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CASE REPORT

TAKAYASU ARTERITIS PRESENTING WITH MASSIVE CEREBRAL ISCHEMIC INFARCTION WITHSEVERE UNILATERAL RENAL ARTERY STENOSIS IN A 28-YEAR-OLD MALE: A CASE REPORT

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ABSTRACT

Takayasu arteritis is a relatively rare type of large-vessel arteritis that primarily affects the aorta and its major branches, the coronary arteries, and the pulmonary arteries. Depending on the different groups of blood vessels involved in the disease process, the clinical presentation of Takayasu arteritis varies.Takayasu arteritis (TA) is a chronic vasculitis disease of unknown etiology. Clinically significant renal disease is relatively common, and renovascular hypertension is the major renal problem. Here we report a case of male presenting with a debilitating massive cerebral ischemic infarct that turned out to be a relatively rare presentation of Takayasu arteritis. We reported a case of 28 yr old male presented to the Emergency Department with quadriparesis, pain andnumbness of his arms and absent bilateral radial, brachial and subclavian pulse. His laboratory results showed an elevated leucocytes counts, raised C-reactive protein and erythrocyte sedimentation rate. MRI brain withmr-angiography revealedacut with chronic cerebral ischemic infract with narrowing and occlusion of the major branches of his aortic arch with many collaterals. Contrast ct abdomen revealed significant right renal artery stenosis. ***Conclusion:*** Takayasu arteritis is a relatively rare disease with various and sometimes devastating clinical manifestations, such as massive cerebral ischemic infarction as in our case. Currently, there are multiple diagnostic tools and treatment options available, and more under investigation. Early, appropriate diagnosis and initiation of proper therapy could avoid further progression and reduce complications of the disease.

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INTRODUCTION

Takayasu arteritis (TAK) is a rare autoimmune, idiopathic, and chronic granulomatous large vessel vasculitis mainly affecting the aorta and its large branches. It induces a variety of nonspecific inflammatory (systemic and local) and ischemic symptoms due to the formation of stenotic lesions and thrombi. The inflammatory process initially leads to thickening of the arterial wall which may result in stenosis, occlusion, dilatation, aneurysm formation or rupture of the involved arteries (Jolly and Curran, 2005). Symptoms of the disease can be very diverse depending on the location and degree of involvement of the inflamed vessels. Clinical manifestations include fatigue, weight loss, fever, faintness, headache, and differences in arterial pressure between bilateral upper or lower limbs. The most severe symptoms include pulmonary thrombosis, aortic regurgitation, congestive heart failure, cerebrovascular events, degeneration of vision or blindness, and hearing problems. In severe cases, it is very difficult to feel a pulse in patients, and for this reason TAK is called 'pulseless disease' (Terao *et al*., 2014). Correct diagnostics, early recognition and appropriate therapy are very important for prevention of severe complications. Glucocorticoids and immunosuppressive drugs are primarily used for treatment, but biological drugs are playing an increasing role in therapy.

It is critical to elucidate the pathophysiological mechanisms involved in TAK for further treatment development. The exact etiology is assumed to be a cell-mediated inflammatory process within the vasculature, which can result in occlusion, aneurysmal dilatation, and constriction in afflicted segments because of mononuclear and granulomatous infiltrates, which causes a variety of symptoms[1]. During the disease's acute phase, patients may also have symptoms such as limb weakness or pain, headaches, syncopal attacks, and uneven blood pressure. Fever, weight loss, and exhaustion are examples of constitutional symptoms that may occur first[2]. Takayasu arteritis is a relatively rare type of large-vessel arteritis that primarily affects the aorta and its major branches, the coronary arteries, and the pulmonary arteries. Here we report a case of a male presenting with debilitating massive cerebral ischemic infarction that turned out to be Takayasu arteritis. The most frequently used diagnostic pattern in clinical and epidemiologic studies of TAK is ACR Classification Criteria, which includes five clinical and one imaging criteria (Arend *et al*., 1990). A diagnosis of Takayasu arteritis according to the criteria of ACR from 1990, might be made if a patient has at least three of these six criteria present:

1. Age <40 year at disease onset
2. Claudication of extrimities
3. Decreased brachial artery pulse
4. Difference of >10 mmhg systolic blood pressure between arms.

