (score 10/10 on the visual pruritus scale) associated with fatigue, exhaustion, depression and suicidal thoughts. In addition, from 2013 to 2017 there was an unintentional weight loss of 13 kg
- Laboratory diagnostics at the time of presentation to the Charite Berlin confirmed strongly positive anti-M2, anti-gp210 and ANA titers. The anti-sp100 titer was negative. In addition, there were clearly elevated IgM titers with a normal IgG titer as well as clearly elevated bile acids (>140 µmol/L). Biopsy showed no evidence of autoimmune hepatitis, but high primary biliary cholangitis activity and advanced fibrosis without cirrhosis. The clinical picture of the skin showed excoriations and severe prurigo nodules
- Administration of colestyramine, naltrexin, sertraline, bezafibrate, obeticholic acid, rifampicin as well as phototherapy did not lead to a satisfactory therapeutic outcome
- As a therapeutic option for the treatment of cholestatic pruritus, plasmapheresis with the Plasorba BR-350 adsorber (Asahi Kasei Medical) under citrate anticoagulation was then initiated (therapy of 2.5 times plasma volume), resulting initially in a slight reduction in bile acid plasma concentrations, while the visual pruritus scale (VPS) remained unchanged
- Due to the relatively moderate treatment effect of plasmapheresis, CytoSorb hemoadsorption was initiated in standalone mode

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Treatment

- Several CytoSorb treatments were performed for the next days (each treatment between 5-6 hours)
- CytoSorb was used in hemoperfusion-only mode
- Treated blood volume per treatment: 60-70 litres
- Anticoagulation: citrate

Measurements

- Bilirubin
- Bile acids
- Subjective itching sensation (visual pruritus scale)

Results

- During treatment, bilirubin levels were kept stable and at a low level
- Treatment with CytoSorb also resulted in a rapid reduction in plasma bile acid concentrations and stabilization at non-toxic levels (<25 µmol/L)
- Furthermore, treatment was associated with a significant reduction in subjective itch sensation (visual pruritus scale lowered to 5)

Patient Follow-Up

- Over time there was excoriations started to heal
- According to the patient, the quality of sleep improved significantly with the use of a blanket at night now possible
- Three months after therapy discontinuation, the patient again complained of severe pruritus (visual pruritus scale 10/10), marked excoriation and massive sleep disturbances
- At the time of documentation, an application to the liver transplantation list had not yet been made, and the restart of CytoSorb therapy was not possible due to the patient's personal situation

Conclusions

- In this patient with therapy-resistant pruritus due to primary biliary cholangitis, treatment with CytoSorb hemoadsorption resulted in the reduction of plasma bile acid concentrations as well as a clear improvement in the subjective itching sensation
- According to the medical team, CytoSorb appears to interfere with the suspected pathophysiology of PBC-associated pruritus, which is centred around cytokines, damage-associated molecular patterns (DAMPs), pathogen-associated molecular patterns (PAMPs) and, most importantly, cholestatic markers such as bile acids - thus contributing to a significant improvement in the clinical condition as well as the quality of life for patients
- CytoSorb was safe and easy to use in standalone mode