Hemorrhage Because of Amyloid-Related Factor X Deficiency After Insertion of Tenckhoff Catheter

Editor:

Bleeding is a common complication of immune light-chain (AL) amyloidosis and is reported in up to 28% of cases (1). Although intracutaneous bleeding is the most common manifestation, severe bleeding after peritoneal dialysis (PD) catheter insertion has not previously been reported.

We recently electively inserted a Tenckhoff catheter in an 80-year-old man with longstanding immunoglobulin Gκ monoclonal gammopathy complicated by systemic amyloidosis with cardiac and renal involvement leading to stage 5 chronic kidney disease. He had no prior history of major or minor bleeding and was on no antiplatelet or anticoagulation therapy at the time.

A swan-neck coiled Tenckhoff PD catheter was successfully placed deep in the pelvis under laparoscopic guidance. An arcuate subcutaneous tunnel was formed, and the exit site was created in the right lower quadrant. There was no immediate bleeding observed at either the exit site or the port sites, and the patient's immediate postoperative course was uncomplicated.

On postoperative day 2, the patient felt a pulling sensation when bending over, and he soon after noted oozing from the exit site, with subsequent filling of the catheter with blood. The exit site bleeding became more brisk, and the patient was seen in clinic. The bleeding was controlled with pressure on the tunnel and subcutaneous desmopressin. Subcutaneous vitamin K was given for an international normalized ratio of 1.3, and the catheter was flushed until the effluent was clear.

The bleeding recurred after the patient returned home, and he subsequently required admission to hospital because of persistent brisk bleeding from the tunnel. On admission, he was noted to have diffuse abdominal ecchymoses and a palpable hematoma throughout the tunnel tract. In hospital, he was put on bedrest and received further treatment with vitamin K, desmopressin, fresh-frozen plasma, and tranexamic acid. Permanent hemostasis was achieved 2 days after re-admission, and the catheter remained functional.

During this time, the patient's hemoglobin concentration fell to 98 g/L from 124 g/L. In the postoperative period, his platelet count ranged between 116×10^9/L and 120×10^9/L (reference range: 124 – 400×10^9/L); his partial thromboplastin time, between 27 s and 30 s (reference range: 22 – 30 s); and his international normalized ratio, between 1.2 and 1.3 (reference range: 0.9 – 1.2). Factor X measured on postoperative day 3 was low at 0.55 U/mL (reference range: 0.7 – 1.52 U/mL).

DISCUSSION

This case is the first reported of severe bleeding associated with PD catheter placement in a patient with AL amyloid–associated factor X deficiency. It represents an uncommon but potentially severe complication in this population. Earlier large case series have reported rates of major bleeding after PD catheter insertion to be in the 2% range among unselected uremic patients (2). Patients with AL amyloidosis are at increased risk of bleeding because of both blood vessel fragility associated with amyloid infiltration of small vessels and amyloid-associated coagulopathy (3,4). Elevated thromboplastin time and prothrombin time are the coagulation abnormalities most commonly seen. A few patients also have factor X deficiency, thought to occur through selective binding of factor X to amyloid fibrils in the liver and spleen (5,6).

Observational studies of patients with AL amyloidosis report factor X deficiency in 6.3% – 14% of them (6,7). A recent series of 60 patients with mild-to-severe factor X deficiency undergoing invasive procedures or surgeries reported an overall 11% risk of bleeding (6). In many cases, preoperative treatment with platelets, fresh-frozen plasma, and plasmapheresis was given in anticipation of potential bleeding, but the efficacy of those precautionary measures is uncertain.

Our patient presented with delayed bleeding after catheter insertion, and the bleeding appears to have been precipitated by minor trauma to the catheter associated with bending. That observation suggests that the bleeding may well be related both to blood vessel fragility and to the demonstrated factor X deficiency, which presents as impaired secondary hemostasis.

Before inserting PD catheters in patients with underlying AL amyloidosis, nephrologists and surgeons should screen for factor X deficiency. If the deficiency is severe, then preventive measures as outlined earlier should be instituted. These patients should also be cautioned to

avoid physical exertion for a few days post insertion. If bleeding occurs, then medical attention should be sought immediately.

We hope that this case increases awareness of this potential complication of PD catheter insertion in patients with AL amyloidosis.

DISCLOSURES

The authors have no financial conflicts of interest to declare.

G. Harman
B.B. McCormick*

Division of Nephrology
University of Ottawa
The Ottawa Hospital
Ottawa, Ontario, Canada

*email: bmccormick@ottawahospital.on.ca

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