Journal of Advances in Medicine and Medical Research

32(11): 15-25, 2020; Article no.JAMMR.58901
ISSN: 2456-8899
(Past name: British Journal of Medicine and Medical Research, Past ISSN: 2231-0614, NLM ID: 101570965)

Chronic Periaortitis – Review and Presentations

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Authors' contributions

This work was carried out in collaboration among all authors. Author ST designed the study, completed all the clinical research work, wrote the protocol, retrieved all the images from the radiology department and wrote the first draft of the manuscript. Authors SB and PT summarized the clinical data and carried out an extensive literature search and researched the topic and helped with the first draft. Authors VS and VB were the consultants responsible for clinical care of the patients and managed their treatment after extensive multidisciplinary team discussions. They revised the paper and summarized the discussion and conclusions. All authors read and approved the final manuscript.

Article Information

DOI: 10.9734/JAMMR/2020/v32i1130525
Editor(s):
(1) Thomas I. Nathaniel, University of South Carolina, USA.
Reviewers:
(1) S. N. Kumar, APJKTU, India.
(2) Abhinav Mahajan, Sri Guru Ram Dass University of Health Sciences and Research, India.
Complete Peer review History: http://www.sdiarticle4.com/review-history/58901

Case Study

Received 03 May 2020
Accepted 09 July 2020
Published 22 July 2020

ABSTRACT

Chronic periaortitis is a rare inflammatory condition predominantly affecting the abdominal segment of the aorta. This can present as IgG4 related inflammatory disease, idiopathic retroperitoneal fibrosis, perianeurysmal retroperitoneal fibrosis and inflammatory abdominal aortic aneurysm (IAAA). Aortitis can also be a manifestation of a number of rheumatological large vessel vasculitides such as Takayasu arteritis and giant cell arteritis (GCA). We present three interesting cases of chronic periaortitis and a literature review. The first case shows a classic picture of IgG4 periaortitis. The second case illustrates periaortitis with retroperitoneal fibrosis, ureteric involvement and hydronephrosis, following abdominal aortic aneurysmal stenting. The final case presents as widespread periaortitis due to Takayasu's disease involving the entire aorta including the arch and root of the subclavian artery.

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Keywords: Periaortitis; IgG4 related disease; retroperitoneal fibrosis; Takayasu's disease; FDG- PET scan; CT scan; corticosteroids.

1. INTRODUCTON

Immunoglobulin G4 related disease (IgG4-RD) is a systemic inflammatory disease associated with high levels of serum IgG4, a dense lymphoplasmacytic infiltrate rich in IgG4 plasma cells and fibrosclerosis. [1] It is characterised typically by plasma cell infiltration of organs it affects [2]. It is thought to be autoimmune in nature and is seen most commonly in middle aged to elderly age men.

IgG4-RD has been associated with organ dysfunction of the lungs, pancreas, kidneys, biliary tree, prostate and even the cardiovascular system [3,4,5,6]. of note, significant associations have been made with autoimmune pancreatitis and retroperitoneal fibrosis [3]. The most common presentations of IgG4 related disease tend to be pancreatic disease where the disease causes sclerosing autoimmune pancreatitis. In addition, where IgG4 related disease affects the kidneys it commonly causes tubulointerstitial nephritis [7]. Other common presentations include proptosis where IgG4-RD affects the orbit, also associated with diplopia due to tethering of the extra-ocular muscles. All salivary glands (parotid, sublingual, submandibular) can also be affected by IgG4-RD where it causes significant swelling and symptoms of dry mouth, similar to other autoimmune diseases [7]. Associated with this IgG4-RD can cause dacryoadenitis, where the lacrimal glands swell as a result of being affected with this disease, presenting as periorbital swelling.

These manifestations are all a result of lymphoplasmacytic infiltrates rich in IgG4+ immunoglobulin in plasma cells [8]. Where IgG4 related disease presents as a mass, as described in presentations above, histological diagnosis is key to diagnosing this condition [7]. Serum markers shown in presentation of the disease demonstrate eosinophilia and hypergammaglobulinemia, however these are non-specific and hence histological diagnosis and extensive imaging is helpful for diagnosis and subsequent management [7]. IgG4 related periaortitis have been reported in the literature a handful of times over the last few years [9,10,11,12].

