JIDAM
eISSN 2582 - 0559
"An Official Journal of IDA - Madras Branch"©2019.
Available online

CASE REPORT

TAKAYASU ARTERITIS – A CASE REPORT: WITH MAXILLOFACIAL SURGEON'S PERSPECTIVE AND BRIEF REVIEW OF LITERATURE

Dr. Ramesh Srinivasan V, Dr. Priyadharsana PS, Dr. Effie Edsor, Dr.Vigneswaran T, Dr.Gowri Shankar.P*.
Department of Oral and Maxillofacial Surgery
*Department of Oral Pathology
RVS Dental College and Hospital, Coimbatore, Tamilnadu, India.

To access & cite this article
Website: jidam.idamadras.com

DOI:10.37841/jidam_2020_v7_i1_05

ABSTRACT

Takayasu arteritis is a chronic inflammatory disease that affects major vessels, mainly the aorta and its branches. The diseased condition leads to occlusion, stenosis, aneurysm or dilatations along the path of the affected artery. The etiology remains unknown. The disease has been reported worldwide, with an increased incidence in young Asian women. Like other medical condition, Takayasu arteritis patients will seek dental treatment. Hence the purpose of this article is to highlight the disease features and what a dentist should know or take precautions before treating them.

KEYWORDS: Takayasu Arteritis, Pulseless disease, Autoimmune.

Address for correspondence:

Dr. Effie Edsor, MDS,
Assistant Professor, Department of Oral and Maxillofacial Surgery,
RVS Dental College and Hospital.
Coimbatore, Tamilnadu, India.
Email id: effieedsor@gmail.com

Received – 28.01.2020
Accepted – 27.02.2020
Published – 27.03.2020

INTRODUCTION

Takayasu arteritis (TA) is also known as pulseless disease, aortoarteritis, aortic arch syndrome, idiopathic aortitis, occlusive thromboaortopathy, occlusive coagulant aortic syndrome, obstructive productive arteritis, reverse coarction or Martorell syndrome.[1,2,5] TA is a chronic inflammatory arteritis affecting large vessels, predominantly the aorta and its main branches. The most affected branches are brachiocephalic, carotid, subclavian, vertebral, renal arteries, coronary and pulmonary arteries.[3,4] Inflammatory disease leads to wall thickening, stenosis, fibrosis or thrombus formation.[5]

HISTORY

The disease is named after Mikito Takayasu, an Ophthalmologist, who reported ocular changes like aneurysms and arteriovenous anastomoses in patients with TA (1908).[1,2] The disease is named after Takayasu who first reported a patient with TA, but there are other case reports of patients with TA in literature reported earlier. In 1830, Rokushu Yamamoto described a 45-year-old man with TA with features of persistent fever who later had no pulse in the right radial artery and weak pulse in the left radial artery.[2] In 1856, Savory presented a case report of TA in 22-year-old woman, with pulseless disease in both upper extremities and left neck. He later noticed on follow-up that the patient lost her vision.[2] However, it was uncertain whether these cases truly suffered from TA.

Minoru Nakajima in 1921 compared his cases with previous reports and proposed that they should be considered as one disease. He characterized this disease by the following four criteria: (i) cases in young women affecting bilateral eyes; (ii) formation of arteriovenous anastomosis around the optic disc and microaneurysm in retinal vessels; (iii) decreased or loss of vision; and (iv) unpalpable radial artery. It was Minoru who termed it as 'Takayasu disease'.[2] Later in 1948, neurosurgeons, Kentaro Shimizu and Keiji Sano from the University of Tokyo, called the disease as 'pulseless disease'.[2]

The disease was entitled as 'occlusive coagulant aortic syndrome' by Maekawa and Kakei and 'obstructive productive arteritis' by Nasu.[2] In 1965, Riehl et al based on the pathological and immunological findings, concluded that TA is an autoimmune disease.[2] American College of Rheumatology published a classification criteria for the disease in 1990 and described it as 'Takayasu arteritis'. Although both 'Takayasu arteritis' and 'Takayasu's arteritis' are used, in the Online Mendelian Inheritance in Man (OMIM) it is registered as 'Takayasu arteritis' and this term is more commonly used.[2]

