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Takayasu Arteritis: A current perspective on diagnosis and emerging treatments

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Abstract

Takayasu-Arteritis (TA), also known as the pulseless disease or aortic arch syndrome is a rare but serious condition that primarily affects mainly the large blood vessels. This inflammatory vascular disease is characterised by intimal fibrosis and vessel narrowing, especially affecting the aorta, its branches and the pulmonary arteries. Patients typically present asymptomatic but such conditions as weak or absent pulses in arms or legs, angina pectoris and stroke-like symptoms can appear in severe cases.

A comprehensive understanding of TA disease is essential for prompt diagnosis and effective management. This review will examine the historical context, symptoms, diagnostic criteria, new therapeutic approaches, and future directions in research and treatment.

Keywords: Takayasu-Arteritis, vasculitis, pulseless disease

Introduction

Takayasu-Arteritis (TA) also known as the pulseless disease or aortic arch syndrome, is a rare and severe form of chronic inflammatory panarteritis of the large vessels that predominantly affects the large arteries such as the aorta with its main branches, sometimes the pulmonary and coronary arteries (Rizzi and Bruno, 1999). This condition is characterised by a chronic granulomatous inflammatory reaction resulting in intimal and adventitial fibrosis. This in turn leads to progressive occlusion of the affected blood vessels and causes stenosis and aneurysm.

The precise aetiology of TA remains unclear; however, there is a strong association between HLA-B52 and the strongest genetic risk factor. The disease manifests clinically in a variety of ways, encompassing a broad spectrum of symptoms. These include myalgias, fever, weight loss, angina pectoris, and claudication of the upper limbs, resulting in a systolic blood pressure difference.

Historical Background and Discovery

The history of Takayasu-Arteritis can be traced back to the early beginnings of the 20th century. The initial description of this inflammatory arthritis is attributed to Mikito

Takayasu, a Japanese ophthalmologist, who in 1908 described a singular case of a 22-year-old woman who suffered from gradual loss of vision in both eyes. Further investigation showed wreath-like anastomosis and aneurysmal dilatation of the retinal vessels in the fundus schema (Takayasu, 1908). Simultaneously Yoshiakira Ohnishi presented a similar case at the 12th Annual Meeting of the Japanese Ophthalmology Conference. Since then the disease has been known as “Takayasu disease” or “Takayasu-Ohnishi disease” (Sugiyama et al., 2009). Later on, Nakajima discovered similar cases and classified the symptoms as an independent disease, which criteria included inter alia fundus findings with arteriovenous anastomoses, affecting both eyes of young women of the age of 20 and an impalpable radial artery (Nakajima, 1921). In 1948 the name “pulseless disease” was introduced by Simizu and Sano and the lesions were not only detected in the aortic arch but also the abdominal aorta and renal arteries (Shimizu and Sano, 1948). The name “Takayasu Arteritis” was officially implemented worldwide in 1990 as the American College of Rheumatology published the typical criteria for this inflammatory arteritis (Arend et al., 1990).

Since its discovery, the medical community has made significant progress in understanding the causes and development of Takayasu-Arteritis. Ongoing research in this field has deepened our understanding of Takayasu-Arteritis, leading to enhanced diagnostic criteria and more effective treatment strategies.

Epidemiology and Genetic Role

The disease typically affects individuals in their youth or middle age, with a peak onset between 20 and 30 (Onen and Akkoc, 2017). The data on the occurrence of TA is limited due to the fact that the condition is often diagnosed late, which complicates the undertaking of incidence studies. Though first described in Japan and linked to Southeast Asia, its incidence there (1–2 per million per year) is similar to Europe. A 2021 systematic meta-analysis by Rutter et al. estimated an incidence rate of 1.11 per million person-years (95% CI 0.70–1.76) with a higher prevalence observed in women, though significant variability across populations was also identified (Rutter et al., 2021). Recent studies report prevalence rates of 8.4 per million in the U.S. (Sanchez-Alvarez et al., 2021), 13.2 in Sweden (Mohammad and Mandl, 2015) and 25.2 in Norway (Gudbrandsson et al., 2017) - approaching figures in South Korea (28.2) (Park et al., 2017). However, small sample sizes in non-Scandinavian populations limit interpretation.