Retroperitoneal fibrosis (RPF) is a condition characterized by a highly fibrotic retroperitoneal mass that frequently causes ureteral obstruction. This can arise around a dilated or undilated aorta. RPF is considered to be idiopathic in the majority of cases however secondary conditions like iatrogenic drug use, radiotherapy or infections can be strongly associated with this. [13] Retroperitoneal fibrosis can also be IgG4 related or non-related with 20%- 53% of IgG4 RD patients having this finding [14]. Pathogenesis behind this remains uncertain however it is thought that there is a local inflammatory response to low density lipoprotein (LDL) and ceroid found in atherosclerotic plaque of the aorta. A combination of this with damage and thinning of the aortic wall is thought to trigger the process.

2. CASE 1 (IGG4 RELATED DISEASE)

A 78-year-old gentleman presented to the emergency department with a 3-week history of central abdominal pain, change in bowel habits, abdominal distension, fatigue, loss of appetite and significant weight loss (approximately 12 kg) over a 2-month period. His background medical history consisted of ischaemic heart disease, heart failure, atrial fibrillation, chronic kidney disease stage 3, peripheral vascular disease, idiopathic pulmonary fibrosis, hypothyroidism, previous perforated duodenal ulcer and femoro-popliteal bypass. He was treated for deep vein thrombosis & pulmonary embolism in the past and was on long term warfarin. He had been a heavy smoker and had previous asbestos exposure. General, abdominal and systemic examination was unremarkable. Blood investigations reported normal renal and liver function tests. He had a raised C-reactive protein at 50 mg/L.

CT scan of chest, abdomen and pelvis (CAP) during this admission suggested abdominal aortic mural thickening and significant periaortic inflammation at the origin of superior mesenteric artery (SMA) without any evidence of SMA stenosis or distal mesenteric ischaemia. CT scan raised a suspicion of periaortitis with underlying connective tissue disorder. He received suitable analgesics, dietician's input and dietary meal supplements as well as laxatives for symptom control. Outpatient colonoscopy with appropriate bowel preparation and whole body PET (Fluorodeoxyglucose Positron Emission Tomography) scan was arranged and he was discharged home.

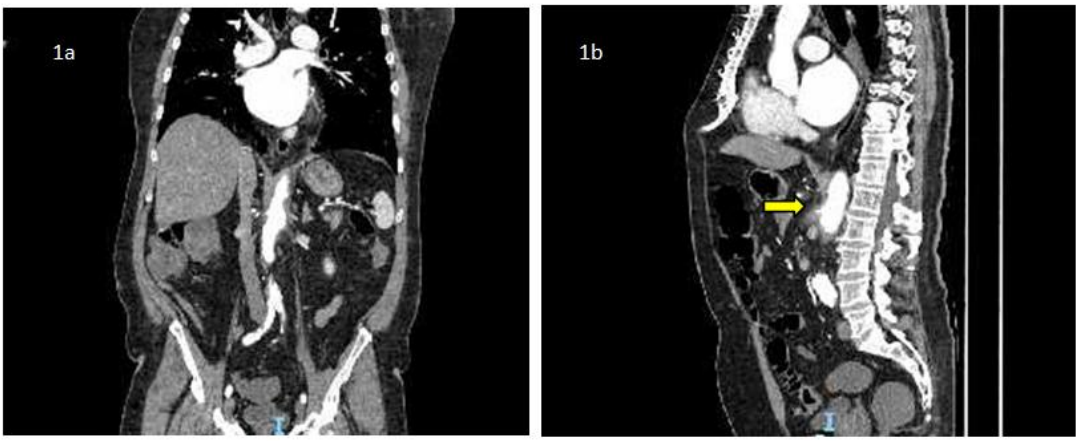

Fig. 1a. Contrast enhanced CT CAP coronal view demonstrating IgG4 related periaortic inflammation. Fig. 1b. CT CAP sagittal view with evidence of inflammation around the SMA origin with associated mural thickening (solid arrow)

