Post-operative Hemorrhage in Neurofibromatosis Type 1
Richard E Overman, M.D., Obeth Bahena Gutierrez, B.S., Michael D Honaker, M.D

Background
Previous reports have suggested a connection between neurofibromatosis type 1 (NF-1) and surgical bleeding complications, vasculopathy, or coagulopathy. To date, a single, specific mechanism for abnormal bleeding in NF-1 patients has yet to be elucidated and guidance in management is limited to case reports.

Discussion
Vasculopathy in NF-1 is rare (3.6% incidence) but is a main contributor of early death NF-1 due to the risk of hemorrhage.1, 2 Factors contributing to bleeding include: vessels with a thickened intima and a thin, fibrotic media; abnormal vascular structure within neurofibromatous tissue; and possible coexisting coagulopathies, including von Willebrand’s disease.3, 4

A variety of mechanisms have been postulated to explain peri-operative bleeding in NF-1 patients. While no definitive link exists, pre-operative evaluation for co-existing coagulopathy would be prudent.

Case
A 65-year-old female with a history of NF-1, anemia, and CKD, presented to the hospital in septic shock due to cystitis. While hospitalized, the patient developed a lower GI bleed. CT (Figure 1) demonstrated colo-colic intussusception and a colonic mass, felt to be both the lead point for the intussusception and likely the source of bleeding. Colonoscopy was unable to reduce or traverse the intussusception and the patient remained obstructed, prompting surgical intervention.

The patient underwent an uneventful Hartman’s procedure (Figure 2 – gross pathology specimen), with no intra-operative bleeding issues. Postoperatively, however, she developed hemodynamic instability, required blood transfusion, and increasing abdominal distention. At re-exploration, 2.4 L of hemoperitoneum was encountered without appreciable clot. Damage control sequence with re-exploration two days later was undertaken after correction of the patient’s coagulopathy, and the abdomen was closed.

Unfortunately, the patient developed several post-operative complications, including chronic respiratory failure, and multiple aspiration pneumonias. Her family elected comfort care and she died on post-operative day 24. Pathology demonstrated an unclassified high grade spindle cell sarcoma.

Conclusion
In patients with NF-1 undergoing surgery, pre-operative assessment for coagulopathy may be warranted, and a high index of suspicion for post-operative bleeding complications should be maintained.

References
1. Hassan S, et al. Spontaneous intestinal haematoma associated with neurofibromatosis type-1. DOI: 10.1136/bcr-2013-010512
2. Zhang K et al. Massive spontaneous hemorrhage in giant type 1 neurofibromatosis in soft tissue of chest wall. DOI: 10.2016/j.jtcvs.2012.05.046

Figure 1 CT scan demonstrated colo-colic intussusception (blue arrow), and intussuscepted mesenteric vessels (orange arrow)