

Aortic aneurysm in a patient with syphilis-related spinal pain and paraplegia

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Abstract

The tertiary stage of syphilis is nowadays extremely rare, showing predilection for the cardiovascular and nervous systems.

A 57-year-old Caucasian man sought medical assistance due to back pain that evolved to paraplegia of the lower limbs. A thoracic CT scan demonstrated an important aneurysmatic lesion of the descending thoracic aorta causing erosion of the vertebral bodies and VDRL and FTA-abs positivity. Although rare, syphilitic aortitis, the hallmark of cardiovascular syphilis, should be considered in the differential diagnosis in patients with thoracic aneurysm when in the absence of classic risk factors for atherosclerosis, especially in cases that progress with erosion of vertebral bodies.

Key words: back pain, tertiary syphilis, syphilitic aneurysm.

Introduction

Back pain is one of the most common reasons for visiting a clinician, but less than 5% will have a serious underlying systemic pathology such as infection or malignancy [1].

Syphilis is a disease known for a long time, which peaked during the Second World War, with a swift decline after the arising of penicillin, but it remains one of the most prevalent sexually transmitted diseases [2]. It is a chronic systemic affection originated by *Treponema pallidum* infection, which may be either congenitally or sexually transmitted, alternating activity and latency periods during the time span of the infection.

Due to widespread use of antibiotics, the tertiary stage of syphilis is currently extremely rare, being the cardiovascular and central nervous systems the most commonly affected sites, even if asymptomatic [2–4]. Among the cardiovascular complications caused by tertiary syphilis, aortitis is the most common, in which 10% of patients develop meaningful critical complications [5].

In non-treated syphilis, aortitis occurs in over 70% of cases, with such symptoms as aortic regurgitation, stenosis, and aortic aneurysm occurring in up to 15% of patients [2, 5–7]. Classically, syphilitic aneurysms occur in 90% of cases on the thoracic aorta, and in 10% in the abdominal aorta [3, 7–9]. Infection of aortic wall develops during the secondary or bacteraemic phase of syphilis, having a latent period from infection until the clinical presentation ranging from 5 to 50 years [5, 8].

For unknown reasons *T. pallidum*, after infecting the body and spreading through vascular dissemination, may cause an inflammatory process in the *vasa vasorum* in the inner layer of the aorta leading to strictures. There is also obstruction due to granuloma formation and focal necrosis of the elastic and muscular layers and substitution for fibrotic tissue that afterwards turns into calcification (syphilitic aorta) [10, 11]. This process may carry on and generate extreme strictures with saccular or fusiform aneurysms [8–10]. Due to intense scarring of the media layer, rupture resulting from a dissection is highly unlikely to occur, as opposed as non-syphilitic

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aortic aneurysms, in which the rupture is the main cause of death.

Case report

A 57-year-old Caucasian man, living in the countryside, sought medical assistance referring to back pain that had started 6 months before, with progressive worsening in the last 45 days, until it became excruciating, disabling the patient from performing minimal tasks. The clinical picture had evolved in a 7-day time span with symmetrical paraesthesia and subsequently paraplegia on lower limbs. He denied weight loss or other constitutional symptoms. As past history, he referred to unprotected sexual activity during his youth. In the physical examination, we found pain to minimal movement and light touch of vertebral bodies and aortic holosystolic murmur without fremitus. During the investigation, a thoracic CT scan was performed, which demonstrated an important aneurysmatic lesion of the descending thoracic aorta causing erosion of the vertebral bodies corresponding to T4 to T7 (Fig. 1). It also showed an important cardiomegaly and bilateral pleural effusion. The haemodynamic study diagnosed a severe aortic valve insufficiency and a descending aorta aneurysm of the thoracoabdominal portion. The serum marker Venereal Disease Research Laboratory (VDRL) test was 1 : 64 and FTA-abs was positive. Patient was referred for surgical treatment; unfortunately, the lesion had a spontaneous rupture before any action was taken. Due to the known cause of death, autopsy was not performed.

Discussion

We describe here a rare case of a huge pulsatile syphilitic aortic aneurysm presenting with back pain as the first manifestation and evolving with neurological dysfunction due to osteolytic vertebral erosion.

During the first half of the 20th century, thoracic aortic aneurysms were much more common than their abdominal counterparts, due to the prevalence of syphilitic thoracic aortic aneurysms [3, 11]. In 1952 the thoracic/abdominal aneurysms ratio was 2 : 1, until 1964 when this ratio equalised, making the syphilitic cardiovascular involvement very rare in current days [8].

During the pre-antibiotic era, about one third of untreated syphilitic patients would develop recognisable tertiary syphilis clinically. Sclerosis initialis or *ulcus durum*, typical for first period of syphilis, may not occur or be unusual in appearance, or may be localised to a site that is not available for medical examination, but they also need to be differentiated from changes such as herpes simplex or *Haemophilus ducreyi*-induced ulcer, and the detection of diseases can occur in its subsequent stages. Confirmation of syphilis in patients with VDRL-positive results, with or without typical symptoms, is performed by FTA-ABS or TPHA.

