Case Report

Journal of Gastroenterology & Digestive Systems

Crohn Disease in Patients with VW Disease

Hassan Moteirek

Completed my MD in 2003 at the BSMU in Minsk, and there continued my education and training at the gastrointestinal and endoscopy department.

***Corresponding author**

Dr. Hassan Moteirek, Completed my MD in 2003 at the BSMU in Minsk, and there continued my education and training at the gastrointestinal and endoscopy department. At mean time works in south Lebanon, Phone: 009613295276; Email: dr.hassanmt@gmail.com

Submitted: 11 Sep 2018; **Accepted:** 20 Sep 2018; **Published:** 28 Sep 2018

Keywords: Von Will Brand Disease (Vwd), Crohn Disease, Colonoscopy, Lower GI Bleeding, Mesaslazine.

Case History

29 years old male with a history of recurrent rectal bleeding, joint pain and sickle cell disease. He underwent cholecystectomy in 2005 with post operation bleeding complication stopped after blood transfusion. In March 2017 he referred with complains on abdominal pain and recurrent rectal bleeding. Colonoscopy was performed and showed mild ileitis with superficial ulcerations, compatible with crohn disease which was confirmed by pathology. The patient was put on meclizine 4 g daily, and Imuran 50 bids. Symptoms partially resolved. After one year he came for control colonoscopy, which was performed in March 2018. The patient was scoped to terminal ilium where ileitis and ulcerations persisted, biopsy was taken. At the site of biopsy started oozing, hemostasis performed with adrenaline injection which was successful. The patient was admitted to the hospital for observation, after 12 hours started to see fresh blood per rectum. Hematocrit persisted 40, PTT 42 (slightly elevated)... correction test was positive suggesting coagulation disorder. Factor VIII and VWF requested. We started treatment with plasma 2 unit's q 4 hrs. Bleeding stopped then persisted until his hematocrit dropped to 25, we scoped again and put 3 clips at the site of biopsy but bleeding persisted. His hematocrit dropped to 22 and had blood transfusion, then after consultation with hematologist and surgeon we went for right hemi colectomy. Patient was discharged after 2 days. Till this moment we were waiting the results for the factors VIII and VW since it was not available at our hospital. After 24 hours, this patient came back to the ER with fresh blood per rectum and was admitted to start plasma again. On the second day we gat the results for the factors and we had VWF deficiency and secondary factor VIII deficiency. We started giving the factors "WY late" depending on his weight (30 units per kg q 8 hours), and exacyl PO q 8 hours.

After 24 hours bleeding stopped completely and the patient was discharged home and completed the course of VWF treatment and till now he had no other complications.

Discussion

Crohn's disease is a chronic inflammatory disease of the gastrointestinal tract. Crohn's disease is a relapsing and remitting disease. Crohn disease 3.1- 20.2 cases per 100 000. This illness usually appears early in life; about one-sixth of patients present before the age of 15 and often with severe disease. The average age at diagnosis is 27 years. The cause of Crohn's disease is unknown, although strong genetic influences are suggested by the occurrence of this disease in families, with a higher incidence in Jews than in the general population. Genetic influences are more prominent in the younger onset subgroup of patients than those who present after the age of 40. Crohn's disease is associated with extra intestinal manifestations that may be more problematic than the bowel disease. Colitic arthritis is a migratory arthritis that affects knees, ankles, hips, wrists, and elbows that may accompany Crohn's disease (although it is uncommon when Crohn's is confined to the small intestine).

Treatment for crohn depends on its location, severity and extra intestinal manifestation. According to ECCO guidelines treatment for mild crohn starts from mesalazine, Imuran and going forward for anti – TNF such as remicade and humira especially when extra intestinal manifestation. New biologics such as entyvio and stellara have a role in complicated non responding cases.

VW disease is 1/ million

Von will brand disease (VWD) is a genetic disorder caused by missing or defective von Will brand factor (VWF), a clotting protein. VWF binds factor VIII, a key clotting protein, and platelets in blood vessel walls, which help form a platelet plug during the clotting process.

We conclude that FVIIIvWF-mediated agglutination requires both functional platelet FVIIIvWF binding sites and platelet-platelet cohesion sites, and that platelet surface cohesion sites are altered by AET and PGI2. PGI2 from adjacent intact endothelial cells may prevent excessive platelet accumulation on exposed sub endothelium without suppressing an essential hemostatic process--the binding of platelets to sub endothelial FVIIIvWF.

It is possible that mesalamine diminishes inflammation by blocking cyclooxygenase and inhibiting prostaglandin (PG) production in the colon. Certain prostaglandins are likely to be beneficial in the treatment of gastric ulcers, stress ulcers, duodenal ulcers, and perhaps gastritis and certain forms of inflammatory bowel disease.

Additional contraindications per specific Canadian product labeling: Existing gastric or duodenal ulcer, urinary tract obstruction, use in children <2 years of age (Asacol, Asacol 800, Mesasal, Pentasa, Salofalk); hemorrhagic diathesis (Mesasal); patients unable to swallow intact tablet (Asacol, Asacol 800); renal parenchymal disease (Pentasa)

In our case we noticed that our patient had cholecystectomy with mild bleeding complication that was not noticed to establish the VW disease. Again he was scoped a year ago before his last colonoscopy, and in first procedure, there was no bleeding at the site of biopsy (notice that it was self limited as the patient lately confirmed), but in the last colonoscopy was a massive bleeding, that led to surgery and then the use of VW factor. The only thing that could trigger this disorder was Mesalazine as we showed recently in the pathogenesis of IBD and the action of mesalazine at some level. But still needs further studies to prove it.

Conclusion

In patients with crohn disease and VWD, we recommend avoiding the use of mesalazine.

Also all patients with crohn and VWD should start replacement therapy with VWF before colonoscopy in one week, to avoid bleeding post biopsy.

Replacement therapy should be continued for two to three days in patients who had a mucosal biopsy. For patients who underwent a therapeutic procedure, replacement should continue for up to two weeks.

References

Medscape, John Hopkins, ECCO guidelines, Journal of the Canadian Association of Gastroenterology (2018) 1: 310.