Furthermore, vascular involvement in TA varies by ethnicity. The widely used angiographic classification divides TA into five types based on vessel involvement. Type V, affecting the aorta and its branches, is the most common globally, including in Japan, China, Turkey, France, Italy, and Mexico (Numano et al., 2000; Bicakcigil et al., 2009; Soton et al., 2008). In contrast, Type I, limited to the aortic arch branches, is predominant in Northern Europe. A Norwegian study confirmed these patterns, with Type I common in Northern Europeans and Type V in Asian patients (Gudbrandsson et al., 2016).

Evidence suggests a genetic role in Takayasu-Arteritis, with HLA-B52 being the strongest genetic risk factor. First identified in a Japanese family study, this association has been confirmed worldwide (Terao, 2016). Research on the HLA genomic region in TA has also identified associations with HLA-Cw12:02, and HLA-DRB107, along with effects in the CCHCR1, HLA-B/MICA, and HLA-DRB1/HLA-DQB1 loci. The HLA-B52 association is well-validated and may explain other links due to high linkage disequilibrium. Further studies are needed to confirm the HLA-DRB1*07 and HLA-DRB1/HLA-DQB1 effects (Renauer et al., 2017).

Aetiology and Pathophysiology

The underlying cause of TA appears to be a chronic inflammatory process targeting the walls of the aorta, driven by the infiltration of cytotoxic immune cells, such as natural killer (NK) cells and CD8+ T cells, along with the increased expression of immune molecules like major histocompatibility complex (MHC) proteins and the MICA/MICB family (Arnaud et al., 2011).

Histologically, TA is characterized by the presence of activated NK cells, γδT cells, and CD8+ T cells within the affected aortic tissues. These cells are thought to play a pivotal role in the disease's progression, with activation markers such as NKG2D, MICA, and MHC class I molecules being highly expressed (Arnaud et al., 2011). Studies like the one from Teraro C. et al., 2013 have shown that certain genetic factors contribute to the susceptibility to TA, including interleukin-12p40 (IL-12p40), MICA, MICB, and genes in the leukocyte immunoglobulin-like receptor (LILR) family. These genes may promote the activation of cytotoxic lymphocytes by regulating the expression of cytokines and other molecules critical for immune responses. IL-12p40, which is essential for the development of Th1 and Th17 cells, has been identified as a key player in the pathogenesis of TA. Variants in the IL12B gene, particularly the risk allele of rs6871626, are associated with higher levels of IL-12p40,

correlating with clinical manifestations of the disease such as aortic regurgitation and elevated inflammatory markers (Teraro et al., 2013).

In addition to IL-12p40, genetic studies such as Renauer PA et al., 2015 have highlighted other significant loci, such as HLA-B and LILRB3, which appear to influence the immune system's response in TA. LILR family receptors, which modulate the activity of leukocytes, may contribute to the inflammatory process. Specifically, LILRB3 is involved in inhibiting immune cell responses, while LILRA3 may function as a decoy receptor, preventing excessive immune activation (Renauer et al., 2015). These findings underscore the complex genetic underpinnings of TA and suggest that the interactions between MHC class I molecules, LILRs, and NK cells are central to the disease process.

The role of B-cells in Takayasu-Arteritis is debated. Some studies show increased anti-aorta antibodies and anti-endothelial cell autoantibodies (AECA) (Wang et al., 2010), while others report conflicting results. Elevated B cell-activating factor (BAFF) in active TA suggests potential B-cell activation as reported by Nishino et al. though further research is needed (Nishino et al., 2010).

The research by Salwalha et al. has also emphasized the role of NK cells in TA. Enhanced NK cell activity has been noted in patients with TA, and the presence of genetic variants in the MICB region, along with the association of HLA-B52 with the disease, points to the critical role of the MIC family in disease development. These insights offer potential targets for future therapeutic interventions.

