A CASE REPORT ON PAKSHAGHATA WITH REFERENCE TO TAKAYASU’S ARTERITIS

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ABSTRACT

Takayasu’s arteritis is a rare, chronic, inflammatory, granulomatous large vessel vasculitis predominantly affecting the aorta, its major branches and the pulmonary arteries due to which it presents as pulseless upper extremities. The neurological symptoms vary depending on the degree and nature of blood vessel obstruction. A 28 year old female arrived with a short history of fever and chills followed by partial weakness of left upper and lower limb (hemiparesis) since three days. History of the patient was recorded, examinations were performed which yielded knowledge on absence of radial pulsation on right upper limb and feeble to absent radial pulsation on left upper limb. The condition can be diagnosed as margaavaranajanyapakshagata. This article highlights the ayurvedic understanding of pathogenesis of Takayasu’s arteritis leading to stroke as a complication. The critical understanding of pathogenesis of the disease has been described on the basis of the concept of Sahajasatmya and its de-stability triggering autoimmune reaction. The sequence of autoimmune reaction culminates in granulomatous changes. This process of formation of granulomatous change is the vitiation of raktha by pitta dosha. Ultimately it leads to rakthavrittavata ending in margaavaranajanyapakshagata (hemiparesis). The diagnosis is made on the demonstration of vascular lesions in large and middle sized vessels on angiography, CT, magnetic resonance angiography; contrast angiography. The earliest detectable lesion is a local narrowing or irregularity of the lumen, which may develop into stenosis or aneurysms and occlusion.

Keywords: Takayasu’s arteritis, margaavaranajanyapakshagata, Sahajasatmya, raktha

INTRODUCTION

Takayasu disease or Takayasu’s arteritis is a rare¹ chronic progressive idiopathic inflammatory disease characterised as chronic granulomatous vasculitis resulting in occlusive or ectatic changes² mainly in the aorta and its immediate branches as well as the pulmonary arteries and its branches³,⁴. The classic presentation is characterised by a biphasic pattern of manifestation involving an initial systemic
phase – with a beginning non inflammatory vascular stage followed by an inflammatory vascular stage presenting with nonspecific symptoms such as fever, fatigue, etc. A second occlusive phase is later manifested, depending on the site of occlusion, various areas of the body or various systems are affected exhibiting their respective features. We report a case, which presented straight away with a major catastrophic complication of the disease and was marked by atypical angiographic findings.

Case report:
Presenting concerns:
A 28 year old female, a plantation worker, presented to us with a short history of fever and chills followed by partial weakness of left upper and lower limb since three days.

Historical data:
There was no past history of arthralgia, myalgia, night sweating, anorexia, claudication or other systemic manifestations.

Clinical findings:
On examination patient was moderately built and nourished.
The arterial pulsation in both the upper limbs were not palpable (on left upper limb radial and brachial pulses were feeble to absent, whereas on right upper limb pulses was absent). In lower limb the arterial pulsations were palpable. Carotid pulsations were palpable on both sides. Blood pressure was not able to be recorded in upper limb. Gait was hemiplegic. Cardiovascular examination revealed an apex beat in the fifth intercostal space in the mid clavicular line and a systolic murmur of grade 1 over the mitral area and radiating to the left infraclavicular area. There were no renal vascular bruits. Other symptoms revealed no significant abnormality. The fundoscopic examination was normal. Higher mental status examination of patient was intact. All twelve pairs of cranial nerves were functioning within normal limits. Motor system assessment revealed hypotonic muscles with a reduced muscle bulk for the left upper and lower limb. Muscle strength was assessed as 3/5 for left upper and lower limb. Sensory system and coordination of the patient was intact. Deep tendon reflexes were brisk on the left side.

