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Case Report

Report of a rare case of periaortitis at the aortic arch and successful therapeutic strategy with prednisolone

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ABSTRACT

Periaortitis is one of the less common manifestations of the aorta pathology. Periaortitis mostly arises from the outer layers of the abdominal aorta and iliac arteries. However, other large arteries may also be involved, but rarely the thoracic aorta. Here we present a successful conservative treatment using prednisolone therapy for periaortitis of the aortic arch which shows various clinical symptoms.
<**Learning objective:** Periaortitis is one of the less common manifestations of the aorta pathology. Periaortitis rarely has an impact on the thoracic aorta. The treatment strategy for this complication, especially the dose of prednisolone, is not well established. This case report supports documenting periaortitis in the aortic arch. Not only did we begin initial prednisolone treatment at 45 mg/day, but also performed a downward titration after only half a year.>

Introduction

Periaortitis is one of the less common manifestations of the aorta pathology. Periaortitis has an impact mainly on the abdominal aorta and the iliac arteries, however, the thoracic aorta and main branches of the aorta may also be involved. Periaortitis mostly arises from the outer layers of the abdominal aorta and iliac arteries. Periaortitis associated fibro-inflammatory diseases usually involve the surrounding retroperitoneum such as the ureters and inferior vena cava, but hardly involve the thoracic aorta. This case report supports documenting periaortitis in the aortic arch. Not only did we begin initial prednisolone treatment at 45 mg/day, but also performed a downward titration after only half a year.

Case report

A 56-year-old Asian male, who was operated for a subarachnoid hemorrhage one month previously, experienced persistent low grade fever and chest oppression during rehabilitation. While computed tomography (CT) without enhancement findings raised suspicions of an acute aortic dissection, he was re-admitted to our hospital proposed for an emergent aortic operation. His body temperature was 37.5 °C, pulse was regular, heart rate 68 beats/min, and blood pressure 120/70 mm Hg. Abnormal laboratory findings included high levels of C-reactive protein (CRP: 10.86 mg/dL; normal range: <0.2 mg/dL), with a normal leukocyte count [white blood cells (WBC: 6700/mL; normal range: 4,500–10,000/mL]. By additional examination, immunoglobulin G (IgG) was 1810 mg/dL (normal range:870–1700 mg/dL) and immunoglobulin G4 (IgG4) was 40 mg/dL (normal range:4–80 mg/dL). But enhanced CT showed a soft tissue density mass around the aortic arch without aortic dissection or swollen lymphoid (Fig. 1a, b, c). Furthermore, the lateral contrasted adventitia at the arch was similar to the aortic mantle sign. While we waited and saw the patient to ascertain that an infection did not coexist, finally we arrived at the diagnosis of periaortitis of the aortic arch with neither dissection nor aneurysm. At the time, the patient experienced hoarseness and an otolaryngologist confirmed that he had a left recurrent nerve paralysis. We initially suggested prednisolone therapy for the periaortitis, but the patient wanted to return to the rehabilitation hospital during which, prednisolone therapy was not started. One week later, he was discharged and

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a

b

c

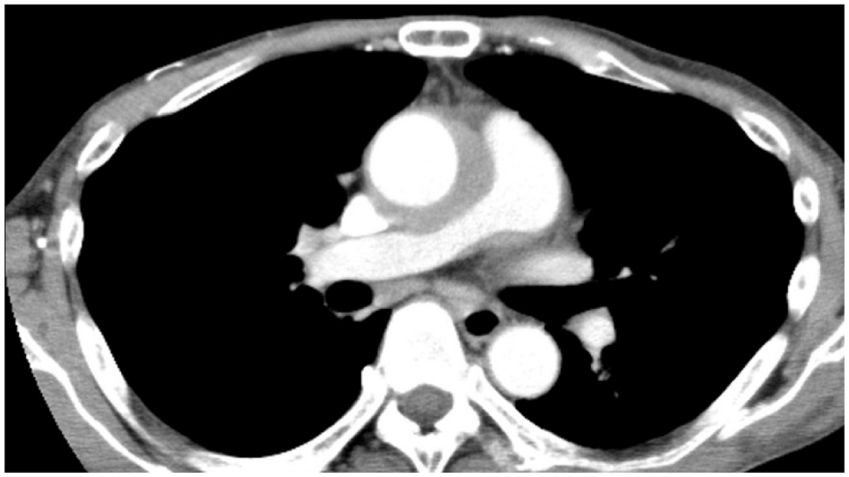
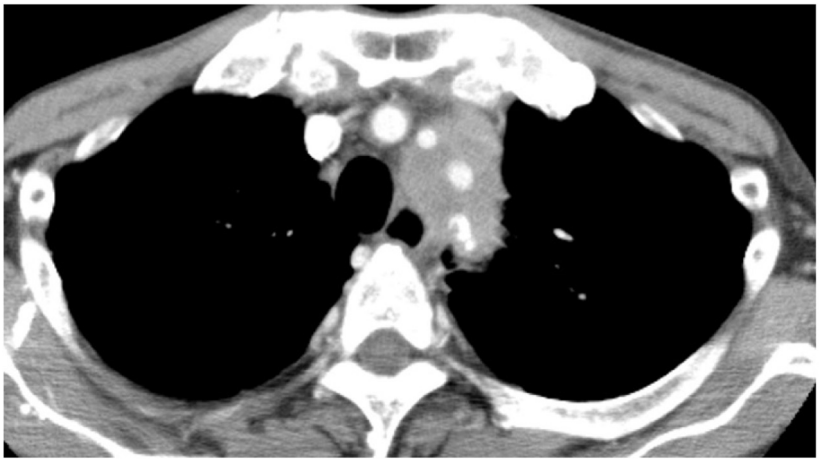