On the treatment front, targeting cytokines involved in TA has shown promise. IL-12p40 and IL-6, both of which are implicated in the disease’s inflammatory response, have become key targets for therapy. A randomised controlled study by Nakaoka et al. has demonstrated the effectiveness of IL-6 receptor inhibitors, such as tocilizumab, in managing refractory TA, offering new hope for patients with difficult-to-treat disease (Nakaoka et al., 2018). Other cytokine-blocking therapies, including TNF inhibitors, are also being explored as potential treatments, signalling a move toward more personalized, targeted approaches in managing TA.

In summary, the pathophysiology of Takayasu-Arteritis is complex, involving a combination of genetic susceptibility, immune cell activation, and cytokine-driven inflammation. Continued research into the molecular mechanisms and genetic factors behind TA will likely lead to better diagnostic tools and more effective treatments for this challenging condition.

Clinical Manifestations and Symptoms

The clinical manifestations of TA vary widely, with symptoms differing in both severity and duration. These symptoms can greatly affect quality of life, making early intervention crucial to prevent further progression. Recognizing these symptoms at the earliest stage is essential for ensuring effective management.

Generally, the arteritis can be divided into three different phases. The first phase is very unspecific and can be constitutional. It can be characterized by a fever of unknown origin, weight loss, malaise, anaemia, arthralgias and myalgias. In the second stage, the large arteries are affected due to mural inflammation causing angina pectoris, dorsal pain and carotidynia, a pulse pain periodicity located in the neck zone, especially on the left side. The third and last phase is known for diminished or absence of pulses in the arteries and/or differences in the blood pressure mainly in the upper and to a lesser extent in the lower extremities. Limb signs or symptoms are among the most common manifestations of Takayasu arteritis, occurring in approximately 85% of patients (Bhandari et al., 2023). One reliable indicator for diagnosing this condition is the systolic blood pressure difference between both arms (>10mmHG). Systemic hypertension often complicates the course, especially when the renal arteries are affected (Keser et al., 2018).

Further clinical features of Takayasu-Arteritis may include:

- **Cardiac Manifestations:**

Up to 40 percent of patients experience cardiac involvement, leading to arrhythmias, pericarditis, and ischemic heart disease. These findings encompass aortic regurgitation secondary to aortic root dilation and ostial or segmental coronary artery stenosis, which can result in ischemic heart disease. Congestive heart failure is reported as the most frequent cause of mortality in this patient population (Weyand CM et al., 1995).

- **Neurological Involvement:**

Neurological complications may manifest as transient ischemic attacks (TIAs), ischemic or hemorrhagic strokes, dizziness, lightheadedness, headaches, seizures, and visual disturbances or vision loss (Bhandari et al., 2023).

- **Pulmonary Involvement:**

TA can affect the pulmonary arteries, potentially leading to complications such as pulmonary hypertension due to vascular inflammation and stenosis. This may result in impaired lung function, with associated symptoms including shortness of breath, coughing, and chest discomfort. Hemoptysis may occur in some instances due to vascular damage, while pleuritis can cause pleuritic chest pain and respiratory distress (Bhandari et al., 2023).

- **Ophthalmic Manifestations**

Takayasu's retinopathy with microaneurysm formation and arteriovenous anastomoses is rare, likely due to early diagnosis and treatment (Vanoli et al., 1995). Eye changes are typically bilateral and strongly associated with the extent of common carotid and/or vertebral artery involvement.

Diagnosis

Currently, there is an absence of universally recognised diagnostic criteria for systemic vasculitides, including TA. Therefore, a set of classification criteria was devised for patients who had already been diagnosed with vasculitis. The initial set of criteria was established in 1990 by the American College of Rheumatology (ACR) and comprised six fundamental elements. A minimum of three of these criteria had to be fulfilled for a diagnosis of Takayasu arteritis to be made:

1. Onset at age ≤ 40 years.
2. Claudication of an extremity.
3. Decreased brachial artery pulse.
4. A difference of more than 10 mm Hg in systolic blood pressure between the arms.
5. A bruit over the subclavian arteries or the aorta.
6. Arteriographic evidence of narrowing or occlusion of the entire aorta, its primary branches, or large arteries in the proximal upper or lower extremities. (Arend et al., 1990).

