Abstract:
Introduction: Retroperitoneal fibrosis (RPF) is an uncommon condition in which a fibrotic and inflammatory mass envelops and potentially obstructs retroperitoneal structures, including either or both ureters. Extension of the retroperitoneal plaque to involve the superior mesenteric artery (SMA) is rare and to the best of the author's knowledge this is the first reported case of mesenteric angina due to RPF. Case presentation: We report the case of a lady with retroperitoneal fibrosis resulting in obstructive uropathy, who was initially managed with ureterolysis, omental wrapping and medical therapy (steroids and Tamoxifen). She improved with the above management. However, over the course of 12 months, her disease progressed and she presented with complaints of persistent post prandial upper abdominal pain. The repeat CECT abdomen and pelvis showed progression of the retroperitoneal plaque to involve the superior mesenteric artery. SMA stenting was done after which her symptoms improved. Conclusion: Retroperitoneal fibrosis is rare and involvement of the superior mesenteric artery is even rarer. In a patient with RPF and persistent abdominal symptoms, there should be a high index of suspicion of SMA involvement. SMA stenting is the preferred minimally invasive option in SMA involvement in RPF.

Keyword:
Retroperitoneal fibrosis, Superior mesenteric artery, mesenteric angina

Introduction:
Retroperitoneal fibrosis (RPF) is an uncommon condition in which a fibrotic and inflammatory mass envelops and potentially obstructs retroperitoneal structures, including either or both ureters. We report the case of a lady with retroperitoneal fibrosis resulting in obstructive uropathy, who initially improved with ureterolysis, omental...
wrapping and medical therapy (steroids and Tamoxifen), but later progressed to involve the superior mesenteric artery (SMA). Extension of the retroperitoneal plaque to involve the SMA is rare and to the best of the author’s knowledge this is the first reported case.

Case Report:
A 52 year old lady, on evaluation of fever and loin pain was found to have B/L hydronephrosis and acute renal failure and managed initially with bilateral DJ stenting elsewhere. Further investigations, after renal functions returned to normal, revealed features of retroperitoneal fibrosis as evidenced by CECT findings of a mantle of tissue from below the origin of the SMA to the iliacs and to the pelvis on the left, encasing the aorta and common iliac vessels, and both ureters. (Fig 1,2)

Her ESR was 110mm/h; CRP was raised at 13.8mg/dl; C4-49.1 and C3C 172mg/dl. After optimization she underwent bilateral ureterolysis and omental wrapping. Per operatively a firm fibrotic plaque was found overlying the lower lumbar vertebrae extending into the sacral promontory with both ureters were medially deviated and encased in the plaque. Left ureter was thickened and densely adherent at the L4-L5 level. Right ureter was more supple compared to the left. The biopsy of the plaque had fibrosis, hyalinization and moderate chronic inflammation. She was started on steroid and Tamoxifen therapy on which she improved. Her renal function improved and ESR came down to 11mm/hr; CRP-0.433mg/dl. However, over the course of 12 months, her disease progressed and she presented with complaints of post prandial pain. The repeat CECT abdomen and pelvis showed progression of the retroperitoneal plaque to involve the superior mesenteric artery. (Fig 3,4)

Fig 1&2 CECT showing the retroperitoneal plaque beginning from below renal arteries and no plaque above(2)
Fig 3&4 CECT showing entrapment of SMA in retroperitoneal plaque

SMA stenting was done after which her symptoms improved.

Fig 6 & 7 Post SMA stent placement angiography and CECT

Discussion:
Albarran (1905), is credited for the earliest report of RPF. Ormond described and defined retroperitoneal fibrosis and thus RPF has had the eponym Ormond disease.(1,2) Retroperitoneal fibrosis appears as a fibrous, whitish plaque encasing the aorta, inferior vena cava, and their major branches, the ureters, and other retroperitoneal structures, Intraperitoneal structures including the gastrointestinal tract may also be affected at times. Its longitudinal axis usually extends from the renal hilum to the pelvic brim, but it may extend into the pelvis, mediastinum. and even the optic orbit. Histologically, a fibrous component and a chronic inflammatory infiltrate composed of lymphocytes, macrophages, plasma cells, and eosinophils are present. Both perivascular and diffuse infiltrates may be present. The fibrotic component consists of myofibroblasts and type-1 collagen.(3) Many mechanisms for the etiology of RPF have been proposed. Development of vasculitis in the adventitial vessels of the aorta and peri-aortic small vessels is one of them. Subsequent release of antigens from atheromatous plaque such as ceroid, a complex lipoprotein, has been proposed which induces an autoimmune antigenic response.(3) Other immunologic events may play a role. Both CD3+ and CD20+ lymphocytes and IgG4-positive plasma cells have been identified.(4) Asbestos exposure as a risk factor for the development of ‘retroperitoneal fibrosis’ was proposed in 2004 by Uibu et al.(5) Epidemiology is not well defined. Incidence in Finland was reported as 0.1 per 100 000 person-years and a prevalence of 1.38 per 100
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000. It is more common in males, with a 2 to 3:1 male-to-female ratio. Mean age at onset is typically 50 to 60 years, but it may manifest in children as well as the elderly.(3) Genetic influence is not thought to play a major role, because there have been no reports of familial clustering. Retroperitoneal fibrosis is associated with HLA-DRB1*03, an allele linked to a number of autoimmune diseases.(6) Medications have been associated with the development of retroperitoneal fibrosis. A definitive etiology of retroperitoneal fibrosis is found in only 30% of cases. (7) The term “idiopathic retroperitoneal fibrosis” should be used only when an inciting etiology is not defined. An underlying malignancy should always be considered, because one is reported to be present in 8% to 10% of such cases. CECT reveals a wellmarcated retroperitoneal mass, isodense with muscle on unenhanced studies, allows superior soft tissue discrimination and can more accurately distinguish the plaque from the great vessels than unenhanced CT.(8) Biopsy to exclude malignancy should be performed, either percutaneously with CT, MRI, or ultrasound guidance. If there is obstructive uropathy at presentation, ureteral stenting, or percutaneous nephrostomy is indicated.(3)

Medical therapy consists of steroids and tamoxifen (thought to alter the TGF beta). Ureterolysis is undertaken if medical therapy fails or if the patient is not a candidate for medical therapy. Traditionally, this has been done by an open surgical approach. Laparoscopy and robotic options exist.

Conclusion:

Retroperitoneal fibrosis is rare and involvement of the superior mesenteric artery is even rarer. In a patient with RPF and persistent abdominal symptoms, there should be a high index of suspicion of SMA involvement. SMA stenting is the preferred minimally invasive option in SMA involvement in RPF.

References:


Fig 5 Pre stenting angiography