Unsuspected Cardiovascular Involvement in Relapsing Polychondritis

A Case of Aortitis With Critical Coronary Artery Stenosis Secondary to Relapsing Polychondritis

Gavin Sugrue, MD; Laura Durcan, MD; Leann Bell, MD; Ferdia Bolster, MD; Suzanne Donnelly, MD; Eoin Kavanagh, MD

A 51-year-old man presented with a prodrome of arthralgia, intermittent oral ulceration, and raised inflammatory markers. He was extensively investigated; however, no firm diagnosis could be established, and he remained under regular surveillance by rheumatology and infectious disease services. After 12 months, he presented with painful ear swelling. Clinical examination revealed a swollen tender ear (Figure 1) and a soft diastolic heart murmur. Serum inflammatory markers were raised, with a C-reactive protein (CRP) of 28 mg/L and an erythrocyte sedimentation rate (ESR) of 25 mm/h.

Biopsy of affected cartilage showed characteristic features of auricular chondritis (Figure 2). Echocardiogram (Movie I in the Data Supplement) revealed mild aortic incompetence, a left ventricular ejection fraction of 60%, aortic root dilatation, and an incidental patent foramen ovale (Movie II in the Data Supplement). Computed tomography (CT) of the thorax demonstrated no tracheal abnormality; however, aortic root dilatation of 4.6 cm was noted (Figure 3). A fast-positron emission tomography-CT (PET-CT) showed increased F-18 fluorodeoxyglucose accumulation in the ascending aorta (Figure 4), and a diagnosis of relapsing polychondritis with secondary aortitis with structural aortic dilatation was made.

Therapy with oral prednisone 1 mg/kg and oral methotrexate (25 mg weekly) was commenced immediately. Inflammatory markers failed to settle after 4 months of therapy, and a new femoral bruit was heard, suggesting clinical disease progression that was confirmed on ultrasound scan. The anti–tumor necrosis factor agent infliximab (intravenous infusion 5 mg/kg 6 weekly) was added to therapy, and oral prednisolone was reduced to <10 mg/d. The patient maintained in the normal range (CRP, 8 mg/L; ESR, 10 mm/h) and the CRP and ESR were maintained in the normal range (CRP, 8 mg/L; ESR, 10 mm/h) and oral prednisolone was reduced to <10 mg/d. The patient remains asymptomatic.

Relapsing polychondritis is a rare inflammatory condition (reported incidence 3.5 cases/million) affecting cartilaginous tissues such as the ears, nose, and trachea. Diagnostic criteria (McAdam Criteria) for relapsing polychondritis requires ≥3 of the following clinical features without biopsy confirmation:1

- Recurrent chondritis of both auricles
- Nonerosive inflammatory polyarthritis
- Chondritis of nose cartilage
- Inflammation of ocular structures—keratitis, scleritis, episcleritis, and uveitis
- Chondritis of the respiratory tract—laryngeal and tracheal cartilages
- Cochlear and vestibular damage causing sensor neural hearing loss, tinnitus, and vertigo.

However, the diagnosis is definitive when ≥1 of the above criteria is met in conjunction with biopsy confirmation.2 The patient fulfilled this criteria based on biopsy highlighting...
auricular chondritis in conjunction with recurrent auricular chondritis and nonerosive inflammatory polyarthritis.

Cardiovascular involvement is a rare complication of this rare condition; aortic involvement in the form of aortic aneurysm and aortic insufficiency is encountered in 4% to 6% of patients with relapsing polychondritis and is associated with significant mortality. The finding of aortitis with associated coronary ostial lesion is exceedingly rare: there are only 5 cases reported in the literature. Coronary ostial stenosis results from contiguous aortic inflammation that promotes intimal proliferation and luminal contraction in the ascending aorta around the coronary ostia.

The diagnosis of aortic root dilatation secondary to aortitis in this case was made definitively on PET-CT findings. PET-CT is now considered to be a robust clinical tool in the diagnosis of steroid naïve patients with large vessel vasculitis. It has advantages compared with current imaging modalities such as CT, angiography, and MRI because it identifies early inflammatory changes before the development of structural changes, facilitating the introduction of therapeutic agents with the potential to prevent tissue damage and significant morbidity and mortality.

This case of aortitis with critical coronary artery stenosis demonstrates an extremely rare cardiovascular complication of the rare condition relapsing polychondritis. We highlight the clinical use of PET-CT in diagnosing and monitoring progressive inflammation of the aorta in an asymptomatic patient with persistently raised inflammatory markers (CRP >10 mg/L) for whom there was a strong clinical suspicion of disease activity, despite high levels of immunosuppression including a biological agent. In this case, interval PET-CT was key to therapeutic decision making, and we note that CRP settled on high-dose infliximab plus methotrexate and steroid, providing further evidence for biological therapies as effective treatment options in refractory aortitis. In summary, PET-CT was an important diagnostic and therapeutic monitoring tool that guided therapeutic decision making in an extremely rare clinical context.

Disclosures

None.

References


KEY WORDS: biological therapy • polychondritis, relapsing

Figure 1. Tender, erythematous left ear.

Figure 2. Left ear biopsy identified cartilage with perichondrial inflammatory granulation tissue.

Figure 3. Coronal computed tomography of thorax with contrast demonstrating aortic root dilatation of 4.6 cm at the level of the sinuses with mural thickening at the aortic root. The aortic root wall measures ≤9 mm in thickness. The left ventricle is mildly dilated likely secondary to aortic incompetence. The ascending aorta is borderline enlarged at 3.9 cm.
Figure 4. Initial fasting 2-dimensional positron emission tomography–computed tomographic study performed from skull base to proximal thighs after intravenous administration of 348 MBq of F-18 fluorodeoxyglucose (FDG). Circumferential FDG accumulation at the aortic root with a maximum standardized uptake value of 2.30 consistent with a diagnosis of aortitis.

Figure 5. Coronary angiogram demonstrating 90% ostial stenosis of the left main stem (yellow arrow, A). A stent is deployed over the stenosis (black arrow, B) with successful positioning (white arrow, C).

Figure 6. A, Follow-up fasting 2-dimensional positron emission tomography–computed tomographic (PET-CT) study from skull base to proximal thighs after intravenous administration of 348 MBq of F-18 fluorodeoxyglucose (FDG) performed 3 years after initial diagnosis. B, Fused FDG PET-CT. Both A and B demonstrate increased FDG activity in the aortic root with maximum SUV of 5.0 (previously 2.3). New FDG activity is seen in the left atrium with maximum SUV of 7.1, and new patchy FDG activity is seen in the descending abdominal aorta with maximum SUV of 4.0.
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_Circ Cardiovasc Imaging_. 2014;7:409-411
doi: 10.1161/CIRCIMAGING.113.001290

_Circulation: Cardiovascular Imaging_ is published by the American Heart Association, 7272 Greenville Avenue, Dallas, TX 75231
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Print ISSN: 1941-9651. Online ISSN: 1942-0080

The online version of this article, along with updated information and services, is located on the World Wide Web at:
http://circimaging.ahajournals.org/content/7/2/409

Data Supplement (unedited) at:
http://circimaging.ahajournals.org/content/suppl/2014/03/18/CIRCIMAGING.113.001290.DC1

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**Video 1:** Transthoracic ECHO demonstrating aortic incompetence in the parasternal long axis view.

**Video 2:** Transesophageal ECHO demonstrating right to left flow of agitated saline across a patent foramen ovale.

**Video 3:** Coronary angiogram demonstrating high grade ostial stenosis of the left main stem and successful deployment of a stent over the ostial stenosis.