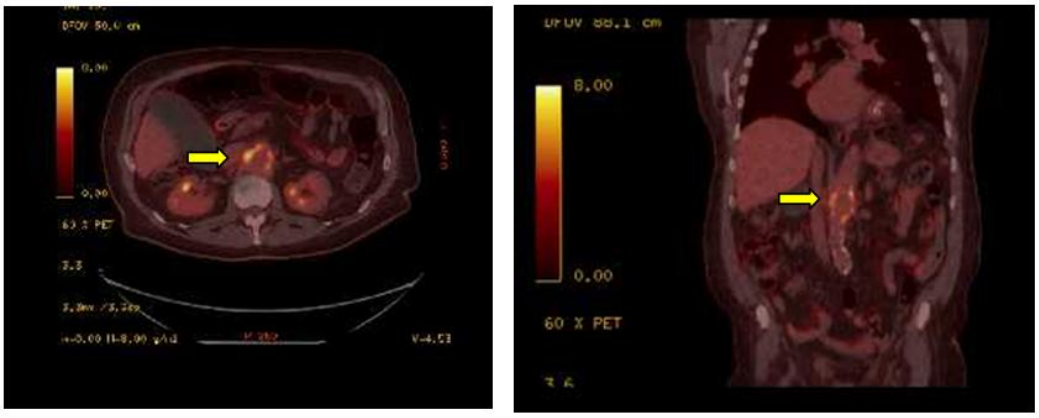

Fig. 2a. FDG PET scan axial view showing intense accumulation of ^{18}F – FDG around the abdominal aorta (solid arrow). Fig. 2b. FDG PET coronal view showing increased uptake around the SMA origin (solid arrow)

Subsequently he underwent a whole body FDG PET scan in 2 weeks 'time which suggested moderate grade FDG activity correlating with the distribution of the aortic mural thickening on the recent CT scan. The involved section of abdominal aorta measured approximately 5.5 cm in cranio- caudal diameter commencing at the level of the coeliac axis and terminating below the level of the renal arteries. There was involvement of the origins of both renal arteries, SMA and to a lesser extent of coeliac axis. Normal bio-distribution of FDG was noted elsewhere within the neck, chest, abdomen and pelvis. Thereafter he underwent ultrasound scan of the craniofacial soft tissue which indicated normal temporal and axillary arteries. Colonoscopy was not attempted due to persistent abdominal pain. Instead he underwent CT colonoscopy which suggested presence of moderate sigmoid diverticulosis otherwise unremarkable appearance of the rest of the colon.

His immunological profile reported positive antinuclear antibody. Anti-neutrophil cytoplasmic antibody (ANCA) was negative. The immunoglobulin (IgG) Subclass assay suggested raised total IgG –17.4 g/L (normal range 6.0-16.0) , raised IgG1 – 11.68 g/L (normal range 3.8-9.3), IgG 2 – 4.39 g/L (normal range 1.2-6.6), IgG3 – 0.66 g/L (normal range 0.2-1.8) and raised IgG4 -1.81g/L (normal range 0-0.9).

He was started on prednisolone 40 mg once daily for 4 weeks followed by 30 mg once daily for 4 weeks. He was discharged home and outpatient follow up appointment was arranged. On further clinical review in 2 months' time, by the rheumatologist and gastroenterologist, his abdominal symptoms settled and bowel habits more or less regularised. However, he complained of new onset dysphagia, early satiety, reduced appetite and weight loss. He was started on high dose omeprazole and an urgent upper GI endoscopy was organised which he later declined. He continued to remain on maintenance dose of prednisolone 15 mg once daily. He declined any further imaging investigations and did not attend follow up appointment. Sadly, he passed away 9 months later due to unrelated causes.

