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Takayasu Arteritis: An Overview Review

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Abstract: A rare chronic granulomatous vasculitis, Takayasu arteritis (TA) mainly affects big arteries including the aorta and its major branches. Although autoimmune pathways are strongly implicated, the precise etiology of this condition, which is most commonly observed in young Asian women, is yet unknown. The illness develops gradually, starting with vague constitutional symptoms and ending with vascular problems like ischemia, aneurysms, hypertension, and pulselessness. Clinical examination, laboratory indicators of inflammation, and sophisticated imaging methods like CT angiography and MRI are all part of the diagnostic evaluation process. Corticosteroids, immunosuppressive medications, and biologics are commonly used in treatment; in more severe cases, revascularization may be required. Early identification and specialized multidisciplinary management are crucial because of its systemic nature and its consequences. With an emphasis on recent developments and treatment concerns, this review offers an updated synopsis of Takayasu arteritis's categorization, pathogenesis, clinical characteristics, diagnostic standards, and therapy approaches.

Keywords: *Takayasu Arteritis, Large-Vessel Vasculitis, Aortic Inflammation, Pulseless Disease, Angiographic Classification, Autoimmune Vasculitis, Corticosteroids, Biologic Therapy, Vascular Stenosis, Imaging in Vasculitis.*

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I. INTRODUCTION

Takayasu arteritis (TA), also referred to as pulseless disease, is a chronic inflammatory arteritis that damages the medium and large arteries and their branches. A common consequence is stenosis, occlusions, or aneurysmal degeneration of the aorta and its main branches, especially the renal, carotid, and subclavian arteries.[1]

II. CLASSIFICATION

➢ *New Angiographic Classification of Takayasu Arteritis*

Angiography's pattern of vascular involvement is used to categorize Takayasu arteritis. The disease is restricted to the aortic arch's branches in Type I. The aortic arch, its branches, and the ascending aorta are all involved in type IIa. The thoracic descending aorta is also involved in Type IIb, which consists of all the vessels involved in Type IIa.

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Type III involves neither the ascending aorta nor the arch, but rather the thoracic descending aorta, the abdominal aorta, and/or the renal arteries. Only the renal arteries and/or the abdominal aorta can have type IV.

Both the upper and lower segments of the aorta and its major branches are widely involved, as Type V is a combination of Type IIb and Type IV. This classification aids in the diagnosis, treatment, and tracking of the course of the disease.[2]

➢ *Ishikawa Clinical Classification of Takayasu Arteritis*

Takayasu arteritis can also be categorized according to the presence of complications and clinical severity. Patients in Group I may or may not have pulmonary artery involvement due to their uncomplicated disease. Patients with a mild to moderate single complication and otherwise uncomplicated disease are included in Group IIA. People in Group IIB have both uncomplicated disease and a severe single complication. Lastly, patients in Group III have both uncomplicated disease and two or more complications. This classification aids in estimating the burden of disease and choosing the best course of treatment.

III. ETIOLOGY

Takayasu arteritis's etiology is not well understood. Fundamentally, it is an inflammatory condition, and it is believed that autoimmune cell-mediated immunity could be the cause. The development of alternating regions of pseudoaneurysm and ischemic changes are ultimately caused by the transmural fibrous thickening of the arterial walls**.**

There is a theory, though, that the aortic tissue's 65 kDa heat-shock protein is triggered by an unidentified stimulus, which causes the vascular cells' major histocompatibility class I chain-related A (MICA) to be induced. The aortic arch is typically affected by TA, a chronic granulomatous panarteritis of major arteries, although one-third of cases also impact the pulmonary arteries15 and the rest of the aorta and its branches. In most cases, the aorta and its branch artery wall exhibit uneven thickening with intimal wrinkling on gross morphologic examination. The orifices of aortic branch vessels to the upper body may be significantly constricted or even completely destroyed when the aortic arch is affected. There may be equal effects on the renal and coronary arteries. Histological findings can include significant medial mononuclear inflammation or an adventitial mononuclear infiltrate with perivascular cuffing of the vasa vasorum (channels supplying blood vessels). Aortic valve insufficiency and dilatation may result from involvement of the aortic root. Coronary ostia may narrow as a result of myocardial infarction.[6]