Diagnostic focus and Assessment:
Diagnostic results:
The complete blood investigation revealed a raised erythrocyte sedimentation rate, with a borderline reduction of haemoglobin percentage and serum potassium level. Urine analysis showed no cast or other abnormalities. Chest X – Ray showed no abnormalities. The echocardiogram revealed trivial mitral regurgitation with mitral valve prolapse. Magnetic resonance angiogram was done, indicating major cervical vessel occlusion, except right vertebral artery. Gross wall thickening of left subclavian artery was observed and both common carotid, where right common carotid occlude more than the left common carotid. Magnetic resonance imaging of brain revealed acute infarct on right M1lenticular striate territory.

Diagnostic reasoning
A diagnosis of Takayasu disease was done, as the patient satisfied four of six criteria mentioned according to the diagnostic criteria framed by the American
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College of Rheumatology [Table 1]. An initial phase treatment was given from an allopathic institute – antipyretics. But, unfortunately after two days, patient succumbed with stroke due to an acute infarct. Hence, second phase of treatment was given with anti-inflammatory drugs, antilipidemic and blood thinners.

**Therapeutic focus**

Rehabilitation from strokeresulted as a complication from Takayasu disease.

**DISCUSSION**

Takayasu disease, also known as pulse less disease, Martorell’s syndrome, Young female arteritis, occlusive thromboaortopathy or Ruder – Harbitz syndrome is a chronic inflammatory arteritis affecting large and small vessels predominantly the aorta and its main branches.

Gian Bathista in 1830 published the description of arteritis. Morgagni in 1761, described the entity. The disease is named after Dr. Mikito Takayasu, who in 1908 described it as a wreath like appearance of retinal vessels with absence of radial pulse, supported by Dr. Onishi and Dr. Kagoshima.

Table 1: American College of Rheumatology criteria for classification of Takayasu disease

<table>
<thead>
<tr>
<th>Sl. No</th>
<th>Diagnostic criteria</th>
</tr>
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<tbody>
<tr>
<td>1.</td>
<td>Age of disease onset less than or equal to 40 years of age</td>
</tr>
<tr>
<td>2.</td>
<td>Claudication of extremities</td>
</tr>
<tr>
<td>3.</td>
<td>Decreased brachial artery pulse</td>
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<tr>
<td>4.</td>
<td>Blood pressure difference of greater than 10mmHg</td>
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<tr>
<td>5.</td>
<td>Bruits on subclavian arteries or aorta</td>
</tr>
<tr>
<td>6.</td>
<td>Arteriogram abnormality</td>
</tr>
</tbody>
</table>

Takayasu disease has worldwide distribution but prevalent in Asian population especially in Japan and India. Takayasu disease is the commonest cause of renovascular hypertension in India.

Etiology of Takayasu disease is still unknown. Takayasu continues to be an enigma even a century after its discovery. The exact cascade of pathogenic process is yet to be established. Till now the aetiologies have been postulated based on a post infection (especially tubercular infection) based on studies conducted in India and Japan. Genetic link to the predominance of disease in Asian population is due to HLA B52 haplotype. HLA B52 link possess more with aortic regurgitation, ischemic heart disease and pulmonary infarction are much more common. Renal artery stenosis occurs more frequently in HLA B39 positive patients. Also, involvement of microbes and other abnormalities in cellular mediated immunity can trigger the pathogenesis. But, they have not been absolutely proved.

Symptoms are initially often non – specific and may include the different stages of the diseases. Initial stage symptoms include fatigue, rapid, unintended weight loss, muscle or joint pain and low – grade fever. It is possible that swelling could damage arteries for months or even years before the next phase symptoms occurs.
In the second stage, inflammation has caused arteries to narrow enough to reduce the amount of blood, oxygen and nutrients reaching to certain tissues or organs. The second phase symptoms include arm or leg weakness or pain with use, light headedness, dizziness, fainting, headache, difficulty thinking and remembering, visual disturbances, high blood pressure, difference in blood pressure between both arms, diminished or absent pulse in the wrists and sometimes ankles, anaemia, which may make you feel tired or weak, chest pain, shortness of breath and fatigue\textsuperscript{20}. Repeated inflammation (swelling and healing) of the arteries can lead to the following complications like – hardening and narrowing of blood vessels, inflammation of the heart – either of the heart muscle itself (myocarditis) or sack surrounding the heart (pericarditis)on the heart valves (valvulitis), heart failure, aneurysm in the aorta, higher blood pressure, ischemic stroke or transient ischemic attack (a temporary stroke that has all the symptoms of an ischemic stroke without causing damage), heart attack and pulmonary artery problems\textsuperscript{21}.