3. CASE 2 (IDIOPATHIC RETROPERITONEAL FIBROSIS)

A 78-year-old gentleman, with a background medical history of myocardial infarction requiring coronary stents and ankylosing spondylitis was under regular surveillance with the vascular surgeons for monitoring of infra-renal abdominal aortic aneurysm (AAA) by 6 monthly ultrasound scans. Over a period of 3 years, the AAA increased in size from 37 X 37 mm to 58 X 58 mm. Hence various treatment options were offered to him including open versus endovascular repair of the AAA when he preferred a minimally invasive approach. He underwent an uneventful endovascular repair of this aneurysm. Subsequent 2 months' post-procedure surveillance US scans suggested patent aortic stent graft and iliac limbs with no evidence of endoleak.

He required hospitalisation with acute abdominal and severe back pain 6 months following the procedure. His CT scan of the abdomen and pelvis suggested left hydronephrosis and evidence of soft tissue thickening around the aorta thought to be periaortitis with retroperitoneal fibrosis.

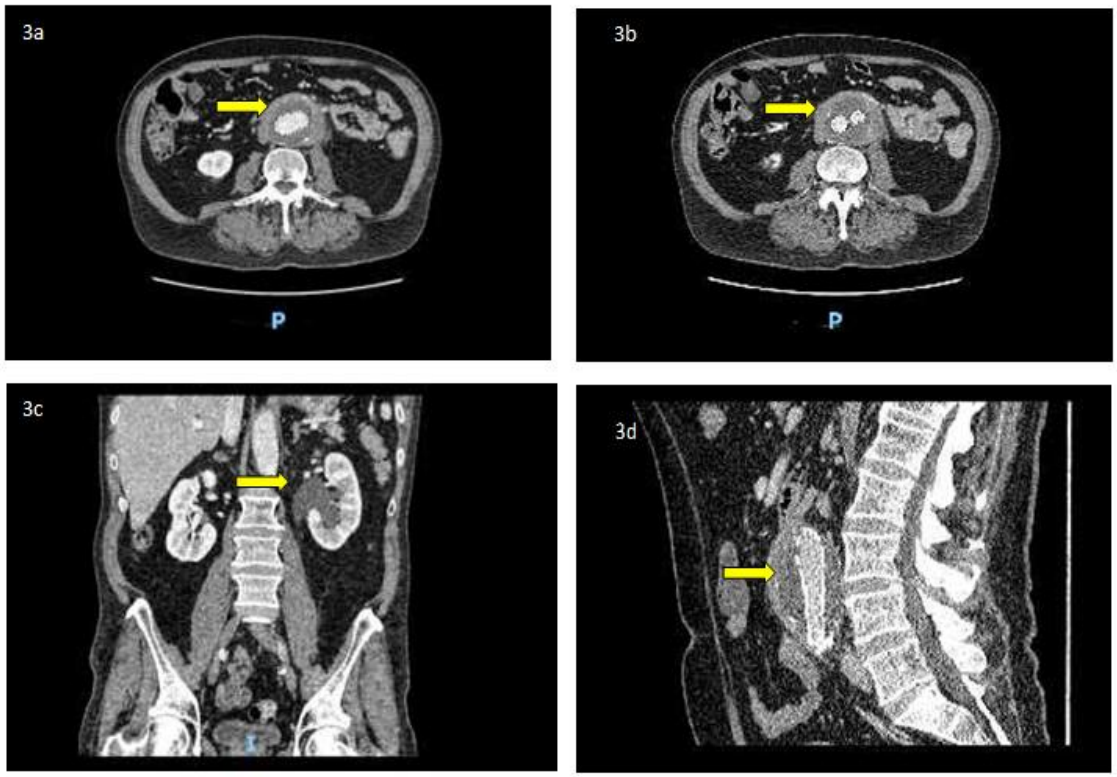

Fig. 3a &3b. Contrast enhanced CT CAP axial view showing evidence of inflammation and mural thickening around the endovascular stent repair. Fig. 3c. Contrast enhanced CT CAP coronal view indicating hydronephrotic left kidney. Fig. 3d. Contrast enhanced CT CAP sagittal view

Serial blood cultures to check for infective causes were negative. His blood investigations indicated normal full blood counts, renal and liver function tests. Antinuclear antibody screen was negative at 25 LIU (normal range 0-48 LIU), Anti-Neutrophil cytoplasmic antibodies were negative at 5 Light intensity units (normal range 0-37 LIU) and extractable nuclear antibody (ENA) was negative.