INCIDENCE

Takayasu arteritis is a rare disease with worldwide distribution, being more common in Asian population. The incidence of TA was found to be approximately 1–2 per million population across the globe.[6] Based on the recent epidemiologic study in Europe, the incidence of TA is more with an estimate varying from 0.4 to 1.5 cases per million population. The highest prevalence of TA is recorded in Japan (40 cases per million population) and the lowest in US (0.9 per million).[6] The disease is more common in female with the female to male ratio of upto 8:1.[2] The disease commonly presents in 2nd or 3rd decade of life but diagnosed in the later stage.[5]

PATHOPHYSIOLOGY

The predisposing and etiological factors of TA are still not clear. An autoimmune basis, influenced by genetic and environmental factors, is strongly suggested; the resulting inflammation is largely a cell-mediated immune response.[7] An association between the extent of vascular involvement and the major genetic risk factor HLA-B*52 was found in Turkish TA patients, suggesting that genetic factors might influences the severity of the disease. [8,9,10]

CLINICAL FEATURES

Clinical manifestations of TA are nonspecific. The most common clinical features are decreased blood pressure (BP) and feeble pulse in the upper extremities, clammy skin and numbness in the fingers.[11]

The clinical course occurs in two stages; an early active inflammatory phase and late chronic phase. The duration of active phase lasts for weeks to months and may have a remitting and relapsing course. It is characterized by systemic symptoms like fever, malaise, night sweats, loss of appetite, headaches, dizziness, loss of weight, arthralgia, skin rashes etc. The former phase does not occur in all patients, but indigenous symptoms are often seen in children with TA.[12]

The chronic phase is due to arterial stenosis and/or occlusion and ischemia of organs.[13] feeble pulses in 84-96%, vascular bruits in 80-94%, hypertension in 33-83%, retinopathy upto 37%, aortic regurgitation 20-24%, congestive cardiac failure, seizures, postural dizziness, pulmonary artery involvement.[5] Atypical presentation of homocystinuria in a TA has been recorded in 2013.[14]

DIAGNOSIS

Laboratory test for TA disease tends to be nonspecific. The erythrocyte sedimentation rate may be high, generally greater than 50 mm/h, in early disease but it is often paradoxically normal later. Leukocyte count may be normal or slightly elevated. A moderate, normochromic anaemia may be present in patients with advanced disease. Hypoalbuminemia with increased levels of C reactive protein, gamma globulin and fibrinogen are frequent findings.[15]

The American Rheumatological Society 1990 considers three of the following six criteria necessary for a definite

diagnosis of TA.[16]

1) Age of onset before 40 years
2) Claudication of extremities especially upper extremities
3) Decreased brachial artery pulse – unilaterally or bilaterally
4) BP difference > 10 mm Hg in systolic blood pressure between both arms
5) On auscultation bruit over subclavian arteries or abdominal aorta
6) Arteriogram abnormality – narrowing or occlusion

ISHIKAWA'S CRITERIA FOR THE DIAGNOSIS OF TAKAYASU'S ARTERITIS [17]

Obligation criterion

Age < 40 year (Symptoms of more than 1month duration)

Two major criteria.

Stenosis or occlusion in the
1) Left mid subclavian artery
2) Right mid subclavian artery.

Nine minor criteria.

1) Raised ESR (persistent high ESR >20 mm/h)
2) Carotid artery tenderness (Unilateral or bilateral)
3) Persistent high blood pressure > 140/90mmHg brachial or >160/90mmHg popliteal
4) Aortic regurgitation
5) Pulmonary artery lesion (Lobar or segmental arterial occlusion)
6) Lesion in left mid common carotid artery
7) Distal brachiocephalic trunk lesion
8) Descending thoracic aorta lesion
9) Abdominal aorta lesion

In addition to obligatory criterion, the presence of major criteria or of one major and two or more minor criteria or of four minor criteria suggests high probability of the disease.[17] Conventional radiographic angiography [digital subtraction angiography (DSA)] is the gold standard technique for diagnosis of TA.[18] Recently non-invasive imaging methods including magnetic resonance angiography (MRA), colour doppler ultrasound (CDU), computerized tomography angiography (CTA), PET with 18F-fluorodeoxyglucose (18F-FDG) and 18F-FDG PET/CT have recently gained ground on DSA.[19,20,21]

