Takayasu Disease with Absence of Major Intracranial Arteries: A Case with Minimum Symptoms

Takayasu Disease (TD) is a relatively uncommon chronic and progressive vasculopathy. Aorta and its major branch involvement is typical feature of this disease. Absence of major branches of arch of aorta is not that common and is a fatal condition.

We present a 58-year-old female with absent bilateral carotid arteries and right vertebral artery (VA) with only left VA preserved with minimal symptoms. She had giddiness and mild headache with past history of hypertension since several months back. On examination she had mild weakness of left upper limb and could walk normally without support. Magnetic resonance image (MRI) revealed right frontal water shed infarction. Her carotid doppler study showed bilateral carotid stenosis which was confirmed by CT angiography (CTA). Conventional cerebral angiography showed absent brachiocephalic artery (BA) and its branches and absent left CCA. Subclavian artery was very tortuous with a prominent left vertebral artery (VA) arising from it. Left VA supplied bilateral anterior and posterior circulation through a prominent right posterior communicating artery (P Com A) and intact anterior communicating artery (A Com A). Bilateral normal anterior, middle and posterior cerebral artery (ACA, MCA and PCA) and their distal branches were well visualized.

The lady had only one, i.e. left VA, out of six major vessels, namely right external carotid (ECA), right internal carotid (ICA), right VA, left ECA, left ICA and left VA, of head. The patient is stable with just antiplatelet therapy.

**Key Words:** absent carotids, cerebral infarction, prominent VA, takayasu disease
head and 3 out of 4 main vessels of cerebral circulation, namely two common carotids arteries (CCA) and two vertebral arteries (VA), are absent and only one of them has covered the whole cerebral circulation with minimal symptoms. Therefore, the main objective of this article is not to highlight about TD but to highlight the fact that there is very minimal symptom despite blockage of all the major intracranial circulation in our case.

**Case Report**

A 58-year-old female patient presented to our outpatient clinic with the complaints of giddiness and mild headache with past history of hypertension for which she was under regular medication. On examination she had mild weakness of left upper limb with normal gait. MRI of head revealed right frontal infarction (**Figure 1**). Her carotid doppler study showed bilateral carotid stenosis. Computerized tomography (CT) and magnetic resonance (MR) angiography of neck vessels showed bilateral CCA complete stenosis. Conventional cerebral angiography was performed which showed absent brachocephalic artery and its branches and absent left CCA (**Figure 2**). Subclavian artery was very tortuous with numerous neovascularization. Left VA was prominent which supplied bilateral anterior and posterior circulation through a prominent right posterior communicating artery (P Com A) and intact anterior communicating artery (A

**Figure 1**: T2W MRI showing infarction in right frontal area.

**Figure 2**: Conventional angiography showing absent brachocephalic and left common carotid artery and prominent left vertebral artery
Com A) (Figure 3). Bilateral normal ACA, MCA and PCA and their distal branches were well visualized. Therefore, this case had absent right VA, right CCA and left CCA and only left VA was present.

She also had abnormal and stenosed left subclavian artery. So she had bilateral almost absent subclavian artery in addition to above mentioned cerebral vascular abnormalities.

She didn’t have any other cardiac, renal or pulmonary features of Takayasu Disease.

Discussion

Our case fulfilled the American College of Rheumatology criteria for the diagnosis of Takayasu Disease. Our case fulfilled four out of six criteria mainly claudication of extremities, decreased brachial pulse, audible bruit in left supraclavicular area and arteriographic vascularization.
abnormalities. The contradictory points in our cases were late onset of disease beyond 40 years of age and absence of significant difference in blood pressure between two arms, probably because of the involvement of bilateral subclavian arteries. Our case is a case of Takayasu Disease with extensive and massive arteritis.

Takayasu Disease was initially classified according to Nasu’s classification. Type I was involvement of branch of arch of aorta, Type II was involvement of arch of aorta and its 3 major branches and part of early descending aorta, Type III was involvement of abdominal aorta and its branches mainly renal arteries and Type IV was involvement of whole of aorta and its branches (Figure 4). However, with further clinical experience of this rare disease, Nasu’s classification was revised by Hata et al in 1994. According to Hata classification, Type I is involvement of branches of arch of aorta, Type II a is involvement of ascending aorta, arch of aorta and its branches, Type II b is involvement of Type II a and thoracic descending aorta, Type III is involvement of thoracic descending aorta, abdominal aorta and renal arteries, type IV is involvement of only abdominal aorta and renal arteries and Type V is involvement of whole aorta including II b and IV (Figure 5). Furthermore, the involvement of coronary or pulmonary artery is indicated as C (+) or P (+), respectively.

According to above classification, our case falls in type I. In our case all the major branches of arch of aorta were involved. Park MC et al also showed that type I was the most common type of Takayasu Disease in their study. 11

Regarding involvement of major cerebral vessels, Suwanwela et al have analyzed involvement of different major vessels in Takayasu Disease but they have not mentioned about involvement of extracranial carotid or vertebral arteries. It was mentioned that they didn’t find any case involving brachiocephalic artery or its branches or ascending aorta. Similarly Arnaud et al mentioned that most common Takayasu Disease was that of type V in white population but they have not mentioned in detail about the involvement arch of aorta and its major branches. Ringleb PA et al also mentioned that subclavian and common carotid artery were most commonly involved which correlates with our case.

In contrary, our case had complete occlusion of proximal BA and thus right VA and CCA were absent. Three major cerebral vessels namely right VA, CCA and left CCA were absent and only left VA was present. Conventional cerebral angiography showed adequate blood flow from left VA to right anterior circulation through a prominent right P Com A and then to left anterior circulation from right through an intact A Com A. Similarly there was adequate blood flow to bilateral posterior circulation from left VA.

Du Toit has mentioned that one of his three cases had abnormality in BA and CCA. But he has not mentioned about the degree of stenosis of BA or CCA.

Kobayashi et al showed in their study, conducted in Japan, that Takayasu arteritis involved mainly the aortic arch (58.8%), the descending aorta (45%) and the ascending aorta (30%). Among the branches, the left subclavian artery was most often involved (60%), followed by the left common carotid artery (40%) and the brachio-cephalic artery (19%).
Literatures have mentioned that only small portion of Takayasu Disease present with neurological symptoms. Ischemic stroke in Takayasu Disease is either due to major vessel occlusion or due to embolization from atherosclerotic vessels. Patients usually present with vertigo, visual disturbances, headache, ischemic stroke, seizure disorder etc. In recent studies less than one fourth of patients with (TD) have been found to have neurological signs and symptoms. Ringleb PA et al also mentioned that neurological symptoms were benign in spite of massive involvement of vessels. However, in some cases stroke can be the first and only presentation of Takayasu Disease as mentioned by Hamdan et al. Our case also presented with only stroke. Cases with both carotid and coronary artery involvement have been seen. However our case didn’t have coronary involvement.

References
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