Serum immunoglobulins including IgG subclasses levels were within normal limits and he could therefore not be classified as IgG related disease. He underwent left ureteric stent insertion and was started on prednisolone 40 mg once daily dose with tapering reduction in the dosage over a period of 3-4 months. On subsequent rheumatology review, his abdominal pain had mostly settled and ESR reduced from 51 mm /hr to 14mm/hr, indicating settled periaortitis.

Latest surveillance ultrasound scan performed 3 years since the endovascular repair indicated reduction in the size of the abdominal aortic aneurysm with no evidence of endoleak and minimal aortic wall thickening anteriorly. He continues to remain under annual surveillance.

4. CASE 3 (PERIAORTITIS RELATED TO TAKAYASU'S DISEASE)

A 35-year-old male presented to the accident and emergency department with sudden onset severe central abdominal pain. He experienced similar episodes of pain over a period of 5 years with each episode lasting for a few days associated with nausea, constipation, sweating and shivering. He had been an ex-smoker. Bloods including liver function tests, renal function tests, serum amylase were entirely within normal limits. His ESR was raised at 88 mm /hr. He had negative antinuclear, negative cardiolipin antibodies and normal complement levels. His CT CAP suggested diffuse circumferential thickening of the lower abdominal aorta and proximal aspect of the common iliac arteries, with evidence of periaortic stranding. This was consistent with the findings at the whole body FDG PET scan.

He was commenced on prednisolone 40 mg once daily and methotrexate 15 mg /week in combination with folic acid 5 mg/week. Oral administration of methotrexate was subsequently changed to subcutaneous dose of 25 mg weekly. After obtaining good response to the methotrexate, his prednisolone dose was tapered over the following few months based on the ESR results. Over the next few months, he developed relapse of his symptoms of abdominal pain despite of maximum dose of methotrexate and prednisolone. His repeat FDG PET, 8 months since his diagnosis suggested persistent periaortitis and inflammatory markers continued to demonstrate an upward trend. Hence, he received 6 pulses of cyclophosphamide at 15 mg/kg with methylprednisolone 10 mg /Kg dose. The dose of prednisolone was tapered gradually and mycophenolate was added to his medications. Nearly one and a half year since commencing the treatment of periaortitis, he started experiencing left upper limb claudication. His left upper limb pulses were not palpable and blood pressure was not recordable although the limb remained viable.

He underwent a repeat whole body FDG PET scan which demonstrated increased 18F – FDG uptake around the arch of aorta, left subclavian artery and right vertebral artery typical of Takayasu's disease.

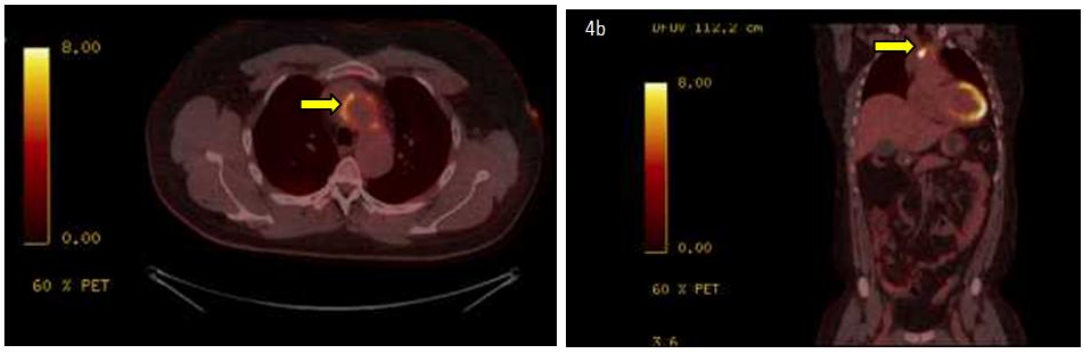

Fig. 4a & 4b. FDG PET scan axial showing intense accumulation of ^{18}F – FDG around the arch of aorta and left subclavian artery (solid arrow)