The updated classification criteria were established by the American College of Rheumatology (ACR) and EULAR in 2022 and are outlined in the table below.

Section	Criterion	Points
Absolute Requirements	Age ≤ 60 years at time of diagnosis	Mandatory
	Evidence of vasculitis on imaging[1]	Mandatory
Additional Clinical Criteria	Female sex	+1
	Angina or ischemic cardiac pain	+2
	Arm or leg claudication	+2
	Vascular bruit[2]	+2
	Reduced pulse in upper extremity[3]	+1
	Carotid artery abnormality[4]	+2
	Systolic blood pressure difference in arms ≥ 20 mm Hg	+1
Additional Imaging Criteria	Number of affected arterial territories (select one)[5]	
	— One arterial territory	+1
	— Two arterial territories	+2
	— Three or more arterial territories	+3
	Symmetric involvement of paired arteries[6]	+1
	Abdominal aorta involvement with renal or mesenteric involvement[7]	+3
Sum the scores for 10 items, if present. A score of ≥ 5 points is needed for the classification of TA.		

FOOTNOTES

1. Evidence of vasculitis in the aorta or branch arteries must be confirmed by vascular imaging (e.g., computed tomography/catheter-based magnetic resonance angiography, ultrasound, positron emission tomography).
2. Bruit detected by auscultation of a large artery, including the aorta, carotid, subclavian, axillary, brachial, renal, or iliofemoral arteries.

3. Reduction or absence of pulse by physical examination of the axillary, brachial, or radial arteries.
4. Reduction or absence of pulse of the carotid artery or tenderness of the carotid artery.
5. Number of arterial territories with luminal damage (e.g., stenosis, occlusion, or aneurysm) detected by angiography or ultrasonography from the following nine territories: thoracic aorta, abdominal aorta, mesenteric, left or right carotid, left or right subclavian, left or right renal arteries.
6. Bilateral luminal damage (stenosis, occlusion, or aneurysm) detected by angiography or ultrasonography in any of the following paired vascular territories: carotid, subclavian, or renal arteries.
7. Luminal damage (stenosis, occlusion, aneurysm) detected by angiography or ultrasonography involving the abdominal aorta and either the renal or mesenteric arteries.

Table 1. Own elaboration based on ACR/EULAR criteria for Takayasu-Arteritis (2022). (Grayson et al., 2022)

In cases where TA is suspected, it is imperative that the diagnosis is confirmed through the utilisation of imaging methodologies. Computed tomography angiography is the recommended modality for diagnosing TA and is considered more efficacious than magnetic resonance angiography due to its superior image resolution and cost-effectiveness. (Moriwaki al. 1997):

- **Type I:** classic type involving solely the aortic arch branches
- **Type II:**
    - **IIa:** Ascending aorta, aortic arch and its branches
    - **IIb:** Ascending aorta, aortic arch and its branches, thoracic descending aorta
- **Type III:** involvement of the thoracic and abdominal aorta distal to the arch and its major branches (e.g. descending thoracic and abdominal aorta +/- renal arteries)
- **Type IV:** sole involvement of the abdominal aorta and/or the renal arteries
- **Type V:** generalized involvement of all aortic segments

Laboratory test results

There are no standard biomarkers to reliably assess disease activity in TA. While the Erythrocyte Sedimentation Rate (ESR) and C-reactive protein (CRP) reflect systemic inflammation, they don't always correlate with vascular inflammation. IL-18 and IL-6 levels are elevated in active TA, with IL-18 particularly correlating with disease activity and serving as a potential marker. Pentraxin 3 (PTX3), which correlates with vascular inflammation, may be a better marker than C-reactive protein (CRP), although it's nonspecific and elevated in other autoimmune diseases. More research is needed to identify reliable biomarkers for TA (Chandhu et al. 2023).

Treatment

The management of Takayasu-Arteritis involves a multifaceted approach, with the aim to control inflammation, induce remission and prevent complications.