Today, in industrialised countries, specific treatment and disseminated use of antibiotics in the treatment of other infections has almost wiped out tertiary syphilis, except for cases of neurologic involvement in HIV-infected patients [4].

The tertiary stage of syphilis is a non-contagious form of the disease, occurring any time after the secondary stage, affecting about 35% of untreated patients [12]. The tissue reactivity characterised by vasculitis and necrosis is severe and highly suggestive of hypersensitivity phenomena. In the past, the most common form of tertiary stage was the gumma, normally a benign granulomatous lesion, extremely rare nowadays.

Cardiovascular syphilis is characterised by obliterating endarteritis, which may affect small-calibre vessels irrespective of location, but it is much more devastating whenever it affects the aorta's *vasa vasorum*, jeopardising the blood supply of the aortic wall, leading to destruction of the elastic tissue and weakening of the

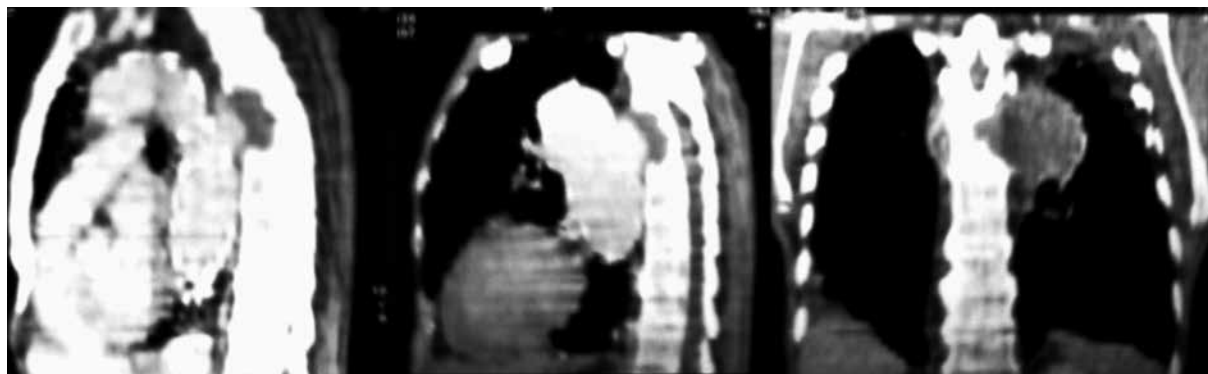


Fig. 1. Thorax CT showing an aneurysmatic lesion of the descending thoracic aorta causing erosion of the vertebral bodies corresponding to T4 to T7.

medial aortic tunic, particularly in the ascendant and transverse segments of the thoracic aortic arc [3, 8–10, 12]. The macroscopic aspect is classically described as “tree bark” due to inner layer alterations and parietal calcification [7, 10, 11].

The cardiovascular syphilitic lesions become symptomatic in only 10% of untreated patients. Among the cardiovascular lesions, 85% of patients present with aortic insufficiency, coronary *ostium* stenosis in 25 to 30%, and the presence of aneurysms in 5 to 10%, 75% of these being of saccular morphology [3].

As documented in our case, a special particularity, even though rare, of the aortic aneurysms is the pulsatile destruction of nearby structures such as sternum, ribs, vertebrae, and skin [9]. Heggteit [7] analysed 100 syphilitic aortitis cases, and in only two were there vertebral bodies erosions. Leung et al. found the same lesion in 20% of syphilitic aneurysm patients [13].

The tertiary syphilis diagnosis may be difficult because clinical features may be deceptive, having strong similarities with other granulomatous diseases, and the serologic titles can be low or negative [4]. Diagnostic exams include treponaemic (FTA-abs) and non-treponaemic tests (VDRL). The VDRL sensibility depends on the level of circulating antibodies, and in tertiary syphilis it may be negative in up to 25% of patients [4]. Whilst the FTA-abs is positive in almost 100% of tertiary syphilis, in our case the diagnosis was made with the radiologic characteristics associated with the presence of either VDRL e FTA-abs.

Once the diagnosis is made, the patient should be treated with benzathine penicillin (three doses of 2.4 million/UI in a weekly basis) and the cerebrospinal fluid tested for neurologic syphilis, even though there is some controversy about the route and duration of penicillin therapy in cases of cardiovascular syphilis [6]. The aneurysm treatment is surgical and urgent whenever neurological symptoms are present, followed by antibiotic therapy.

Our case highlights that patients with back pain and increasing neurological symptoms require full evaluation and rapid treatment to avoid complications. Although the morbidity of syphilis is decreasing, clinicians must be aware that this disease may be present in people with risky sexual behaviour and is still a problem of the modern world. Furthermore, syphilis should always be investigated when there is an insidious involvement of the cardiovascular system, mainly in cases of aortitis and aortic aneurysms in patients who do not have any risk factors for atherosclerotic disease.

The authors declare no conflict of interest.

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