At the age of 69 years, a woman with recurrent gastrointestinal bleeding underwent investigation by gastroscopy and colonoscopy. Extensive telangiectatic changes were observed in the canalis and antrum of the ventricle and in the colon. In parallel, the woman suffered from nephrotic syndrome, hypertension, and progressive renal failure attributable to chronic glomerular changes with extensive interstitial fibrosis. The progressive renal failure and recurrent gastrointestinal bleeds made frequent transfusions and erythropoietin injections necessary. Because of those complications, and because CAPD avoids the intermittent overhydration of a therapy such as hemodialysis and the risk of using anticoagulants, it was decided to perform continuous ambulatory peritoneal dialysis (CAPD) when dialysis became necessary.

After CAPD was started, the woman’s bleeding episodes decreased within 1 week. After 70 months of CAPD, the woman is well, without gastrointestinal bleeding. No transfusions have been necessary since the start of CAPD. For the now 77-year-old woman, data are as follows: epoetin beta dose, 12,000 U/week; body weight, 67 kg; hemoglobin, 130 g/L. The woman has experienced 1 episode of peritonitis since starting dialysis (6 months before the time of writing), because of a malfunction of the peritoneal dialysis device. No intestinal perforations have occurred.

It seems worthwhile to try CAPD for dialysis in patients with morbus Osler.

Key words
Morbus Osler, Osler–Weber–Rendu syndrome, uremia, telangiectasias, hemorrhagic

Introduction
Morbus Osler is also called Osler syndrome, Osler–Weber–Rendu syndrome, and hereditary hemorrhagic telangiectasias. The prevalence of the disease is approximately 1 – 10 per 100,000 population. The condition is caused by an autosomal-dominant mutation of either the endoglin gene on chromosome 9 (ENG) or the gene for the activin receptor–like kinase (ACVRL1) on chromosome 12 (1,2). Homozygous fetuses probably do not survive birth. The ACVRL1 gene mutation probably causes a lesser degree of clinical manifestation.

These gene mutations cause a disturbance of protein synthesis of the endothelium. As a result of that disturbance, the arteriolae and the capillaries do not develop in a normal way, and arteriovenous fistulae occur. The fistulae can develop in all tissues, but are more common in the mucosa of the mouth and nose. Frequent findings are also made in lung, cerebrum, and the gastrointestinal tract. The diagnosis commonly rests on clinical findings; its classification is helped by DNA tests.

Clinical symptoms—mainly nose bleeding (90% of patients)—begin to develop during or after puberty. Later in life, more extensive lesions of the skin and mucous membranes develop; after the age of 50 years, other bleeding sites become common. Patients having the chromosome 9 defect are more prone to bleeding from the lungs and to cerebral hemorrhages; those with chromosome 12 mutations more commonly suffer from gastrointestinal bleeding; renal involvement with hematuria is another finding.

Treatment is symptomatic, with avoidance of laceration of vulnerable aneurysms. Laser coagulation can only partly prevent bleeding (from intestinal sources, for example). In women, estradiol is sometimes used as an endocrinologic measure to “strengthen” the tissue (3,4).

No reports of morbus Osler in combination with dialysis-dependent renal failure can be found in the literature.

Case study
Our unit treats a 77-year-old woman with no hereditary history of bleeding disturbances. Previous personal history includes a thyroidectomy in 1947 for
nontoxic enlargement, curettage in 1952, and, because of suspected local endometriosis, unilateral ovariectomy. Hypertension was noted in 1991. At the same time, hypercholesterolemia was detected, and statin medication was initiated.

During her teen years, this woman started to suffer from recurrent nosebleeds. After a pregnancy in 1946, she suffered from intermittent back pain and hematuria (see “Renal History,” below). At the age of 69 years, the woman began to suffer from recurrent gastrointestinal bleeding and consequent anemia. Gastroscopy and colonoscopy verified extensive telangiectatic changes in the canalis and antrum of the ventricle, and in the ascending and sigmoid colon and the rectum. The bleeding became more and more life-threatening and had to be corrected by repeated blood transfusions. The woman also suffered from microaneurismal changes on the mucosa of her lips and mouth. A diagnosis of morbus Osler was made on clinical grounds.