IV. EPIDEMIOLOGY AND PREVALENCE

Global estimates of TAK incidence range from 1 to 2 cases per million annually, which is extremely low.[9]

Although prevalence estimates vary greatly, ranging from 0.9 to 40 per million, Asian populations typically have higher prevalences.[9]

According to estimates, there are one to two instances of TAK for every million persons worldwide each year. Estimates of prevalence range from 4.7 to 33 per million in Europe, with some studies estimating as low as 0.9 permillion in the US and as high as 40 per million in Japan.[9]

V. PATHOPHYSIOLOGY

The aorta and its branches are particularly vulnerable to Takayasu arteritis, an inflammatory condition affecting large and medium-sized arteries. Panarteritis with intimal proliferation is seen in advanced lesions. The inflammatory process can result in aneurysmal, occlusive, or stenotic lesions. Arterial narrowing can occur in any aneurysmal lesion. The primary consequences of vascular alterations include aortic insufficiency because of involvement of the aortic valve, pulmonary hypertension, aortic or arterial aneurysm, and hypertension, which is typically caused by renal artery stenosis or, less frequently, stenosis of the suprarenal aorta.Although myocarditis, pericarditis, and dilated cardiomyopathy have also been documented, congestive heart failure is a far more frequent finding. The aorta thickens macroscopically during the chronic phase due to fibrosis of all three vessel layers. Multiple areas are frequently affected by the patchy distribution of lumen narrowing. Rapid disease progression may result in insufficient fibrosis and the subsequent development of aneurysms. A common characteristic of many aortitides is the ridged, "tree bark"-like appearance of the intima.[4]

Vasculitis can be separated under a microscope into two phases: an acute florid inflammatory phase and a healed fibrotic phase. A vasa vasoritis is observed in the adventitia during the acute phase. Lymphocytes and occasionally giant cells with neovascularization infiltrate the media. The intima is thickened by fibroblasts, smooth muscle cells, and mucopolysaccharides. Fibrosis and the breakdown of elastic tissue occur during the chronic phase. Biopsy results may not distinguish between these two vasculitides because giant cell arteritis also exhibits similar histopathological findings. Correct diagnosis is typically made possible by clinical manifestations.[5]

VI. CLINICAL PRESENTATION

➢ *Early Stage (Prior to Pulselessness)*

- Symptoms of the Constitution : These include fever, nocturnal sweats, malaise, exhaustion, and weight loss.[7]
- Increased Inflammatory Indicators : Elevated levels of inflammatory markers such as CRP and ESR may be detected by blood testing.[7]

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➢ *Stage Late (Pulseless)*

- Claudication : Exercise-induced limb pain, particularly in the arms and legs, as a result of decreased blood supply.[8]
- Reduced or Nonexistent Pulses : A discernible variation in pulse intensity among limbs, or even the lack of pulses in certain regions, is a crucial indicator.[8]
- Bruises in the Vascular System : Using a stethoscope over constricted arteries, strange rushing noises were detected.[8]
- High blood pressure : elevated blood pressure, frequently brought on by renal artery stenosis.[8]
- Symptoms of Neurology : Headaches, lightheadedness, visual abnormalities (double or blurred vision), stroke-like symptoms (weakness, trouble speaking), and even convulsions are some examples of these.[8]
- Heart Problems : The aorta and its branches may become involved, resulting in heart failure, aortic regurgitation, and chest pain.[8]
- Additional Symptoms : These may include shortness of breath and stomach pain, particularly after eating.[8]