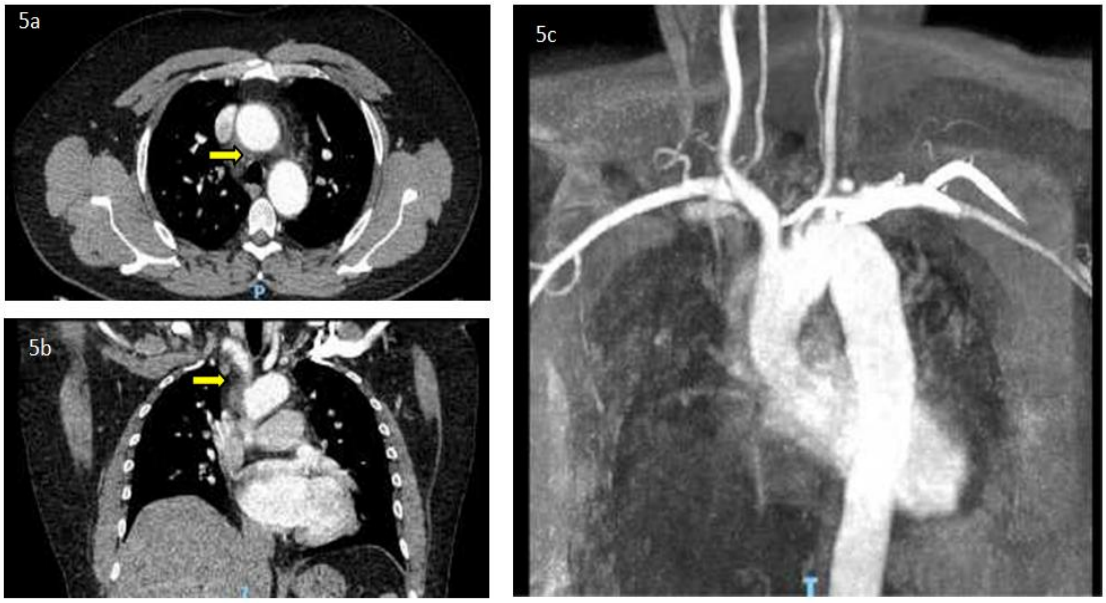

Fig. 5a & 5b. CT aortogram (axial and coronal view) highlighting periaortic inflammation around the thoracic aorta, brachiocephalic vessels and occlusion of left subclavian artery (solid arrows). Fig. 5c. MR angiogram showing significant periaortic inflammation

This was then followed by CT angiogram which demonstrated significant stenosis of the left subclavian artery. Magnetic resonance (MR) angiogram confirmed bilateral subclavian arterial disease with possible under-filling of the left vertebral artery a feature classically seen in Takayasu's disease.

Due to progressive disease, he received monthly infusion of tocilizumab under careful monitoring for a period of further 6 months with significant improvement in the left upper limb perfusion and inflammatory markers. However, his symptoms of left upper limb claudication persisted. CT aortogram during a further admission suggested progressive disease and CT coronary angiogram demonstrated penetrating ulcer along the anterior proximal ascending aorta. It appeared that he was responding to higher doses of prednisolone.

However, he required emergency admission to critical care with chest pain, shortness of breath, productive cough and haemoptysis. CT angiogram indicated no increase in the severity of periaortitis with ground glass changes in the lungs. Echocardiogram suggested globally diminished cardiac function. Despite of antibiotics, antifungals, methyl-prednisolone, cyclophosphamide, tocilizumab, heart failure treatment, renal replacement therapy and ventilatory support he progressively deteriorated and sadly passed away.

5. DISCUSSION

Chronic periaortitis tends to affect the infra-renal abdominal aorta as shown by Ozawa et al [2], similar to non-inflammatory aneurysmal disease. The pathogenesis of IgG4-related periaortitis is unclear and several theories are present in the literature. Castelein et al suggest that atherosclerotic plaque is a key feature of this process [15]. When comparing groups of patients, one with IgG4 related periaortitis and one with idiopathic periaortitis, they found the calcium content (as a marker for atherosclerosis) was significantly higher in those patients with IgG4 related periaortitis [16]. It is speculated that chronic periaortitis is triggered by an immune response to existing atherosclerotic plaque antigens. In particular there is a response to oxidized low-density lipoproteins and ceroid which are common components of the plaque [17]. These antigens are presented to B cells and T cells which are activated in the adventitial aortic layers.