**Table 2:** Angiographic classification of Takayasu’s arteritis (Takayasu conference, 1994)\textsuperscript{22}

<table>
<thead>
<tr>
<th>Type</th>
<th>Vessel involvement</th>
</tr>
</thead>
<tbody>
<tr>
<td>Type I</td>
<td>Branches from aortic arch</td>
</tr>
<tr>
<td>Type IIa</td>
<td>Ascending aorta, aortic arch, and its branches</td>
</tr>
<tr>
<td>Type IIb</td>
<td>Ascending aorta, aortic arch and its branches, thoracic descending aorta.</td>
</tr>
<tr>
<td>Type III</td>
<td>Thoracic descending aorta, abdominal aorta, and/or renal arteries.</td>
</tr>
<tr>
<td>Type IV</td>
<td>Abdominal aorta and/or renal arteries.</td>
</tr>
<tr>
<td>Type 5</td>
<td>Combined features of Type IIb and IV.</td>
</tr>
</tbody>
</table>

**Ayurvedic Perspective on Takayasu Disease leading to Stroke:**

Aetiology:

*Daiva ashetu* (unknown factors or due to past deeds) or *svabhavikahetu* (genetic or natural causes or acquired from present life deeds)\textsuperscript{23}. In this case the initial development of patient was with Takayasu’s Arteritis. The proposed etiological factors or predisposing factors derived from studies conducted throughout the world specifically in Indian sub-continent and Asian countries include –Genetic involvement (HLA B52 haplotype) in the individuals suffering from Takayasu disease, occurrence of *Takayasu* disease in patients with post infection especially tuberculosis and other abnormal cell mediated immune responses to vessels.

Still even after a century of discovering the syndrome the precise etiological factors are unclear. But, the condition is understood as an autoimmune disorder causing granulomatous intimal fibrosis.

**Premonitory symptoms:**

Headache (in headachevata dominated *do-sha* accumulate in head, later vitiate *raktha* – blood and *snayu*– nerves present in head to produce headache)\textsuperscript{24}.

**Clinical symptoms:**

Fever (*Rasa pradoshalakshana*), giddiness (produced due to *raja – manodosh*, *pitta* and *vatadosha*), tiredness (*margavarana-janya*), pain (describes as a symptom of hemiplegia – pricking type of pain in affected limb)\textsuperscript{25}, spasticity (seen in patient
on the final phase of elbow extension of left upper limb due to *snayupraptavata*, where disordered *snayu* (tendons) produces stiffness and contraction\(^{26}\).

**Pathogenesis:**

In normal state, in a healthy human body – *vata*, *pitta* and *kaphadosha* irrespective of opposite qualities co-exist each other rather than contradicting each other. This is termed as *sahaja satmya*\(^{27}\), which the conventional system coined as the so-called immune tolerance. Hence, this is the reason why the body’s own immune cells are not attacking self from Ayurvedic point of view. But, due to the influence of *daiva* (past deeds or unknown factors) or due to *svabhavika karanas*\(^{28}\) (in which we can include all the probable theories or mechanism of initiating autoimmune reaction such as microbial, genetic, immunological) potentially paving way to imbalance or disequilibrium or destability to the *sahajasatmya*, there by the immune cells (leucocytes) lose the ability to identify self from non – self or foreign agents.