5. Bruit audible on auscultation over one or both subclavian arteries or abdominal aorta.
6. Pathological changes (usually focal or segmental) in aortic arteriography and/or its branches

CASE PRESENTATION

A 28-year-old male presented to the Emergency Department (ED) with complaint of generalized weakness in all four limb (quadriparesis) with dizziness and syncope. Patient have history of left hemiparesis with left brachial palsy 5 yr back with history of hospitalization for 3 week. Weakness improved with in 1 month after stroke but left sided body weakness persist. No significant past medical or surgical history with no history of any trauma.

There was no family history of early onset cerebral vascular accident. Patient denied the use of alcohol, tobacco products or recreational drugs. On physical examination, he was conscious and oriented to time place person, heart rate was 88 beats per minute (bpm). no pulse palpated over radial brachial and bilateral subclavian artery. Blood pressure non recordable bilateral upper limb and bilateral lower limb right popliteal artery blood pressure 240/134mmHg and left popliteal artery blood pressure 160/100mmHg. There was obvious left-sided hemiparesis with a positive Babinski sign.Motor system examination bulk reduced all four limb. Powerright sided improved to normal but left sidedpower score 4/5. Left wrist drop with claw finger present. on cvs examination absent right carotid pulsation and carotid bruit with weak pulsation over left carotid. No cardiac murmur or cardiac rub. left renal artery bruit present. Laboratory test results showed white blood cells (WBCs) of 13.9x109/L, hemoglobin (Hb) of 13.2g/L, and platelets (PLTs) of 238x109/L.

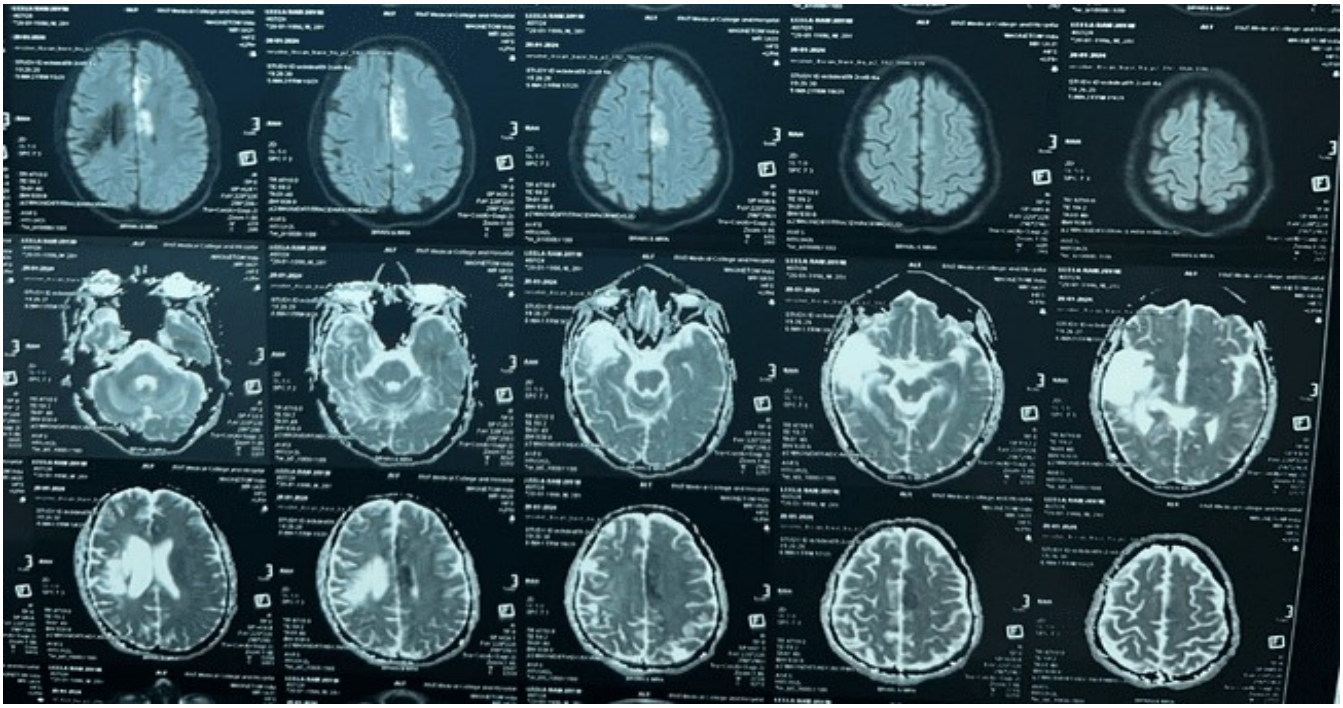

Fig. 1. MRI. T2W hyperintensity in right frontoparieto temporal region in parafalcian location with FLAIR hypointense in right fronto-parieto-temporal region