IgG4 positive plasma cells infiltrate the adventitial layer of the aorta therefore giving the appearance of thickened vascular walls [18]. Other autoimmune vasculitic disease such as

Takayasu arteritis typically affects also the intima and the media however in IgG4-RD, these layers are relatively spared.

5.1 Risk Factors

IgG4 related periaortitis has a predisposition for older patients [2]. In addition, those diagnosed with IgG4 related periaortitis generally have high serum levels of IgG4 [15,10]. Interestingly despite abdominal aneurysmal disease being commonly associated with hypertension and smoking, these factors do not seem to play a role in developing periaortitis [2]. It seems the typical associations with atherosclerosis do not attenuate risk in these patients.

5.2 Clinical Manifestations

Chronic periaortitis often presents with nonspecific symptoms of dull abdominal or flank pain, fatigue, weight loss, low-grade fever, nausea and anorexia. The pain is characterised as persistent and is poorly localized in nature. Ureteric colic and testicular pain have also been reported. Retroperitoneal fibrosis can present with unilateral or bilateral hydronephrosis, obstructive uropathy and ultimately chronic renal failure. The obstruction thought to be associated with oedema rather than fibrosis as it responds to corticosteroid treatment. Further manifestations related to compressive effect of the mass lesions include scrotal swelling, varicocele, deep vein thrombosis and leg claudication [1,18,19]. Inflammatory abdominal aortic aneurysms present with a classic triad of pulsatile tender abdominal mass, back and abdominal pain and raised erythrocyte sedimentation rate. In these cases, a periumbilical bruit may be present.

5.3 Investigations

Chronic periaortic infection must first be ruled out by repeated blood cultures, white cell counts and white blood cell radioisotope uptake scans. Blood tests may show a raised CRP or ESR as seen in 80-90% of patients with chronic periaortitis. Full blood counts and renal function test can show declining kidney function with normochromic anaemia indicating systemic chronic inflammation. Polyclonal hypergammaglobulinemia is a principal feature with raised total serum IgG levels, with a greater rise in IgG1 and IgG4 as demonstrated in our first case. Autoimmune blood markers such as anti-nuclear antibodies, rheumatoid factor, anti-smooth muscle antibodies, anti- dsDNA and anti Ro(SSA)/La(SSB) are not features of IgG4 periaortitis although they may suggest the presence of other autoimmune conditions [20,21].

Imaging modalities include computed tomography (CT), cardiovascular magnetic resonance (CMR), echocardiography and vascular ultrasound. Computed tomography is the modality of choice when assessing and investigating periaortitis [2]. Radiological findings are characterized by thickening of the arterial wall associated with a high-density thin layer. This represents the intima-media inflammatory complex of IgG4 positive plasmacytes and associated fibrosis. IgG4-related periaortic lesions are well circumscribed and display homogenous enhancement during the late phase of contrast enhanced CT imaging [22]. Presence of heterogeneous enhancement, ill-defined margins, cystic changes or lymphadenopathy suggest occurrence of malignancy which is a key differential diagnosis [20]. The periaortitis is mostly noted to affect infra-renal abdominal aorta [23]. Ozawa et el identified the involvement of infra-renal artery regions in >80% of periaortitis cases. Medium sized vessels have also been affected in a few cases with similar radiological findings. Further periaortic lesions can present with luminal changes such as aneurysmal dilation – with accompanied inflammatory aortic aneurysm, small vessel penetration and aortic dissection.

Magnetic resonance imaging can further be used to identify chronic periaortitis. Lesions of inflammation and fibrosis appear hypointense on T1 weighted images and hyperintense on T2 weighted images [21]. Ultrasonography may be used in assessing the degree of aortic dilation in aneurysmal chronic periaortitis and hydronephrosis.

