Case Report

Type I Takayasu's arteritis: An unusual presentation in young female child

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Abstract

Takayasu's arteritis is a rare type of vasculitis of unknown origin in which aorta and its branches are affected. Aortoarteritis causes narrowing of vessel lumen due to inflammatory changes in the vessel wall which in turn leads to thrombus formation, occlusion, or both. However, very rarely these may present exclusively with facial palsy at the time of onset. We present such a rare case of young female child who reported to the outpatient department with unusual findings of facial palsy followed by stroke and was diagnosed as Type I Takayasu's arteritis on the basis of various investigations. Magnetic resonance imaging and magnetic resonance angiography play a significant role in making the diagnosis.

Key words: *Aortoarteriti, Inflammatory changes, Magnetic Resonance Angiography, Thrombus formation*

Takayasu's arteritis, also called as "pulse less disease," is characterized by inflammation and stenosis of medium and small-sized arteries [1]. It is prevalent in adolescent and young females. It is a chronic relapsing and remitting disorder with a gamut of symptomatology mainly attributed to the arterial ischemia related to the involved artery. Main presenting symptoms are headache, visual disturbances, weight loss, fever, arthralgia, and less commonly neurological manifestations like stroke or seizures. Sometimes, the symptomatology at the presentation does not lead to early diagnosis as it is seen in our case, who presented with facial palsy. This in turn may lead to mismanagement of the case.

CASE REPORT

A 12-year-old female child presented to the ENT Department of our hospital for the complaint of sudden deviation of the mouth to the left side along with dribbling of saliva from the corner of the mouth. There was no history of fever, seizures, visual disturbances, unconsciousness, weight loss, malaise, or arthralgia. She has got 2 years old history of off and on headache and neck pain which was relieved with the symptomatic treatment. This was also accompanied by complaint of fatigue and occasional palpitation after exercise. The child was taken frequently took outpatient department treatment for the episodes of headache but without complete relief.

On admission, she was conscious, afebrile, her blood pressure was 148/72 mm of Hg and pulse was 90 beats/min. The pulses were normal on both the sides. Pityriasis alba was seen over the neck region. On the basis of clinical examination, diagnosis of right-sided 7th nerve palsy upper motor neuron type was made, and she was put on conservative treatment with routine review and follow-up.

Two days after this episode, the child developed weakness of the right side of the body and was brought to the hospital again. There was no preceding history

of fever, seizures, visual disturbances, trauma, or unconsciousness. On examination, she was afebrile, conscious, with normal vital parameters. Fundus examination was normal. On central nervous system examination, she had right-sided hemiparesis with right-sided facial palsy. There was no pulsation of the left common carotid in the neck region. There was bruit at the base of left common carotid artery at its origin from the aorta. Clinical diagnosis of acute ischemic stroke was made, and emergency non-contrast computed tomography (CT) was advised.

CT scan showed a large infarct in the MCA territory (Fig. 1). There was mild leukocytosis and ESR was 22 mm/h by Westergren method. Rest of the blood investigations including C-reactive protein was normal. Mantoux test was negative. Magnetic resonance imaging (MRI) with TIW, T2W, Flair, and DW sequences done afterward which confirmed the fresh infarct in left MCA territory (Fig. 2). Magnetic resonance angiography (MRA) revealed stenotic left-sided common carotid and internal carotid artery (ICA) with a compensatory increase in blood flow on the contralateral side (Fig. 3a and b). The blood flow in left ICA was also not much appreciated in color Doppler. The patient was diagnosed as Type I Group IIB Takayasu's disease. Patient was hospitalized for 2 weeks and put on corticoids therapy with low dose of aspirin. The patient was also advised physiotherapy for the hemiplegic side. This has shown good response with power returning in the hemiplegic side, and she has been called for the follow-up after 3 months.

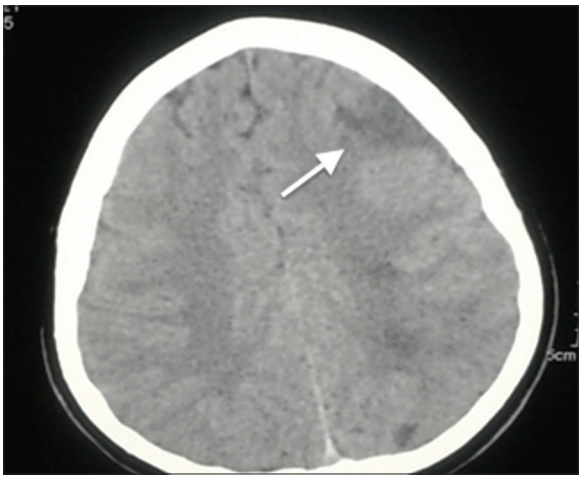

Figure 1: Non-contrast CT brain shows a wedge shaped hypodensity in left frontoparietal region (white arrow) suggestive of infarct

DISCUSSION

Takayasu's arteritis is more common in females than men, and second to fourth decade is the most common age group affected. This is less common in infancy and early childhood. This rare disorder is more commonly seen in Japan, South-east Asia, India, and Mexico [1,2]. In India, it has been noticed in patients between 3 and 50 years age groups. Following are the most commonly involved vessels: (1) Left subclavian artery <50% (2) brachiocephalic trunk, renal arteries, celiac trunk, superior mesenteric artery, and pulmonary arteries <50% (3) Left common carotid artery 20%. Histopathological studies show intimal hyperplasia, tunica media infiltration with mononuclear cells, and thickening of tunica adventitium of the vessel wall. The wall thickening fibrosis and stenosis leads to the ischemic changes [3]. It is diagnosed as per the criteria laid out in Takayasu conference 1994 according to the extent of involvement [4] (Table 1).