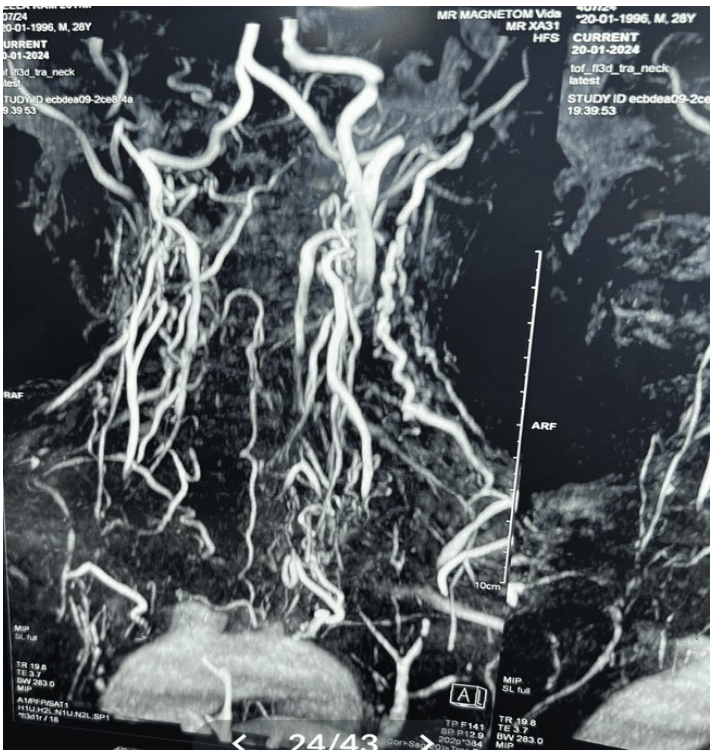

Fig. 2. NECK VESSELS: absent signals in right common carotid internal carotid and external carotid . and fow related absent signal in left common carotid

ESR was 43 mm/1hr. urine examination normal. renal and liver function normal. Serum triglyceride and LDL cholesterol mild elevated. His serum was negative for antinuclear antibody, antinuclear cytoplasmic antibody, and rheumatoid factor. Serology for HBsAg, HCV, and HIV was non reactive. The chest radiograph was normal. No coronary artery abnormalities were found out on 2 D echocardiography. Ultrasonography of the abdomen showed smaller size right kidney (5.6 x 2.3 cm) and left kidney size (10.4×4.4cm). Bilateral renal arteries doppler study showed significant right main renal artery stenosis.echocardiography show normal sinus rhythm. MRI Brain with MRA-acutinftact in left frontal and left parietal region in parafalcine location and left parietal region. Gliosis with encephalomalacia in right fronto-parieto-temporal region and corona radiata region. Reduced caliber of right middle cerebral artery with flow related absent signals in right common carotid artery, right external carotid artery and right internal carotid artery. There are absent flow related signal in left common carotid artery. CT angiogram bilateral upper limb:-calcified as well as non calcified atherosclerotic plaques are seen in arch of arota and decending thoracic arota . there are non opacification of right brachiocephalic trunk, right subclavian artery, right common carotid artery and right axillary artery and right brachial artery opacified due to colletral formation. Bilateral proximal and mid radial and ulnar artery opacified and non opacification of distal radial and ulnar. there is evidence of non-opacification of left subclavian artery and left axillary artery however left brachial artery opacification due to colletral formation. There are evidence of non opacification left common carotid artery from origin till its bifurcation however left internal and external carotid artery opacification due to colleteral formation.

