

RF8. Management of Multiple Arterial Complications in a Patient with Ehlers-Danlos type IV

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OBJECTIVES: Describe minimally invasive open and endovascular approaches towards arterial complications associated with Ehlers-Danlos Syndrome type IV (EDS-4).

METHODS: 32 year-old gentleman diagnosed with EDS-4 (COL3A1 gene mutation), known previously thrombosed ulnar and tibial artery aneurysms, was transferred with a ruptured splenic artery aneurysm. He developed a retroperitoneal hematoma from a spontaneous ruptured ileolumbar artery. Identified on CT were hepatic and superior mesenteric artery (SMA) aneurysms. Events of his stay also included groin hematomas after sheath removal, bilateral femoral vein thrombosis, prolonged mechanical ventilation requiring tracheostomy, and spontaneous bilateral radial artery aneurysms.

RESULTS: The ruptured splenic aneurysm was treated with coil embolization. Groin hematomas were treated by open repair of the femoral artery. The retroperitoneal hematoma was treated with thrombin and coil embolization, followed by operative evacuation of the hematoma. An IVC filter was placed. Bilateral radial aneurysms (right 4 x 3 x 3 cm and left 3 x 3 x 2 cm) were treated by open ligation after confirmation of collateral flow via duplex occlusion test. At 6 months patient had healed all surgical wounds with no upper extremity motor/sensory deficits. Hepatic and SMA aneurysms on serial CT scans are unchanged.

CONCLUSIONS: EDS-4 patients develop arterial complications that can be treated with both open and endovascular techniques. A minimalistic approach is used due to the marked friability of the vessels. Ligation, when feasible, has the benefit of diminished risk of delayed suture line aneurysm and rupture. Observation and close follow up are crucial in the management.