Non-pharmacological assessment

This includes smoking cessation, a healthy diet and sleep schedule and psychological support. As evidenced by the findings of Li G. et al. (2020) and Shao N. et al. (2017), resistance exercises and curcumin have been demonstrated to be efficacious in the management of TA, with a reduction in TNF-α levels observed in pertinent studies.

Medical Approach

Disease-modifying anti-rheumatic drugs (DMARDs) are fundamental in the medical treatment of TA. Synthetic DMARDs such as methotrexate (MTX), azathioprine (AZT), mycophenolate, leflunomide and cyclophosphamide offer better disease control and reduce vascular complication (Misra et al., 2021).

The ACR 2021 guidelines conditionally recommend combining immunosuppressive drugs with glucocorticoids over monotherapy use of glucocorticoids in order to minimize their toxicity. Methotrexate (MTX) is the most commonly used drug, often combined with

oral high-dose steroids as an initial therapy. Additionally, tumour necrosis factor (TNF) inhibitors—such as infliximab, etanercept, and adalimumab—as well as azathioprine may also be utilized in early treatment (Maz et al., 2021). If there is no clinical remission observed it is recommended to either switch to another DMARD or to tocilizumab, a recombinant humanized anti-IL-6 receptor monoclonal antibody, which case reports and case series since 2008 have reported as effective in refractory TA (Nakaoka et al., 2018).

New therapy approaches

JAK-Inhibitors

A contemporary approach involves the inhibition of the JAK/STAT signalling pathway, which has been demonstrated to play a substantial role in the development of TA (Régnier P. et al., 2020). Tofacitinib, a representative JAK inhibitor, is recommended by the Japanese Guidelines (Kawahito et al., 2020) solely in cases where methotrexate (MTX) has proven ineffective. As demonstrated in the clinical trial conducted by Kong X. et al. in 2022, tofacitinib has been shown to be more efficacious than methotrexate (MTX) due to its superior capacity to induce clinical remission, prevent disease relapses and reduce glucocorticoid dependence. Nevertheless, valid concerns regarding the long-term safety of this treatment for young women, particularly its potential connection to cancer, have been expressed. Further trials are required in order to ascertain whether there is any significant impact (Arita et al., 2024).

Ustekinumab

Ustekinumab, an antibody against p40 used for psoriasis, Crohn's disease, and ulcerative colitis, has shown symptom improvement in TA patients as shown in trials such as Gon Y. et al. (2021), but no new clinical trials are underway.

Rituximab

Rituximab is a monoclonal antibody targeting CD20 on B lymphocytes. It treats conditions like non-Hodgkin lymphoma, chronic lymphocytic leukemia, scleroderma, and

pemphigus vulgaris/foliaceus. Some case studies suggest its effectiveness in refractory TA, but more evidence is needed (Mutoh et al. 2019).

Surgical intervention

Endovascular therapy (ET) and open surgery are the primary approaches for invasive treatment. ET, involving balloon angioplasty and stenting, is preferred for treating short lesions, whereas open surgery is recommended for larger lesions or when ET is not possible (Jung et al., 2018). It is recommended to first manage disease activity before proceeding with invasive procedures.

Conclusions

Takayasu's disease is characterised by a set of specific symptoms, including weak or absent pulses, significant differences in blood pressure between the arms or legs, and pain or fatigue in the limbs (claudication) due to reduced blood flow. It is crucial to recognize these symptoms early to ensure an accurate diagnosis. The ability to detect and respond quickly can significantly impact patient outcomes. Diagnosing the condition typically involves a thorough clinical evaluation alongside imaging techniques like angiography to detect vascular changes. Successful management requires a multidisciplinary strategy that integrates both medical treatment and lifestyle modifications.

Recent research presents encouraging possibilities for new treatments such as the increasingly modern JAK-inhibitors. In the future, a shift toward personalized medicine could lead to more tailored treatment plans for individuals with TA. Additionally, continued studies on genetic factors and immune responses may pave the way for targeted therapies that help manage symptoms and slow disease progression.

A comprehensive approach that combines medical treatment with psychological support is increasingly recognized as essential for effectively managing Takayasu-Arteritis.

Staying up to date with new research developments is crucial for improving patient outcomes and adapting to the evolving understanding of the condition.

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