VII. DIAGNOSIS

➢ *Clinical Assessment*

- Physical examination and medical history : A physical examination and a thorough medical history, including the beginning of symptoms, are essential for identifying possible Takayasu arteritis symptoms.[10]
- Signs and symptoms : Fatigue, fever, weight loss, headaches, disorientation, muscle and joint discomfort, and signs of vascular insufficiency (such as absent or reduced pulses, limb-specific blood pressure variations, and bruits over afflicted arteries) are only a few of the symptoms that may be present.[11]

➢ *Imaging Research*

- Angiography : The main imaging technique in the past was conventional angiography, which uses contrast dye but is risky and intrusive. It facilitates the visualization of aneurysms, blood vessel narrowing (stenosis), and other anomalies.[10]
- CTA (or) CT angiography: A common non-invasive imaging method for diagnosing Takayasu arteritis and determining the degree of vascular involvement is CTA.[10]
- MRI and MRA : In situations where radiation exposure must be kept to a minimum, magnetic resonance imaging (MRI) and magnetic resonance angiography (MRA) can also be used to see blood vessels, measure inflammation, and determine the severity of the condition.[10]
- PET/CT : A more modern imaging technique that can aid in the detection of inflammation and is occasionally employed in Takayasu arteritis is Positron Emission Tomography (PET) in conjunction with CT scanning.

➢ *Blood Examinations*

- ESR and CRP : Although they are not unique to Takayasu arteritis, some individuals may have normal levels of the inflammatory indicators erythrocyte sedimentation rate (ESR) and C-reactive protein (CRP).[13]

VIII. RADIOLOGICAL FEATURES

- Thickening of the Arterial Wall : This is a noticeable characteristic that is seen on both MRI and CT, particularly in the early stages.[14]
- Occlusion and Stenosis : Intimal thickening or thrombus formation frequently causes arteries to narrow or become completely blocked.[15]
- Aneurysms : It is possible for the artery wall to dilate or balloon, especially in the aorta.[16]
- CT (or) double-ring enhancement : A distinctive pattern of a brightly enhanced outer ring (signaling media and adventitia) and a poorly enhanced inner ring (signaling intima) can be observed on contrast-enhanced CT.[14]
- Ultrasonic Macaroni Sign : The artery wall exhibits a macaroni-like circumferential thickening in transverse sections that is long, smooth, uniform, and somewhat echogenic.[16]
- Calcification : Particularly in chronic illness, linear calcifications in the descending thoracic aorta and aortic arch may be observed.[17]
- Decrease in Pulsatility : A decrease of typical arterial pulsations in imaging can indicate diminished or absent pulses, which are a clinical characteristic.[16]
- Changes in the Aortic Contour : The descending thoracic aorta may seem wavy or scalloped on radiographs, and there may be a loss of distinct definition.[17]
- Cardiomegaly : There may be cardiac enlargement, occasionally accompanied by rib notching and reduced pulmonary arteries.[17]
- Participation of Pulmonary Arteries : Additionally, pulmonary artery stenosis, blockage, or aneurysms may result from Takayasu arteritis.[18]

IX. CRITERIA

These criteria, which cover both major and minor criteria, are frequently utilized.[19]

➢ *Major Criteria*

- Lesions in the left and right mid-subclavian arteries : blockage or narrowing of these arteries, as shown by angiography.[20]
- Typical symptoms and indicators that last for at least a month : These include fever, limb pulse differences, pulselessness, and other systemic symptoms.[17]

➢ *Minor Criteria*

High erythrocyte sedimentation rate (ESR), carotid artery soreness, hypertension, aortic regurgitation, and abnormalities in other arteries are some of these signs.[21]

- Validation and Grading : Certain criteria systems give points for various observations and imply that a high likelihood of Takayasu arteritis is indicated by a particular cumulative score.[22]
- The significance of angiography : For determining the degree of the disease and visualizing the vascular

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involvement, angiography is regarded as the gold standard.[23]

