Case report

Extra-cranial giant cell arteritis: a diagnostic challenge

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Learning Point for Clinicians

Giant cell arteritis often affects extra-cranial vessels and may not present as a classical cranial arteritis. It must be considered in patients over 50 with constitutional symptoms and raised inflammatory markers. Initial investigations are usually non-specific, but positron emission tomography–computed tomography is normally diagnostic. Prompt treatment with corticosteroids is required for this potentially life-threatening condition.

Case 1

A 61-year-old female was referred to our clinic with a 6-week history of fatigue, weight loss and fevers. She also reported epigastric and bilateral buttock pain. She had no medical history and took no medication. Examination revealed epigastric and left groin tenderness without peritonism. Peripheral pulses were intact. Temporal arteries were non-tender and the remaining examination was normal.

Initial blood tests revealed a normocytic anaemia (haemoglobin (Hb) 9.1 g/dl), white cell count (WCC) 7.5 x 109/l and platelets (Plts) 331 x 109/l. Inflammatory markers were elevated: erythrocyte sedimentation rate (ESR) 103 mm/h and C-reactive protein (CRP) 118 mg/l and alkaline phosphatase was raised at 418 U/l; other liver function tests (LFTs) and renal function were normal.

Autoimmune serology, serial sets of blood cultures and urinalysis were all negative. Computerized tomography scan of the chest, abdomen and pelvis (CT CAP) was unremarkable; however, subsequent positron emission tomography–computed tomography (PET-CT) scan demonstrated an extensive aortitis (Figure 1a) consistent with large-vessel vasculitis and a diagnosis of giant cell arteritis (GCA) was made.

She was treated with 60 mg prednisolone once daily and methotrexate 15 mg once weekly. Symptoms improved after 3 days with normalization of CRP by week four.

Case 2

A 78-year-old female was referred with a 2-month history of anorexia, fatigue, night sweats and weight loss. She was breathless on exertion and had swollen ankles. She had no past medical history and took no medications.

On examination, she was afebrile. She had finger-nail splinter haemorrhages. Cardiovascular examination revealed an ejection systolic murmur and unequal brachial blood pressures (right 129/62 mmHg, left 107/56 mmHg). Peripheral pulses were intact and temporal arteries pulsatile. She had bilateral pitting oedema with bruising at the ankles.

Initial blood tests demonstrated a normocytic anaemia (Hb 86 g/dl), WCC 8.1 x 109/l and a thrombocytosis: Plts 571 x 109/l. Urea and creatinine...
were 6.1 and 80, respectively. Other than a hypoalbuminaemia (27 g/l) all other LFTs were unremarkable, and international normalized ratio and prothrombin time were also normal. Inflammatory markers were raised: CRP 139 mg/l and ESR 117 mm/h. ANA, proteinase 3 and myeloperoxidase antibodies were negative. Serial blood cultures and viral serology were also negative, and

Figure 1. (a). PET-CT scan demonstrated fluorodeoxyglucose (FDG) uptake within the wall of the aorta extending to the common iliac, subclavian and carotid arteries. A magnified image focusing on the heart, ascending aorta and carotid arteries is shown in the right upper region of the figure. (b). PET-CT scan showed FDG uptake in the aortic wall with involvement of the carotid and subclavian arteries as well as the coeliac axis and superior mesenteric artery. A magnified image focusing on the heart and ascending aorta is shown in the right upper region of the figure, and a magnified image of the carotid arteries, subclavian arteries and descending aorta are shown in the right lower region of the figure (Increased FDG uptake indicated in red (online version) and light grey (print copy)).
electrocardiogram and transthoracic echocardiogram were unremarkable. CT CAP revealed small pericardial and pleural effusions. A subsequent PET-CT scan demonstrated extensive aortitis (Figure 1b) consistent with GCA.

Intravenous Methylprednisolone (1 g single dose) and 60 mg prednisolone daily were initiated. Her symptoms improved and inflammatory markers normalized at 2 weeks. However, she began developing petechiae on her abdomen and ecchymoses on all limbs. Blood tests demonstrated a thrombocytopenia (Plts 20 × 10^9/l) and bone marrow aspirate confirmed an associated immune mediated thrombocytopenia requiring platelet transfusion.

Discussion

GCA is a large-vessel vasculitis seen in patients over 50-years-old and is the commonest primary systemic vasculitis. Classically it presents as a cranial arteritis, with headache, visual disturbance and jaw claudication. Temporal artery biopsy can be diagnostic, but is often negative. Importantly, up to 40% of patients have no cranial features, but instead present with non-specific constitutional symptoms. This reflects the systemic inflammatory response associated with the large-vessel inflammation seen in extra-cranial GCA. Our cases highlight the importance of considering extra-cranial GCA when assessing patients with constitutional symptoms and an acute inflammatory blood profile. In such cases, the general physician will usually consider infection and malignancy. We feel that GCA is an important additional differential and PET-CT scanning should be considered if routine investigations are unrevealing. Furthermore, our cases highlight that systemic vasculitis should not be dismissed in the absence of the cutaneous, renal and neurological features commonly associated with ANCA-positive vasculitides.

Conflict of interest: None declared.

References