

# Burned out Takayasu aortitis with superimposed atherosclerosis and left subobstructive coronary ostium in young woman

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## Citation

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## Abstract

Takayasu disease or arteritis (TA) is a chronic, idiopathic, inflammatory disease that principally affects the aorta and its primary branches. The pathological changes depend on stage of the disease. The late "burned out" stage is marked by fibrous scarring, leading to intimal and adventitial thickening, which may contain dystrophic calcification. Similar histopathological findings are also seen in many other causes of aortitis. We here in report chronic "burned out" TA in 28-year-old woman with superimposed atherosclerosis and left sub obstructive coronary ostium. This patient in addition had Aortic stenosis and calcification of ascending aorta.

## INTRODUCTION

Takayasu arteritis is also known as pulseless disease, Mörterell syndrome and occlusive thromboangiopathy.<sup>1</sup> Inflammation of the vessels leads to wall thickening, fibrosis, stenosis and thrombus formation. Strong and growing evidence shows that disease associated with chronic inflammation leads to premature atherosclerosis. There are autopsy reports of atherosclerotic changes in patients with the same disease.<sup>2</sup> Increased atherosclerosis in patients with TA may be multifactorial.<sup>3</sup> Management of TA includes effective suppression of disease activity with effective control of traditional atherosclerotic risk factors. Invariably symptoms reflect end organ ischemia. The female to male ratio appears to decline from eastern Asia towards the west. TA should be considered in differential diagnosis of calcified aorta in young women, even in the absence of occlusive or stenotic lesions.

## CASE REPORT

A 28-year-old female, detected to have heart disease at the time of delivery by caesarian section. She was admitted to our hospital with class III dyspnoea. Echocardiography showed calcified and thickened aortic valve, moderate to severe aortic stenosis & grade II aortic regurgitation. Severe mitral regurgitation (2 jets noted) with perforation in the anterior mitral leaflets was noted. In addition, moderate pulmonary hypertension and grade II tricuspid regurgitation

noted. This patient underwent mitral valve repair.

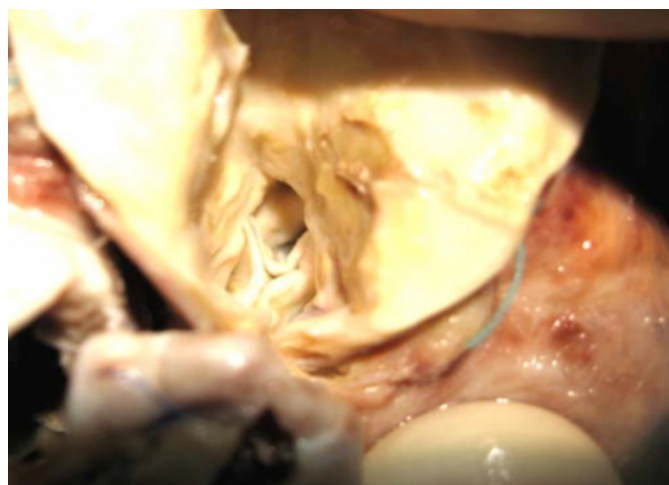
We received explanted heart. On gross examination heart looked to be small for the age. On opening, mural thrombi in right ventricle and left atrium was noted. Tricuspid valve was incompetent. Repaired mitral valve was intact. There was aortic stenosis with annular calcification extending to mitral leaflets. Right ventricle was hypertrophied. Pulmonary artery showed fatty streaks. Aorta was calcified till the arch with skip areas. Left coronary ostium showed subobstructive plaque. Myocardium showed subendocardial infarct involving mainly, left side of interventricular septum and left ventricular outlet near the aortic valve.

Histopathology of the aorta and pulmonary artery showed similar features with varying grades. Tunica intima and adventitia were thickened with fibrosis. Areas of calcification were noted in intima. The media was disorganized, replaced by collagen & patchy necrosis of media. Van-gieson stain showed distorted and fragmented elastic fibers indicative of early features of dissection. The adventitia showed congested and proliferating blood vessels, fibrosis and scattered acute inflammatory cells. The vasa vasorum showed endarteritis obliterans changes with focal chronic perivasculitis. In addition atherosclerotic plaques with lipid crystals as well as calcification were also noted. The myocardium showed areas of necrosis and foci with features of stuttering infarct. Based on these findings a

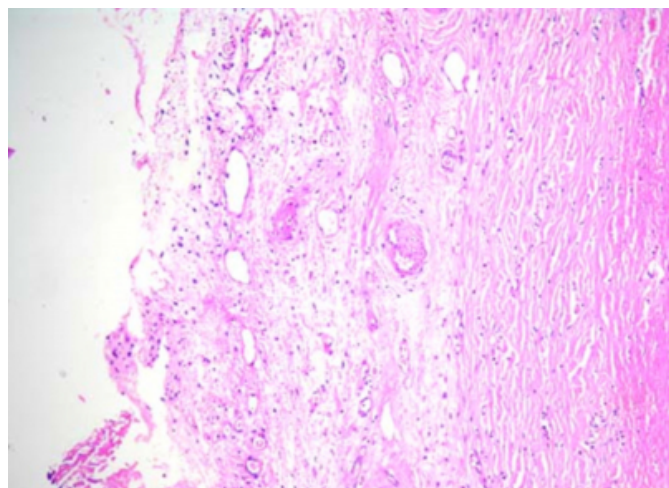
diagnosis of “burned out” Takayasu arteritis with superimposed atherosclerosis was made. The cause of death was due to subendocardial infarction due to subobstructive coronary ostium.

