43. LARGE VESSEL VASCULITIS AND SARCOIDOSIS: CO-EXISTENCE OR ONE DISEASE?

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Introduction: Takayasu arteritis (TA) is a large-vessel vasculitis that preferentially affects the aorta and its major branches, is rare, predominately affects women of child-bearing age and its precise aetiology is unknown. TA causes chronic vascular inflammation. Sarcoïdosis, too, is a systemic inflammatory condition which can affect any organ system; the pulmonary system is the most common site. Large-vessel vasculitis is rare in sarcoidosis, but overlap between the two conditions has been reported. It is unclear whether they co-exist or manifest as one disease entity. We report a case of a 50-year-old lady with pulmonary sarcoidosis on a background of TA.

Case description: A 40-year-old female presented in 2010 with constitutional symptoms, erythema nodosum (confirmed on biopsy), audible murmurs over her carotid and subclavian arteries and raised inflammatory markers (CRP 100). She was diagnosed with Takayasu arteritis following CT angiogram which demonstrated perilaterial cuffing and thickening of her carotids, subclavian and thoracic aorta. Her medical history consist of pericarditis in 1992, a thromboembolic event in 1995, ulcerative keratitis in 2006 and incidental aortic regurgitation in 2009. She was treated with oral corticosteroids and started on azathioprine as a steroid sparing agent. Inflammatory markers normalised. Further cardiology assessments confirmed evidence of a dilated ascending aorta in 2015 and she was also diagnosed with corneal ulceration in September 2016.

In July 2017, intermittent ankle swelling was reported which was associated with mildly raised inflammatory markers (CRP of 12, ESR of 27). Accentuating murmurs noted and in view of raised inflammatory markers, CT angiogram was repeated; that showed stable appearances of TA. In May 2018, her azathioprine was reduced to 100mg from 125mg as she remained clinically and radiologically stable. In July 2018, she reported recurrence of night sweats and she had marginally raised CRP of 7 and ESR of 8. PET-CT, to look for active TA, demonstrated high uptake on bilateral mediastinal lymph nodes and no evidence of active TA. It was noted retrospectively that mediastinal lymphadenopathy was present on her CT back in 2017. She then underwent endobronchial ultrasound bronchoscopy in August 2018 which showed reactive lymph nodes. Other potential causes were excluded by extensive microbiological and immunology studies. Mediastinoscopy and lymph node excision was arranged as a lymphoproliferative/infective disease needed to be excluded in view of prolonged immunosuppression. Biopsy supported the diagnosis of sarcoidosis showing granulomatous changes. Oral prednisolone 40mg initiated and azathioprine was increased to 125mg. ACE levels remained normal.

Discussion: This case report emphasises the need for consideration of other systemic conditions in patients with known inflammatory diseases as they can co-exist. Patients who are presented with symptoms that are not fully consistent with a specific phenotype of a disease as in this case the ocular symptoms (corneal ulceration, ulcerative keratitis) and the erythema nodosum, could raise the possibility of a different or co-existent disease.
It does also suggest that the prevalence of TA, or related forms of arteritis, may be higher than expected and should be considered, especially in younger patients with non-characteristic cardiovascular symptoms and suspected systemic inflammatory disease. Moreover, the association with sarcoidosis in this and other previously described cases suggests that the two diseases may be related, and that TA or TA-like vasculitis may even be a complication of sarcoidosis. Other causes of large vessel vasculitis should be excluded as TB and lymphoproliferative diseases which can also present with lymphadenopathy especially as it is well known that large vessel vasculitis, especially in elderly population, could be part of a para-neoplastic syndrome. Other diseases have been reported associated with TA but rarely sarcoidosis. TA and sarcoidosis may be related as they are characterized by certain nonspecific immunoinflammatory abnormalities. In most case reports sarcoidosis precedes TA diagnosis. In this case, TA was found 9 years before the diagnosis of sarcoidosis was made.

Key learning points: TA can precede the diagnosis of sarcoidosis. In case of relapsing or refractory TA, further investigations should be considered to exclude other co-existent pathologies as sarcoidosis. TA and sarcoidosis may be related as they are characterized by certain nonspecific immunoinflammatory abnormalities. It has been reported that TA stands as pathology-associated with sarcoidosis. Complete vascular clinical examination should be performed to detect inflammatory arteritis, especially in cured sarcoidosis presenting a relapse of the biological inflammatory process.

Conflicts of interest: The authors have declared no conflicts of interest.