Case Reports in Clinical Medicine, 2014, 3, 457-459
Published Online August 2014 in SciRes. http://www.scirp.org/journal/crcm
http://dx.doi.org/10.4236/crcm.2014.38100

Syphilitic Aortitis Mimicking Takayasu's Arteritis

Anna Sophia Pörings[1], Bernd Salzberger[2], Lothar Veits[3], Boris Ehrenstein[1], Wolfgang Hartung[1], Martin Fleck[1,2]

[1]Department of Rheumatology and Clinical Immunology, Asklepios Medical Center, Bad Abbach, Germany
[2]Department of Internal Medicine I, University Medical Center, Regensburg, Germany
[3]Department ofPathology, Klinikum Bayreuth, Bayreuth, Germany
Email: annapoerings@web.de

Received 29 May 2014; revised 28 June 2014; accepted 25 July 2014

Abstract

SIR, Syphilis is a sexually-transmitted infectious disease caused by the bacteria Treponemapallidum. A characteristic manifestation of the third stage is arteritis of the aorta, which leads to necrosis and loss of tissue. Here, we report on a 48-year-old woman admitted with suspected Takayasu's arteritis (TA) due to localized inflammation confined to the aortic root and valve, which could be diagnosed as syphilitic aortitis.

Keywords

Vasculitis, Syphilis, Takayasu's Arteritis

1. Letter to the Editor

A 48-year-old patient was admitted to our tertiary Rheumatology center after composite aortic valve graft replacement had been performed the month before due to 70% stenosis of the aorta combined with an aortic valve insufficiency. The patient had developed 4 months prior to admission rapid signs of cardial decompensation. The pathohistological analysis of the resected tissue revealed aortitis with lympho-plasma cell rich infiltrations around vasa vasorum and lymphatic aggregates in the adventitia (**Figure 1**). In addition, there was a thickened vessel wall with slight fibredesorganisation and intramural lamelliform necrosis of the media with sparse neutrophilic granulocytes, lymphocytes and plasma cells (**Figure 2**). Immunohistochemical studies including stainings for CD68, IgG4 and CD138 did not provide additional information. Off note, there were no granulomas present. In light of these histopathological findings, infectious aortitis as well as localized idiopathic aortitis were considered by the pathologists, however, no information regarding an infection has been provided on request

How to cite this paper: Pörings, A.S., Salzberger, B., Veits, L., Ehrenstein, B., Hartung, W. and Fleck, M. (2014) Syphilitic Aortitis Mimicking Takayasu's Arteritis. *Case Reports in Clinical Medicine*, **3**, 457-459.
http://dx.doi.org/10.4236/crcm.2014.38100

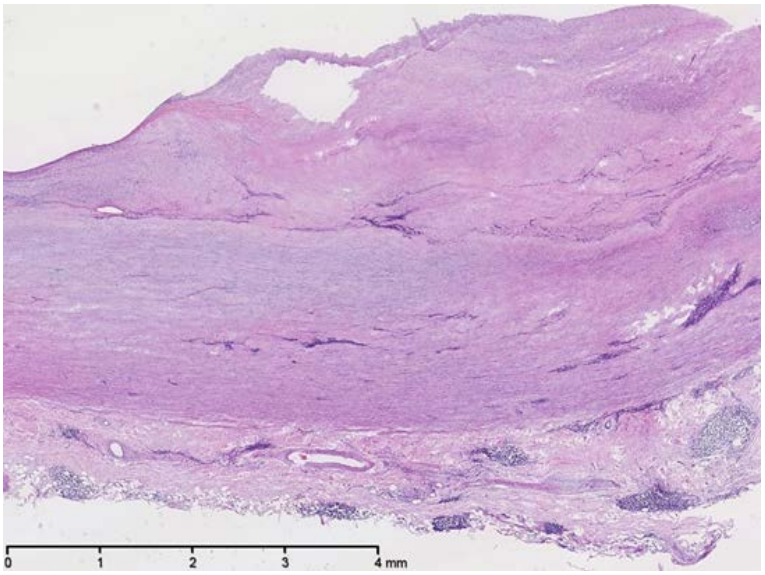

Figure 1. Section of the aorta with lymphatic aggregates near by the vasa vasorum of the adventitia and a thickened intima.

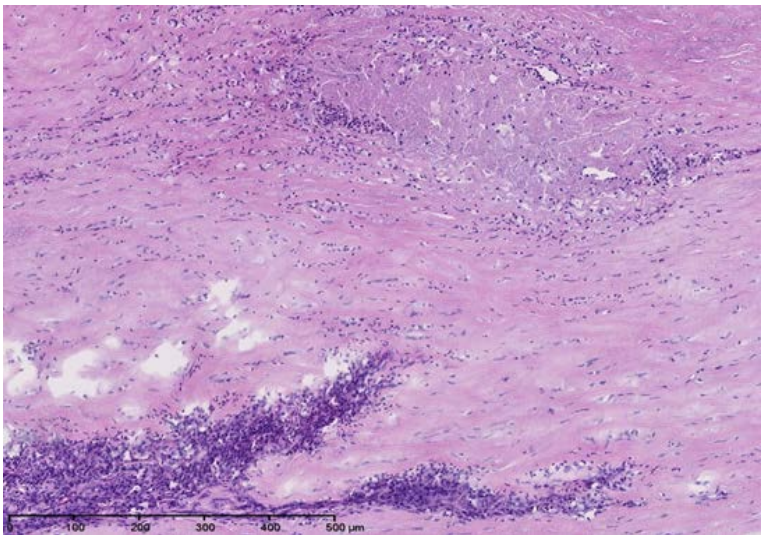

Figure 2. Image enlargement of the media with fibre desorganisation and lamelliform necrosis.

by the treating clinicians.