Fig. 1. Computed tomography (a) revealed the soft density mass around the aortic arch without evidence of any dissection or rupture. Inflammation surrounded cervical branches (a) and arch (b). The lateral contrasted adventitia at the arch was similar to the aortic mantle sign (b). The mass compressed slightly pulmonary main trunk (c).

a



b

c

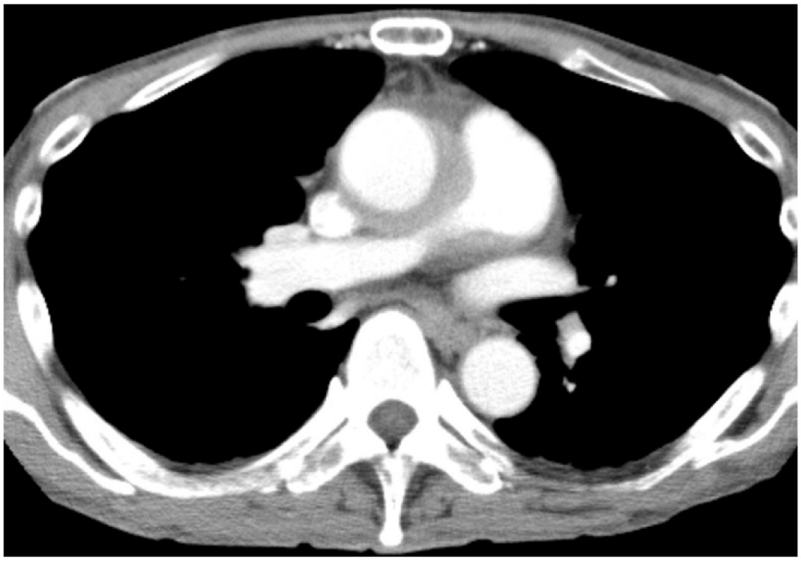


Fig. 2. Computed tomography (a) revealed the huge soft density mass around the aortic arch. Inflammation more strongly surrounded cervical branches (a) and arch (b). The mass compressed strongly pulmonary main trunk(c).

a

b

c

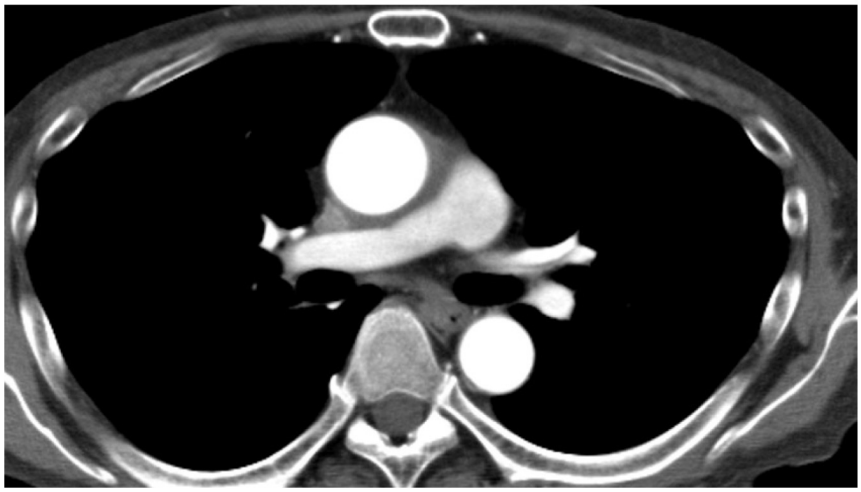


Fig. 3. Computed tomography (a, b, c) revealed that the soft density mass around the aortic arch was remarkably decreasing.

transferred back to the rehabilitation hospital. The patient consented to receiving prednisolone therapy after he was discharged from the rehabilitation hospital after two and a half months.

While visiting an outpatient department, the patient complained of shortness of breath. Echocardiography revealed low cardiac function like dilated cardiomyopathy: left ventricular internal dimension-diastole /-systole 66/59 mm, intraventricular septal thickness-diastole/left ventricular posterior wall thickness-diastole 7/5 mm, ejection fraction 21%. Blood test showed an abnormally high level of brain natriuretic peptide (BNP: 588.1 pg/ml; normal range: <18.4 pg/ml) and CRP (8.70 mg/dL). However, the leukocyte count was within normal range (WBC: 8100/mL). Enhanced CT revealed that the periaortic soft tissue density shadow had spread and presented with bilateral pulmonary artery stenosis due to compression of the mass (Fig. 2a, b, c). Furthermore, cardiac enlargement, as well as pericardial and pleural effusion was pronounced.