To be more specific, on looking into the features told in *vataprakruti* individuals, depicting the features of *vata* itself, it includes certain *gunas* like the *sheegra guna*\(^{29}\), by virtue there are other two *gunas* mentioned in the literature as *alpasmruti* (lesser remembrance – abnormality to WBCs) and *sheegragrahita*\(^{30}\) (early identification of tissue). The above mentioned features occur at the level of *raktha* i.e. to be precise at the level of WBCs in the recognition of body tissues. This mechanism leads to the mistaken judgement of the body tissues as an external antigen whereas, while describing the *pitta prad-hanalakshana* by Charaka, it has been mentioned the feature of *pitta* as *teekshna guna*\(^{31}\) of *pitta* by virtue of its leading to *teekshnaagni* and *teekshnaparakrama*\(^{32}\). This causes the attack of immune system, here to the body’s own tissues\(^{33}\). This cascade of abnormal and noxious reaction drives an autoimmune disorder sequence. This is nothing but the impaired functioning of *dosha* (*ojovisramsa* – as it is the malfunctioning and displacement of *doshas*\(^{34}\)). Then due to the repeated action of *pitta* i.e. repeated inflammation. There is vitiation of blood induced by *pitta dosha* – ultimately leading to *sthayahath* or *ghanambhavathi* (granulomatous formation)\(^{35}\) as mentioned in the context of describing the features of blood vitiated by *pitta*. This is nothing but repeated inflammation and its failure leading to formation of organized mass of macrophages, where organized mass refers to a tight ball of mass and is termed as Granuloma.

This mode of pathology in Ayurveda is nothing but “*Raktavrittavata*” limiting its manifestation to certain regions of the body (*mandalani cha*)\(^{36}\). This pathogenesis resembles the pathogenesis of *vatarakta*, where the complications such as *moorcha*, *mada*, *bhrama* and *vedana* resemble to an extent\(^{37}\). So, in this case the site of *Raktavrittavata* is on to different *dhamani* (artery – based on the definition given in classics). And finally the occlusion in these arteries cause aggravation of *vata* in turn leading to the manifestation of *Pa-kshagata* (hemiplegia). According to Acharya Susrutha, he clearly mentions about the manifestation of contralateral hemiplegia due to aggravated *vata* in *urdhwa* (superior), *adhoga* (inferior) and
tiryaggata (oblique) dhamani (vessels)\textsuperscript{38}, to an extent we can understand it as circle of villus.

**Diagnostic focus and assessment:**
The etiological diagnosis is shonithadushti – vitiation of blood (post infection or cell mediated immunity abnormality), anatomical diagnosis impact on urdhwagadhamani – superior branch of vessels (M1 Segment of MCA), pathological diagnosis margavarana – occlusion (raktavrittavata – obstruction of movement of vatadosha by raktha) i.e. granuloma formation causing stenosis or even obstruction and clinical diagnosis pakshagata (hemiparesis on left side) i.e. left sided contralateral hemiparesis. Treatment foravarana should be followed by intervention of hemiparesis. Here, the case is hemiparesis with the predominance of vatadosha as patient is devoid of any symptoms like heaviness or burning sensation.

**CONCLUSION**
The case is relatively common when overlooking the gender (female) of the patient and the age of incidence (28 years). But, the presence of short duration of non – specific symptoms in patients such as the fever and chills, occlusion of all the major cervical vessels except the right vertebral artery irrespective of absence of any major symptoms elicited in the history and the complication of Takayasu disease ending with stroke made the case relatively rare. Ayurvedic understanding of pathology in Takayasu disease ending in stroke has not been discussed in any of the scientific articles, but through this case the pathogenesis has been plotted with authentic references. Takayasu’s arteritis often called as Aortic Arch Syndrome is a group of features arising from the granulomatous intimal fibrosis. The condition is difficult to be diagnosed though mere clinical features and examinations. Radiological evidence of wreath like appearance in the wall of arteries is confirmatory for this particular condition. In Ayurveda, this syndrome is nothing but a pathological state of raktavrittavata – blood causing obstruction in the path of vatadosha and not a proper disease diagnosis. This particular pathology has the potential to end in stroke or pakshagata; which the modern science acknowledges as a rare complication to Takayasu Disease.

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