Renal history

Stable proteinuria was detected during a pregnancy 1946. Thereafter, the woman suffered from recurrent hematuria. A bleeding focus was never localized despite repeated urologic investigations. A renal biopsy in 1995 revealed glomerulosclerosis and interstitial fibrosis. Ultrasound visualized kidney scars. Progressive renal failure was diagnosed. During the stage of severe chronic kidney impairment, the woman received subcutaneous erythropoietin.

When the renal failure reached a life-threatening stage, discussions were held with the patient about various considerations and risks, and an attempt was made to start dialysis. For various reasons (see “Discussion”), continuous ambulatory peritoneal dialysis (CAPD) was initiated. Within 1 week of the patient starting CAPD, the gastrointestinal bleeding ceased. After 70 months of CAPD, the woman is now well, without evident bleeding—including gastrointestinal bleeding. No transfusion has been necessary since the start of CAPD.

Currently, the patient uses epoetin beta 12,000 U weekly. Her body weight is stable at 67 kg, and her hemoglobin is 130 g/L. She no longer suffers from nosebleeds, except very mild ones in conjunction with severe viral respiratory infections.

The woman experienced 1 episode of peritonitis during the CAPD period (6 months prior to the writing of this article), which was considered attributable to a malfunction of the PD connection system (Stay-Safe: Fresenius Medical Care, Bad Homburg, Germany). She currently performs four 2-L exchanges of peritoneal dialysis fluid daily. No intestinal perforations have occurred. Her peritoneal equilibration test data are within the average for urea, creatinine, and glucose.

The patient is very satisfied with the dialysis modality. Her quality of life, estimated on a scale from 0 (worst ever) to 10 (best ever), is 7. She still drives a car and lives in her own home. Her son suffers from nosebleeds, but her daughter has no bleeding disturbances.

Discussion

The literature contains no reports of experiences with this type of patient. Therefore, before any dialysis type was decided, discussion arose among the woman’s physicians about whether any dialysis modality might worsen her general condition and shorten her life expectancy even further.

On the one hand, hemodialysis would cause intermittent water retention and, in principle, a need for anticoagulation that would worsen bleeding problems. In contrast, CAPD would reduce the extent of intermittent overhydration, and anticoagulation would not be necessary. However, worries existed about intra-abdominal aneurysms and the subsequent risk of extensive intra-abdominal bleeding that PD might induce. Intra-abdominal aneurysms could start to bleed, induced either by direct rubbing by the Tenckhoff catheter or by distension of the tissue caused by peritoneal dialysis fluid. Distension of the tissue could increase, especially when ultrafiltration further increased intraperitoneal volume and, concomitantly, intraperitoneal pressure. In addition, intestinal perforations could occur.

For decision-making, we had the benefit of previous experience at our center in using CAPD for patients with severe congestive heart failure (5). In a study of those patients, we noted that continuous removal of fluid (ultrafiltration) effectively reduced the burden on the heart, enabling improvement of heart size in those patients. Despite dicumarol anticoagulation therapy in some of the patients, they did not develop bleeding complications either preoperatively or postoperatively. Because in CAPD (as compared with intermittent hemodialysis) the ultrafiltration of retained fluid is
distributed evenly throughout the week, the risk of intermittent fluid retention and subsequent edema with increased intravascular pressure is reduced. Therefore, the distension of aneurysms could be reduced.

The intraperitoneal volume used would be kept low initially. In contrast to hemodialysis, CAPD would require no further anticoagulation to perform the dialysis procedure per se. The patient was considered able to perform the CAPD training program, and she was also well motivated.

The woman started CAPD from the immediate postoperative period onward, according to our routine program. We start with 1-L exchanges 4 – 8 times during the first few days, slowly increasing the volume and reducing exchange frequency over time. We also use a surgical girdle to avoid too much intra-abdominal distension when the patient is out of bed during the first week (6,7). In parallel, the training program for CAPD is performed. That protocol functioned well in this case.

Peritoneal dialysis has been a very successful mode of dialysis for this patient with morbus Osler. Despite a somewhat-too-low Kt/V (she does not want to increase the number or volume of exchanges), she is happy with her current situation. Her latest peritoneal equilibration test is within the normal range for urea, creatinine, and glucose (Figure 1).

Conclusions

Based on our limited experience of this patient with morbus Osler, CAPD seems to be a promising peritoneal dialysis modality for patients at risk of severe bleeding complications.

References


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