As the patient's condition continued to deteriorate, he was admitted once more for further examination and started prednisolone treatment. We determined the route of prednisolone administration based on guidelines on aortitis syndrome. As we decided that his inflammatory state was to a moderate degree, we chose not to administer a bolus dose, which was 1 to 2 mg per 1 kg in body weight per 1 day, but decided instead on a middle equivalence administration, which was 0.4 to 0.9 mg prednisolone per 1 kg in body weight per day. As such, we started the patient on 45 mg prednisolone/ day given that his body weight was 55 kg. We confirmed that there were no side effects after starting the patient on prednisolone. Furthermore, CT revealed a reduction in inflammation around the aortic arch after the 14th day of initial prednisolone treatment (Fig. 3a, b, c). Moreover, the dilated cardiomyopathy was due to heart failure so the patient was also started on a beta-blocking agent without any complications. The patient was discharged from the hospital after three weeks.

One month later, the patient's blood test results tremendously improved: CRP, WBC, and BNP were 0.06 mg/dL, 14400/mL, and 20.3 pg/ml respectively. However, the high leukocyte value reflected the effects of the prednisolone and did not worsen the inflammation of the periaortitis of the aortic arch. As his general condition was good apart from having a moon-like facies, we continued prednisolone at 45 mg/ day. After six weeks the CRP levels remained at 0.06 mg/dL and we reduced the dose of prednisolone to 40 mg/ day. Two weeks later, CT findings improved markedly. We thus further reduced the dose of prednisolone to 35 mg/day. With reference to the inflammatory reaction and CT findings, we gradually reduced the prednisolone dose to a final dose of 5 mg/day before stopping treatment completely after 46 weeks. He has been visiting the outpatient clinic every month, and remained free of the inflammation of the periaortitis of the aortic arch for at least five years.

Discussion

Periaortitis is not commonly viewed as both an autoimmune disease as well as an aortitis syndrome [1–4]. Gallium (Ga) scintigram or positron emission tomography (PET) CT is a useful modality to prove the inflammation around the aorta and its branches [2–4]. In this case report, we should have made a diagnosis of periaortitis with such a modality. However, we thought this patient had a periaortitis at the aortic arch, and we used prednisolone administration as a treatment strategy that is usually prescribed for aortitis syndrome [5,6]. Many case reports have described an improvement of clinical manifestations, reduction of inflammation and aortic aneurysm size following prednisolone administration [2,3,6,7]. However, there is much uncertainty regarding both duration and dose of prednisolone administration.

While there have been extensive studies on chronic periaortitis of the abdominal aorta, few cases have reported periaortitis of the thoracic aorta, especially at the aortic arch without aneurysm [7–9]. Hayıroğlu et al. [6] reported a case who was admitted to an emergency department with chest pain due to idiopathic ascending aortitis, diagnosed without Ga scintigram or PET CT. In the abdominal aorta region, both inflammatory abdominal aortic aneurysm and idiopathic retroperitoneal fibrosis (IRF) belong to the same disease groups as chronic periaortitis [2–4]. Among chronic periaortitis, one of the pathophysiological mechanisms was IgG4-mediated. According to Kamisawa et al. [8] 4% of thoracic aorta aneurysms had IgG4. One of the diagnostic criteria has an 'aortic mantle sign' visible on enhanced CT. This sign illustrates the lateral contrasted adventitia which is thickened and surrounds the aorta. This finding is also consistent for the histological characteristic [1–3,7,8]. According to a retrospective study, Palmisano et al. [7] documented 153 patients with chronic periaortitis, only 15 patients had thoracic periaortitis. Mizushima et al. [9] reported the clinical course after corticosteroid therapy in an IgG4-related chronic periaortitis. They detailed every steroid dose for all 40 patients. Only two patients had also thoracic aorta as well as abdominal aorta, both of them were diagnosed with definite IgG4-related diseases. In this case, we should have included in the differential diagnoses an acute aortic dissection with thrombosed false lumen or a ruptured thoracic aorta aneurysm with infection. Although we were unable to obtain an arterial wall biopsy sample, we assumed periaortitis of the aortic arch from enhanced CT image findings and clinical manifestations. We then proceeded to administer prednisolone and successfully completed treatment. Therefore, we believe that it is useful to report our prednisolone doses used and dosing period for periaortitis of the aortic arch [5–9]. Not only did we begin initial prednisolone treatment at 0.9 mg/ kg/day, but also performed a downward titration after only half a year. Prednisolone use is indicated in cases of periaortitis without aneurysmal diameter expansion. In cases with expansion, an extremely careful follow-up including vascular surgical intervention is essential. The follow-up must include a hematological inflammatory score as well as CT imaging.

Conclusion

We experienced periaortitis in the aortic arch. We began 45 mg prednisolone/day at the beginning, and we performed downward titration after only half of a year.

Disclosures

The authors declare no conflicts of interest.

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