The presenting symptoms are mainly ischemia related headaches, visual disturbances, weight loss, fever, arthralgia, and less commonly neurological manifestations like stroke or seizures. Majority of the patients have hypertension which further complicate the symptomatology of the disease. 5-9% patients can present with the history of acute stroke. The association of facial palsy as presenting episode is slightly less common as in this case, and a high index of suspicion is required to diagnose these patients presenting with unusual complaints like facial palsy.

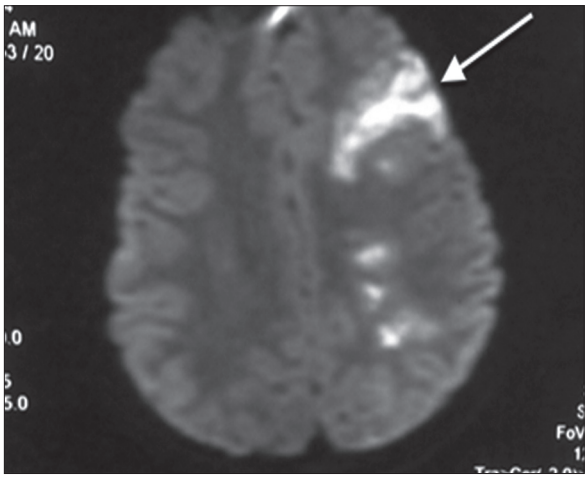

Figure 2: MR Diffusion weighted axial image shows restricted diffusion in left parietal region (white arrow)

Sometimes, the case becomes quite challenging due to a mismatch between clinical, biologic, and radiological information. Ishikawa has clinically divided the disease into three groups [5] as per the presenting features (Table 2).

The early diagnosis is essential to treat them properly without alarming vascular ischemic damage. There is a great advantage for using noninvasive techniques in the pediatric patient to avoid unnecessary complications. MRI, MRA, Doppler ultrasound, CT, and positron emission tomography (PET) are the common modalities to be used for the diagnosis [6,7]. MRI, MRA, and color Doppler are considered as the best for the diagnosis as these are radiation free and are the best to describe the anatomic details of the vessels. MRA has an important role in early diagnosis and post-treatment follow-up [8]. Spin echo T1, T2, FLAIR, and DW images are additional adjunct to the MRI for proper delineation and outline the impact of disease. Color Doppler can highlight the flow direction and spectral analysis in that particular vessel. PET imaging can provide the cellular level activity in the wall of the vessel much early than the other imaging modalities.

There is wide belief that it is a self-limiting disease which leads to a delay in treatment. However, the patients should be treated with corticoids and immunosuppressive drugs. The surgical intervention such as angioplasty and stenting is reserved for the patients who develop complications due to stenotic segment. Oral corticoids are started with 1 mg/kg daily in divided two doses and can be tapered as the symptoms subside. Long-term corticoid therapy may be required, and the consideration of osteoporosis should be tackled along with. Cytotoxic agents such as methotrexate, azathioprine, and cyclophosphamide are reserved for the patients whose disease is steroid-resistant or relapsing in nature. Anti-tumor-necrosis factor agents were used in patients of relapsing type who were not responding to other treatments [9]. 10 years survival is 90% but is less in case of complications.

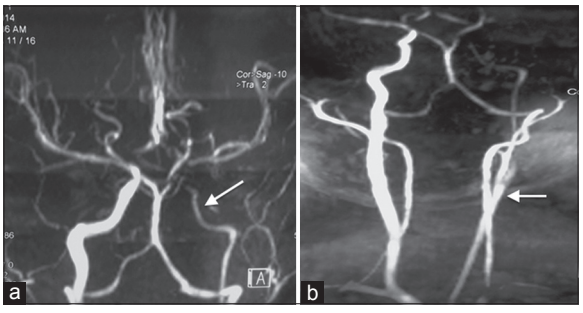

Figure 3: (a and b) Time- of- flight MR angiography image shows markedly attenuated left internal carotid (oblique white arrow) and middle cerebral artery. Right internal carotid and middle cerebral artery are of normal caliber. Left Common Carotid artery is totally compromised (horizontal white arrow)

CONCLUSION

The patient should be thoroughly investigated in the beginning for all the possibilities relating to the symptomatology. The presentation of Type I Takayashu's disease with isolated facial palsy is quite rare as is in our case. MRI has played a significant

Table 1: New angiographic classification of Takayasu's arteritis

Type	Vessel involvement
Type I	Branches from the aortic arch
Type IIa	Ascending aorta, aortic arch and its branches
Type IIb	Ascending aorta, aortic arch and its branches, thoracic descending aorta
Type III	Thoracic descending aorta, abdominal aorta, and/or renal arteries
Type IV	Abdominal aorta and/or renal arteries
Type V	Combined features of Type IIb and IV

Table 2: Ishikawa clinical classification of Takayasu arteritis

Group	Clinical features
Group I	Uncomplicated cases with or without pulmonary artery involvement
Group IIA	Mild/moderate single complication together with uncomplicated disease
Group IIB	Severe single complication together with uncomplicated disease
Group III	Two or more complications together with uncomplicated disease

role in diagnosing the case and to guide for the further management.

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