#### Figure 1

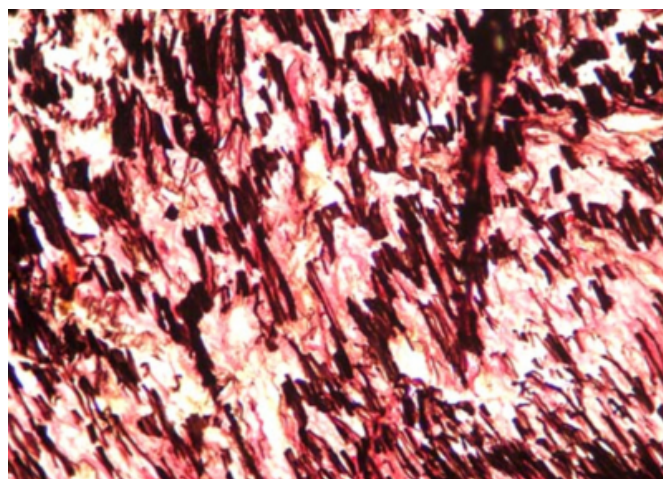
Figure 1(a) Gross photograph showing calcified aorta and aortic valve. (b) Adventitial surface of aorta showing nonspecific aortitis with endarteritis obliterans. (Lowpower view 10x, H&E). (c) Distorted and fragmented elastic fibers (Highpower view 40x, EVG). (d) Tunica media showing marked fibrosis (Highpower view 40x, Masson's Trichrome).



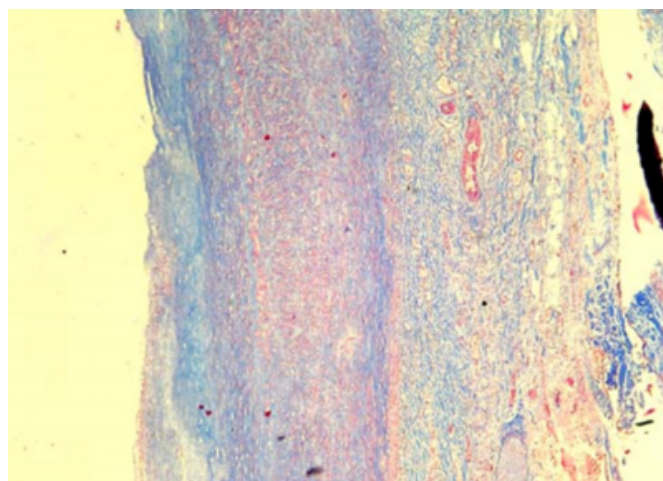
#### Figure 2



#### Figure 3



#### Figure 4



### DISCUSSION

American College of Rheumatology (ACR) defined specific diagnostic criteria for this disorder in 1990.<sup>4</sup> Still date angiography remains the gold standard for diagnosis. Pulmonary artery can also be involved in the disease process, however assessment of pulmonary vasculature by angiography is not universally recommended. It's being reserved for patients with symptoms of pulmonary hypertension.<sup>5</sup> Histological assessment is limited to those causes undergoing revascularization procedures.

Histologically, the inflammatory changes in TA vary according to stage of the disease.<sup>6</sup> The early stage is characterized by edema, patchy necrosis of the media and elastin, and chronic inflammation with infiltration by lymphocytes, plasma cells and rare giant cells in the outer third of media, adventitia and vasa vasorum. The late

“burned out” stage is characterized by marked intimal & adventitial thickening caused by fibrous scarring. Dystrophic calcification has been noted within these lesions, and eventual sequence of events leads to progressive luminal narrowing and stenosis. The thickened adventitia & its vasa vesorum exhibit patchy collection of perivascular lymphoplasmacytic infiltrate. The elastic lamina of the media are disorganized or absent, replaced by collagen. These changes predispose to aneurismal degeneration. Atherosclerotic plaques are of two types; the myohyperplastic plaque and the hypercholesterol plaque.<sup>7</sup> The inflammatory changes in TA affect the adventitia & the media, were in atherosclerosis, intimal changes are more pronounced.<sup>2</sup> In atherosclerosis adventitial inflammation shows a peculiar tropism for nervous structures related to the media at plaque level only (medial neuritis).<sup>7</sup> Increased atherosclerosis in patients with takayasu, may be multifactor. It may be associated with local or systemic disease activity, as well as with traditional atherosclerotic risk factors such as hypertension and hyperlipidemia.<sup>2</sup> An intriguing possibility also exists that it is mainly in areas where primary vessel wall disease is more prominent in Takayasu arteritis that secondary atherosclerosis is more common.

No known serological test that has been able to supplant vascular histopathology in determining disease activity.<sup>1</sup> Differentials needs to be considered are other causes of large vessel vasculitis: Inflammatory aortitis (syphilis, tuberculosis, Systemic lupus erythematosus, rheumatoid arthritis, Behcet’s disease, Kawasaki disease and giant cell arteritis); development abnormalities (coarctation of aorta & morfan syndrome) and other aortic pathologies, such as ergotism and neurofibromatosis. Most of these have specific features that enable diagnosis. Common complication includes Takyasu retinopathy, secondary aortic regurgitation and aneurysm formation. Premature atherosclerosis in Takayasu aortitis needs further clarification. Different hypothesis are proposed they include: an augmented and

sustained acute-phase response in Takayasu arteritis; with unique vessel wall involvement in this condition; with traditional atherosclerotic risk factors; or with all or any combination of the above needs to be clarified.<sup>2</sup>

## CONCLUSIONS

Takayasu should be considered in differential diagnosis calcified aorta in young women, even in absence of occlusive or stenotic lesions: and superimposed atherosclerosis needs to be looked for histopathologically. It is important for physicians to coordinate medical and surgical therapy carefully. Effective control of traditional atherosclerotic risk factors is needed, in addition to the effective suppression of disease activity, for the management of Takayasu arteritis.

## CORRESPONDENCE TO

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