Abstract

Objectives: Intimomedial mucoid degeneration (IMD) is a rare vascular disorder characterized by mucinous deposition in the intima and media layers, which causes aneurysmal degeneration of the vessel wall in young patients. We report 3 patients with IMD (Mean age=30 years, SD=4.3) with variable clinical presentations.

Methods: Patients were diagnosed with IMD based on surgical histologic specimens of the affected vessel. Clinical presentation, diagnostic studies, therapeutic procedures and outcomes were reviewed for each case. Differential diagnoses included collagen vascular disorders, infectious pathologies and vasculitides. Histologic specimens from the involved vessels were analyzed using the Haemotoxylin and Eosin stain.

Results: Clinical presentations included one patient with a known posterior tibial artery aneurysm and new onset of local pain and paresthesias; one patient with a newly discovered
dorsalis pedis artery aneurysm; and one patient who presented emergently with a spontaneously ruptured non-aneurysmal subclavian artery at the thoracic outlet. Two patients underwent repair of the affected vessel with interposition grafts. The patient with the dorsalis pedis artery aneurysm underwent aneurysm excision and vessel ligation without reconstruction. Perioperative follow up was obtained in all 3 cases. One patient was later diagnosed with a symptomatic superior gluteal artery aneurysm and multiple other incidental aneurysms.

Conclusions: IMD is a rare entity affecting different vascular beds in men and women of different racial backgrounds that may present as aneurysmal degeneration or arterial rupture in the absence of a significant injury. Diagnosis is based on histology and, due to the potential for multiple vessel involvement, patients with confirmed IMD may need full body imaging and long term follow up to monitor for other sites of aneurysm formation.

Author Disclosure Block: