Tracheal obstruction caused by innominate artery pseudoaneurysm in a repeat surgical patient with Marfan syndrome

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Introduction: Marfan syndrome (MFS) is a rare genetic disorder resulting in pleiotropic manifestations involving multiple systems. Specifically, aortic abnormalities exist in up to 90% of patients with MFS, and some of these aortic abnormalities present with unique complications. We present a case of severe tracheal compression secondary to thrombosed innominate artery pseudo-aneurysm (IAPA) in a repeat surgical patient with MFS.

Case Presentation: A 35 yr old, 65, 96kg male with MFS presented for resection of a 7x9cm IAPA causing tracheal compression discovered during workup for progressive inspiratory stridor, dyspnea, and facial swelling. He had a complex past surgical history beginning at age 9: AVR, ascending aorta and arch replacement for type A dissection, descending thoracic aortic replacement, aortic arch and head vessel replacement, and aorto-bi-iliac graft placement.

The patient was prepared for awake FOI with inhalational and trans-tracheal lidocaine. An ENT surgeon was available for rigid bronchoscopy and cardiovascular surgeons were prepared for femoral cannulation if airway deterioration was encountered. A fiberoptic scope loaded with a 6.0 anode wire-reinforced ETT was advanced through the area of stenosis. The patient was induced after confirmation of appropriate ETT placement and return of carbon dioxide. Muscle paralysis produced no airway compromise.

Placement of a TEE probe yielded poor windows and intermittent tracheal compression. After sternotomy, our ability to ventilate began to deteriorate, secondary to ongoing surgical manipulation and dissection of the pseudoaneurysm. Progressive hypercarbia ensued, followed by hypotension and elevated CVP. FOB revealed increasing compromise of the distal trachea. Cardio-pulmonary bypass was initiated via femoral cannulation.

The remainder of the case proceeded without complication. Resection of the thrombosed pseudo-aneurysm resulted in improved ventilatory mechanics. Dysphonia, right brachial plexus injury and bilateral occipital CVA resulting in visual impairment complicated the post-operative course.

Discussion: Recurrent or progressive disease following operative repair of the aorta in patients with MFS is a significant problem. In one study, 70% of patients developed dissections or aneurysms, and over 50% proceeded to further operation(1). Although tracheal compression by innominate artery aneurysm in a patient with MFS has been previously described(2), this case highlights the complex challenges of repeated sternotomy and mediastinal mass effect on airway and hemodynamic management considerations in the repeat MFS surgical patient.