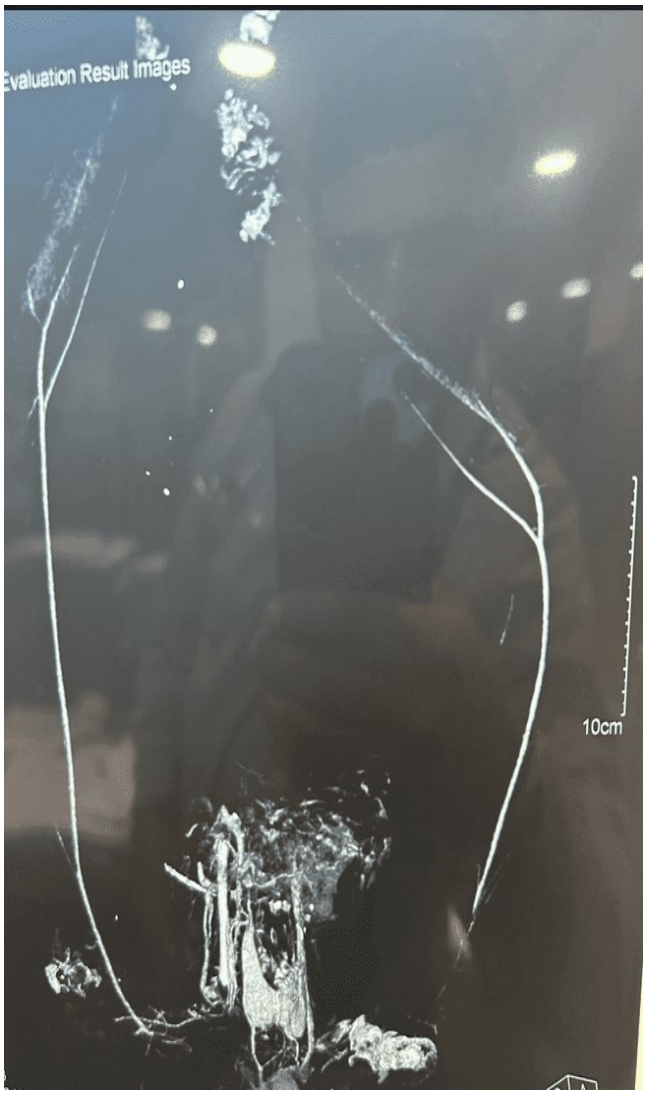

Fig. 3. CT angio bilateral upper limb arterial system non opacification right brachiocephalic trunk and left common carotid till bifurcation

MRI cervical spine: long segmented T2W hyperintense signals changes in chord parenchyma from lower border of C4 to lower brder of C6 vertebrae likely transverse myelities. CT angiography abdominal aorta – narrowing at origin of celiac artery and superior mesenteric artery. There are narrowing at origin and reduced caliber of right renal artery with reduced caliber and small sized right kidney.