MANAGEMENT

Corticosteroids and conventional immunosuppressive agents such as methotrexate (MTX), azathioprine (AZA), mycophenolic acid (MMF) and leflunomide (LEF) are the most commonly used agents for TA management.[19] Biological drugs such as (TNF) tumour necrosing factor inhibitors, rituximab and tocilizumab are used in patients who are resistant to the above mentioned drugs.[19]

The frequency of ischaemic events in TA, may be decreased with the use of antiplatelet drugs.[19] A retrospective study has concluded that antiplatelet therapy was associated with decreased frequency of ischaemic events in patients with TA.[22,23]

Surgical management involves balloon angioplasty or stent graft replacement. In long standing cases, surgical bypass of the affected segment is required.[19] As a general rule, surgical intervention should be avoided during the active phase of the disease.[19] Earlier diagnosis, better assessment of disease activity and clinical trials will improve the prognosis of TA.

CASE REPORT

A female patient aged 45 years reported to the outpatient ward with a chief complaint of shock like pain on left side of the face for past 8 years. Pain was of sudden onset, severe intensity with very short duration, radiating to the left shoulder. The pain aggravated in the morning and relieved in the noon. Patient was treated for trigeminal neuralgia for 4 years (gabapentin plus and zeptol)

Patient had the history of headache and before 4 years, she noticed numbness in both the upper limbs, percutaneous retrogasserian glycerol rhizotomy (PRGR) attempted but failed due to no cerebrospinal fluid (CSF) flow. She complained the history of weight loss of around 8 kg in past 5 years. With previous history and CT angiographic findings (Fig 1), patient was diagnosed as TA. Patient noticed watering of the left eye before 5 months and was advised steroid therapy. On intra-oral examination, patient had poor oral hygiene and grossly decayed 36. Left radial artery couldn't be palpated; the pulse and blood pressure were under normal limit. Since the patient was known case of TA, with physician consent, under antibiotic coverage, extraction of 36 was done due to poor prognosis. Patient reported for review with no fresh complaint, hence oral prophylaxis was done. The patient was advised to maintain oral hygiene and have a periodic dental visit. Since the patient was not on antiplatelet or on long term steroid, procedure was done only on antibiotic cover.

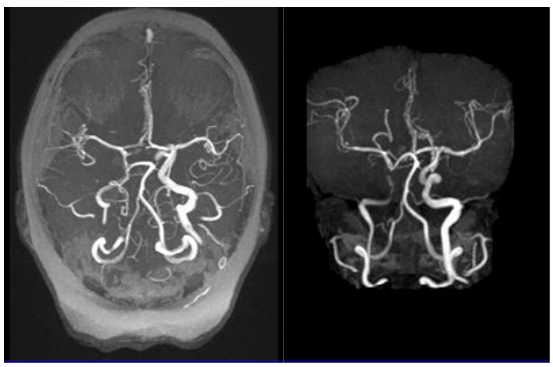

Fig 1: CT Angiogram of patient

MAXILLOFACIAL SURGEON'S PERSPECTIVE

Adequate knowledge and proper diagnosis play a key role in the management of TA patients since it is a rare disease. Like other medical condition, early morning appointments to be made to avoid stress. During all dental procedures, patients head should be positioned in a relaxed way to avoid pressure or stress on the carotid sinus region which might lead to bradycardia.[12] Gupta et al in his article has mentioned that dental surgical procedures preferably to be performed in the inactive phase (remission) of the disease.[12]

Extraction or other surgical procedures should be carried out under antibiotic cover to prevent bacteremia and proliferation in the inflamed vessels. Most of TA patients are on high dose steroid therapy during the active phase. In such situation, the dosage of steroid must be doubled (rule of two) to prevent adrenal crisis.

The TA patients are prescribed antiplatelet therapy to prevent ischemic complications. Hence before surgical procedures cardiovascular monitoring should be done.

CONCLUSION

Maxillofacial and dental problems are inevitable in any patient. Detailed history, knowledge about the disease and management of medically compromised patient with extra precautions is mandatory to avoid complications.

FINANCIAL SUPPORT AND SPONSORSHIP:

Nil

CONFLICTS OF INTEREST:

There are no conflicts of interest.

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