^{18}F-Fluoro-deoxyglucose positron emission tomography (FDG-PET) is a new diagnostic method in identification of periaortitis. It is a reliable tool in assessing the degree of metabolic activity of the lesion. There is increased uptake of FDG in the area of the suspected lesion and this is more prominent during the early stages of inflammation. This technique has been adopted to identify a number of other IgG4 related organ involvement [24].

CT guided biopsy and subsequent histopathology may be possible for extravascular or

retroperitoneal mass lesions. Haematoxylin and eosin staining identify the presence of lymphocytoplasma cells, fibrin deposition, obliterative phlebitis and fibrosis. Immunohistochemistry can be carried out to identify the presence of IgG4 positive plasma cells. However, absence of positive pathological findings is a common feature due to practical challenges of sampling the retroperitoneum, periaortic region, kidney and pancreas [25].

5.4 Management

Corticosteroids are the mainstay of medical treatment used to reduce the degree of periaortic lesion thickness, inflammation and associated fibrosis [29]. Rapid symptom remission occurs within few weeks of commencing treatment. Their use in conjunction with surgical intervention has yielded good results. In a retrospective study on 40 patients treated with corticosteroid therapy for IgG4 related periaortitis the authors identified a significantly positive effect in preventing new aneurysm formation in patients without prior luminal periaortic lesion dilatation [15]. Wagenknecht et al described reduced ureteral re-stenosis rates when used in patients with further retroperitoneal fibrosis. On the other hand, there is some suggestion that corticosteroid therapy may increase the risk of pre-existing aneurysmal rupture [26], therefore its preoperative use in patients with IAAA is still debated.

Therapeutic alternatives such as azathioprine, mycophenolate and methotrexate have been identified in smaller case reports as having the potential for disease clearance [19,26,27). Tamoxifen, a selective oestrogen receptor modulator has also been used as a single agent and in conjunction with corticosteroids [28]. This has greater response in reducing retroperitoneal fibrosis. Further clinical studies comparing the efficacy of the immunosuppressant drugs is warranted.

Surgical involvement may be required in cases of aneurysmal periaortitis. Aim of early intervention and monitoring is to prevent aneurysmal rupture. Endovascular prosthesis insertion and surgical intervention are often recommended once aortic diameter exceeds 5.5cm [29]. The presence of dense adhesions around structures such as the inferior vena cava, ureters and duodenum further challenge management. Mortality rates are between 0.9 – 5% for elective aneurysm repair with values similar to the non-inflammatory type [30]. Open IAAA repair was originally adopted however in light of its increased morbidity and mortality, endovascular aneurysm repair (EVAR) has now become increasingly common. However long-term regression of the aneurysmal fibrosis is found to be lower in endovascular repair versus open repair. This is due to a potentially inflammatory response aggravated by EVAR [31]. Individuals with retroperitoneal fibrosis carries a high possibility of developing chronic kidney disease due to ureteric compression and obstruction. Ureterolysis and ureteric stent insertion is used in patients with hydronephrosis [28].

6. CONCLUSION

Chronic periaortitis is a rare condition associated with several overlapping causes. IgG4 related disease, idiopathic retroperitoneal fibrosis and Takayasu's have similar clinical presentations although the IgG4 levels need to be raised in the first condition. Retroperitoneal fibrosis typically results in ureteric obstruction as noted in the second case and subclavian artery involvement in a young male adult points to a diagnosis of Takayasu's as seen in the third case. FDG PET scans show high uptake around the aorta and is the new mode of investigation. However, CT imaging remains the gold standard in diagnosis and monitoring disease progression. Tissue biopsies, whereby possibly, can clinch the diagnosis although this may be difficult when large vessels are involved. A high index of suspicion should be present in any patient without any infection but presenting with persistent signs of inflammation.

CONSENT

As per international standard patient's consent has been collected and preserved by the authors.

ETHICAL APPROVAL

As per international standard written ethical permission has been collected and preserved by the author(s).

COMPETING INTERESTS

Authors have declared that no competing interests exist.

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Peer-review history:
The peer review history for this paper can be accessed here:
http://www.sdiarticle4.com/review-history/58901