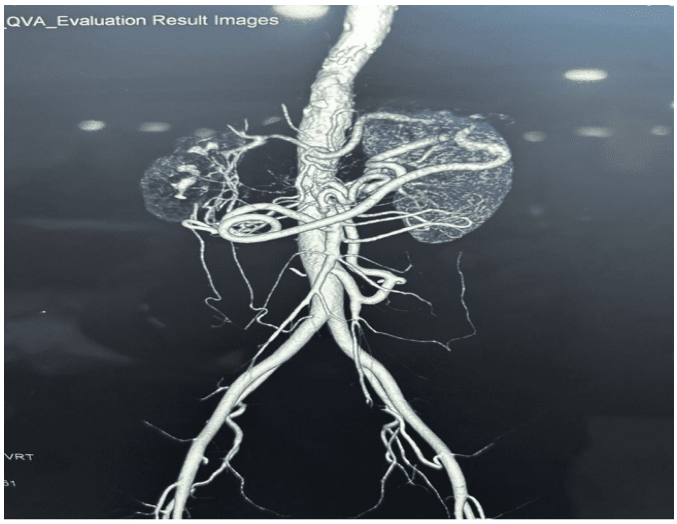

Fig. 4. CT abdomen show stenosis of right renal artery

DISCUSSION

After the Japanese ophthalmologist Mikito Takayasu, who first documented a case in 1905, Takayasu arteritis is so named.[3] The aorta, its major branches, the coronary and pulmonary arteries, as well as other big and medium-sized vessels, are all affected by the chronic inflammatory vasculitis known as Takayasu. Because of the frequent obstruction of big arteries coming from the aorta, it is also known as a pulseless condition. The exact pathophysiology is unknown.[4] However, pan arteritis, which has substantial intimal hyperplasia, medial and adventitial thickening, infiltration of mononuclear cells, and occasionally giant cells, is thought to be the cause.[5] It is mostly observed in Asian-descent women, peaking in the 30s. Takayasu illness affects 2 in 10,000 individuals annually, with a male to female ratio of 8:1.6 Takayasu's national data are still not available, though. Depending on the vessels involved, the takayasu disease can manifest clinically in a variety of ways. It typically begins with constitutional symptoms like fever, weight loss, claudication, and fatigue, and then progresses to other symptoms like weakness, dizziness lightheadedness, high blood pressure, retinopathy, aortic regurgitation, vascular bruits, neurological symptoms like seizures and syncope, etc., depending on the occlusion caused by the inflammatory infiltrates. In almost 20% of instances, neurological symptoms are present.[6] vessels were involved in the distribution of involvement. Three out of the six criteria listed above have a sensitivity of 90.5% and a specificity of 97.8% for diagnosis.[7]The conditions atherosclerotic, inflammatory, infectious, and genetic that affect the major arteries are included in the differential diagnosis of Takayasu disease. Examples include atherosclerosis, fibromuscular dysplasia, TB, and giant cell arteritis. The gold standard for diagnosing Takayasu's Arteritis is angiography. Doppler and non-invasive MRA, however, can also produce just as good results.[8]Acute phase reactants like ESR and CRP also provide additional evidence in favor of the diagnosis. The mainstay of treatment is expected to be systemic glucocorticoids and immunosuppressants, which are believed to reduce inflammation and limit the development of the illness.[09] If irreversible arterial stenosis develops due to conditions such cerebral ischemia, hypertension with critical renal artery stenosis, extremity claudication, or both, surgical intervention (endovascular) may be required.[10]

To avoid problems, surgical intervention is typically discouraged during active disease and advocated during quiescent disease. Our patient is under 30 years old, has bilateral brachial pulse impalpability, significant lumen narrowing of the bilateral subclavian arteries, abnormal thickening of the wall of the arch of the aorta, brachiocephalic artery, right subclavian artery, and right as well as left common carotid artery, all of which meet the diagnostic criteria. Collateral vessels sustain the brain perfusion. Syncope is a brief loss of consciousness brought on by cerebral hypoxia. Our patient experienced dizziness before sporadic syncope. The most commonly used therapeutic agents include CS and conventional IS agents, such as MTX. In patients who remain resistant and/or intolerant to these agents, biologics, including anti-TNF agents, RTX and tocilizumab, seem promising. Antiplatelet treatment may lower the frequency of ischaemic events in patients with TA.In the presence of a critical short-segment arterial stenosis causing life-threatening conditions, the principle of treatment is mainly revascularization of the affected organs by endovascular interventions including balloon angioplasty or stent graft replacements. On the other hand, long-segment stenosis with extensive periarterial fibrosis or occlusion requires surgical bypass of the affected segment, which is clearly associated with superior results compared with endovascular intervention. Both endovascular interventions and surgical procedures should be avoided

during the active phase of the disease. Post-interventional IS treatment is recommended. Earlier diagnosis, better assessment of disease activity and future clinical trials will help improve the management of patient.

CONCLUSION

Takayasu arteritis belongs to rare, idiopathic diseases of the immune system affecting the aorta and its branches. It is increasingly recognized in Europe and clinical practitioners should be aware of this disease. Non-specific systemic symptoms, which are accompanied by no pulse or various types of ischemic symptoms, should be considered along with deep diagnostic imaging. Early diagnosis and proper treatment can protect the patient from dangerous complications. Diagnosis and monitoring disease activity in TAK may be accomplished by the integrated use of non-invasive imaging methods, patient symptoms, clinical findings and acute phase reactants. However, there is no single imaging method that provides all the information required and each method has distinct and complementary roles in assessing disease activity and vascular inflammation. Glucocorticoids and immunosuppressive drugs are mainly used for treatment of the disease, but biological drugs are increasingly used in therapy. More research is needed to better understand the path diagnosis of this disease, as well as to introduce new drugs and treatment regimens so that disease remissions are longer, treatments more effective, leading to a better life quality for patients.

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