Due to the clinical presentation and histological findings, manifestated TA had been suspected by the treating surgeons, and therefore immunosuppression with 20 mg prednisolone per day was initiated in addition to therapeutic anticoagulation, and the patient was admitted to our hospital for further evaluation.

On admission the patient presented in good general condition with no relevant findings in the physical examination. An elevated INR of 2, 3 due to anticoagulation was observed, and all other routine parameters were unremarkable. Vascular ultrasonography was performed of the *A. carotis*, *A. axillaris* and *A. femoralis* of both sides without any abnormal findings. In addition, multiple immune parameters (ANA, ANCA, RF, ACPA) as well as bacterial serologies (TPHA, Borreliaburgdorferi) were analyzed. Surprisingly, TPHA testing revealed a very high titer of 1:10240. Confirmatory tests were performed additionally demonstrating the presence of IgG and IgM specific for Treponapallidum at high titers. Together with the histological findings the final diagnosis of syphilitic aortitis could be established reflecting Syphilis at stage III [1].

The drug of choice to treat syphilis stage III is Penicillin G that was initially administered as a bolus therapy of two mega penicillin G in 50 ml in 30 minutes. The following two weeks the patient was treated with a continous infusion of 50 mega Penicillin G. The reason for this treatment regimen was to secure a robust concentration of Penicillin G and to avoid repeated intramuscular injections due to concomitant therapeutic anticoagulation [2] [3].

With regard to the inflammatory aspects observed macroscopically and the histological findings of an inflammatory aortitis the diagnosis of TA has been initially suspected and consequently immunosuppression was initiated [4] [5]. TA is a rare disease—the incidence observed in a study of North American patients was calculated at 2.6/million/year, and affects most commonly young women [6] [7]. Generally, this disease presents with an inflammation of the vasa vasorum of the adventitia in the acute phase. This leads to a lymphocytic infiltrate of the media and a thickening of the intima due to proliferation of fibroblasts and smooth muscle cells. The chronic phase of TA is characterized by the destruction of the elastic tissue, which leads to fibrosis and ultimately stenosis. In addition, manifestation of aneurysms might occur complicating the disease course [4] [6] [7]. Similar lesions can be observed in syphilitic aortitis with endarteritis obliterans and concomitant infiltration of plasma cells and vessel wall fibrosis challenging the diagnosis. However, a histological feature characteristic of syphilitic aortitis is necrosis of the media, which could be observed in our patient [4] [5]. In contrast to TA, which is a rare disease in Western Europe, the numbers of Syphilis cases in Germany are increasing. The patient numbers increased 2011 from 3033 infections to 3698 in the following year, which results in an incidence rate of 4.5/100.000/year [8].

The histological report of our patient mentioned a remoulding of the elastic fibres, lymphocyte infiltration and necrosis of the media, whereas the immunohistochemistry was inconspicuous. Regardless of the histological findings supporting an infectious etiology over TA, our patient did not suffer from the typical symptoms of TA including weakening of the brachial artery pulses, claudication or fatigue. In addition, the age of our patient did not match very well with the predominant manifestation age.

In summary, rheumatologists should have a high level of vigilance to exclude syphilitic aortitis in patients with suspected TA presenting with atypical or inconsistent findings prior to the establishment of immunosuppressive therapy.

2. Rheumatology Key Message

Syphilitic aortitis is an important differential diagnosis to TA and should be excluded in patients with suspected TA presenting with atypical findings.

References

[1] Ho, E.L. and Lukehart, S.A. (2011) Syphilis: Using Modern Approaches to Understand an Old Disease. *Journal of Clinical Investigation*, **121**, 4584-4592. http://dx.doi.org/10.1172/JCI57173

[2] Mattei, P.L., Beachkofsky, T.M., Gilson, R.T. and Wisco, O.J. (2012) Syphilis: A Reemerging Infection. *American Family Physician*, **86**, 433-440.

[3] Janier, M., Libar, E., Bonnet, A., Meunier, P., Tabet, M., Mathourais, M., Paterour, C. and Porcher, R. (2012) Treatment of Late Syphilis with 2.4 Million Units Benzathine Penicillin G (BPG): Tolerance of Single versus Divided Doses. *Sexually Transmitted Diseases*, **39**, 359-360. http://dx.doi.org/10.1097/OLQ.0b013e318249968c

[4] Tavora, F. and Burke, A. (2006) Review of Isolated Ascending Aortitis: Differential Diagnosis, Including Syphilitic, Takayasu's and Giant Cell Aortitis. *Pathology*, **38**, 302-308. http://dx.doi.org/10.1080/00313020600820898

[5] Johnston, S.L., Lock, R.J. and Gompels, M.M. (2002) Takayasu Arteritis: A Review. *Journal of Clinical Pathology*, **55**, 481-486. http://dx.doi.org/10.1136/jcp.55.7.481

[6] Hall, S., Barr, W., Lie, J.T., *et al.* (1985) Takayasu Arteritis. A Study of 32 North American Patients. *Medicine*, **64**, 89-99.

[7] Keser, G., Direskeneli, H. and Aksu, K. (2013) Management of Takayasu Arteritis: A Systematic Review. *Rheumatology*. [Epub ahead of print]

[8] Robert Koch Institut (2012) Epidemiologisches Bulletin: Erneuter Anstieg der Syphilis-Meldung in 2011, 18.Juni 2012/Nr. 24.

Scientific Research Publishing (SCIRP) is one of the largest Open Access journal publishers. It is currently publishing more than 200 open access, online, peer-reviewed journals covering a wide range of academic disciplines. SCIRP serves the worldwide academic communities and contributes to the progress and application of science with its publication.

Other selected journals from SCIRP are listed as below. Submit your manuscript to us via either submit@scirp.org or Online Submission Portal.