X. MANAGEMENT

The goals of treating Takayasu arteritis are to reduce inflammation, control hypertension, and avoid consequences. Corticosteroids are usually the first line of treatment, and if necessary, immunosuppressants or biologics come next. For severe cases or problems, revascularization techniques such as surgery or endovascular therapies may be taken into consideration.[10]

➢ *Medical Management*

- Corticosteroids : To lessen inflammation, prednisone or prednisolone are frequently administered. Although some drugs may need long-term usage and have possible adverse effects, they can be useful in causing remission and controlling symptoms.[17]
- Immunosuppressants : To lessen inflammation and possibly taper off steroid use, medications such as methotrexate, azathioprine, or mycophenolate mofetil can be administered either in addition to or instead of corticosteroids.[17]
- Biologics : Biologics such as tocilizumab or TNF inhibitors may be employed to alter the immune system if other therapies prove ineffective.[24]
- Antihypertensives : Since high blood pressure can be a serious side effect of Takayasu arteritis, it is imperative to manage it.[24]
- Antiplatelet Treatment : Blood clots and other problems can be avoided using aspirin or other antiplatelet medications, particularly in individuals with critical cranial or vertebrobasilar involvement.[24]

➢ *Revascularization*

- Bypass Surgery : For severe or long-segment stenosis with significant fibrosis, bypass grafting may be an option.[10]
- Endovascular Techniques : In certain situations, narrowing arteries can be widened by angioplasty or stenting.[25]

XI. DISCUSSION

The prevalence of Takayasu Arteritis (TA), a rare but important form of chronic vasculitis, ranges from 0.9 to 40 per million people worldwide, predominantly affecting women under 40, particularly in Asian nations.[9] An early inflammatory phase with systemic symptoms like fever, malaise, and elevated inflammatory markers (e.g., ESR, CRP) is the typical biphasic pattern in which the disease manifests,[7][13] and a final stage that is characterized by ischemic symptoms as a result of artery occlusion and constriction.[8]

Panarteritis, adventitial vasa vasoritis, and intimal proliferation are all hallmarks of the chronic granulomatous inflammatory process that underlies the pathophysiology of TA.[4][5] It has been hypothesized that heat-shock protein molecular mimicry may cause autoimmune activation.[6]

An important part of diagnosis is imaging. For vascular assessment, traditional angiography is still the gold standard,[23] because non-invasive imaging techniques like CTA, MRA, and PET/CT can measure vessel wall inflammation and disease activity, they are becoming more and more popular.[10][14] Disease stratification is aided by a number of classification schemes. Moriwaki et al.'s angiographic categorization distinguishes six kinds (I–V) according to vascular involvement,[2] on the basis of complications, Ishikawa's classification offers a clinical severity assessment.[3]

TA management is a multidisciplinary process. The cornerstone for bringing about remission is high-dose corticosteroids,[17] while steroid-sparing immunosuppressants (e.g., methotrexate, azathioprine) and biologics like tocilizumab are used for maintenance and refractory cases.[24] However, biologics like tocilizumab and steroid-sparing immunosuppressants like methotrexate and azathioprine are utilized for maintenance and refractory cases.[10][25] Monitoring disease activity and avoiding relapses remain difficult despite advancements in treatment. Although they are nonspecific, biomarkers such as CRP and ESR are useful. Clinical follow-up and imaging are still essential for evaluating treatment response.[13][14]

XII. CONCLUSION

Takayasu arteritis is an uncommon, complicated vasculitis that can have serious consequences if left untreated or misdiagnosed. It might manifest as severe vascular problems or as generalized systemic symptoms. It is essential to make an early diagnosis by combining sophisticated imaging with clinical examination. Immunosuppression is necessary for treatment in order to reduce inflammation, and surgery may be necessary if there is vascular compromise. Patient outcomes can be significantly enhanced by prompt interdisciplinary intervention and more knowledge. This study advances our knowledge of TA and encourages its early identification and integrated treatment, which benefits medical professionals and enhances the prognosis for those who are impacted.

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