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Case Report

Cerebral venous involvement in Takayasu arteritis—A rare encounter

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ABSTRACT

Takayasu arteritis commonly affects moderate to large size arteries. Contrast enhanced MR angiography is a sensitive tool to detect Takayasu arteritis. Venous involvement in Takayasu arteritis is an extremely rare phenomenon. A young girl of 19 years age presented with intense headache for one year duration and depressive features for three months. Neuroimaging (MR angiography) reports were suggestive of Takayasu arteritis. Also there were features suggestive of significant involvement of cerebral veins. Venous involvement is a rare phenomenon in Takayasu arteritis. This case report highlights about the rare association of cerebral venous involvement in Takayasu arteritis with literature review. The possible biological mechanisms of depressive symptoms and headache are also discussed.

1. Introduction

Takayasu arteritis is a relatively rare variant of vasculitis, which commonly involves large to moderate size arteries like − aorta and major arteries emerging from aorta. Human Leukocyte Antigen (HLA) is involved in Takayasu Arteritis(Terao, Yoshifuji, & Mimori, 2014). Recently different geno-biological markers like − HLA-DQB1/DRB1, IL12B, MLX and FCGR2A/3A are found to be associated with Takayasu arteritis (Terao et al., 2014). Indian studies report about female preponderance of Takayasu arteritis. These studies revealed that male patients have predominant involvement of abdominal aorta, whereas females have involvement of aortic arch and their branches (Parakh & Yadav, 2007). The average age of presentation in Indian population was also found to be in the third decade of life and hypertension being a common manifestation (Parakh & Yadav, 2007). In an Indian study, it was found that patients with Takayasu arteritis commonly present with hypertension, with aortic (arch of aorta, descending aorta and abdominal aorta) involvement as the most common (approximately 56%) vascular involvement(Jain, Kumari, Ganguly, & Sharma, 1996). Evidences suggest about the association of neuropsychiatric symptoms with vasculitis. Vasculitis involving cerebral vasculature may be manifested in the form of cognitive disturbances (impairment of memory, attention and executive function), mood disturbances, anxiety and changes in personality (Berlit, 2007).

2. Case history

A 19-years-old unmarried girl presented with the chief complaint of headache for the past one year, and sadness of mood, crying spells and disturbed sleep for three months. The headache was episodic, pulsatile in nature, usually unilateral, and used to last 2 to 3 h. She used to have multiple such episodes in a day and over the past three months it was continuous with episodic exacerbations from the baseline. There were no associated features like photophobia and phonophobia, besides occasional sense of nausea. Analgesics (naproxen, paracetamol, tramadol) had been prescribed for headache in previous consultations done elsewhere. Her sadness of mood and crying spells were persistent throughout the day causing impairment of her day to day functioning. She had also expressed ideas of hopelessness and worthlessness. There was, however, no past history of psychiatric illness. There was history of swelling on the right side of neck in the submandibular region. Fine needle aspiration cytology (FNAC) examination of the lymph node was done, which had revealed tubercular granuloma, for which she had received a course of antitubercular medications (rifampicin, isoniazid & pyrazinamide). History of 3syncopal attacks was present in the past 3 months.

Family history was non-contributory. On mental status examination, she had a depressed mood, pessimistic view about future, excessive worries and apprehensions related to her future as well as illness, and death wishes. She was diagnosed with moderate depressive episode and initiated on oral mirtazapine (15 mg/day) with clonazepam (0.25 mg, as and when needed) and for her intractable headache amitriptyline (10 mg/day) was prescribed. She underwent neurological evaluation for her complaints of intractable headache and episodes of syncope to rule out an organic cause. Both fundi were normal and there was no neurological deficit. The radial pulse on the left side was feeble while it

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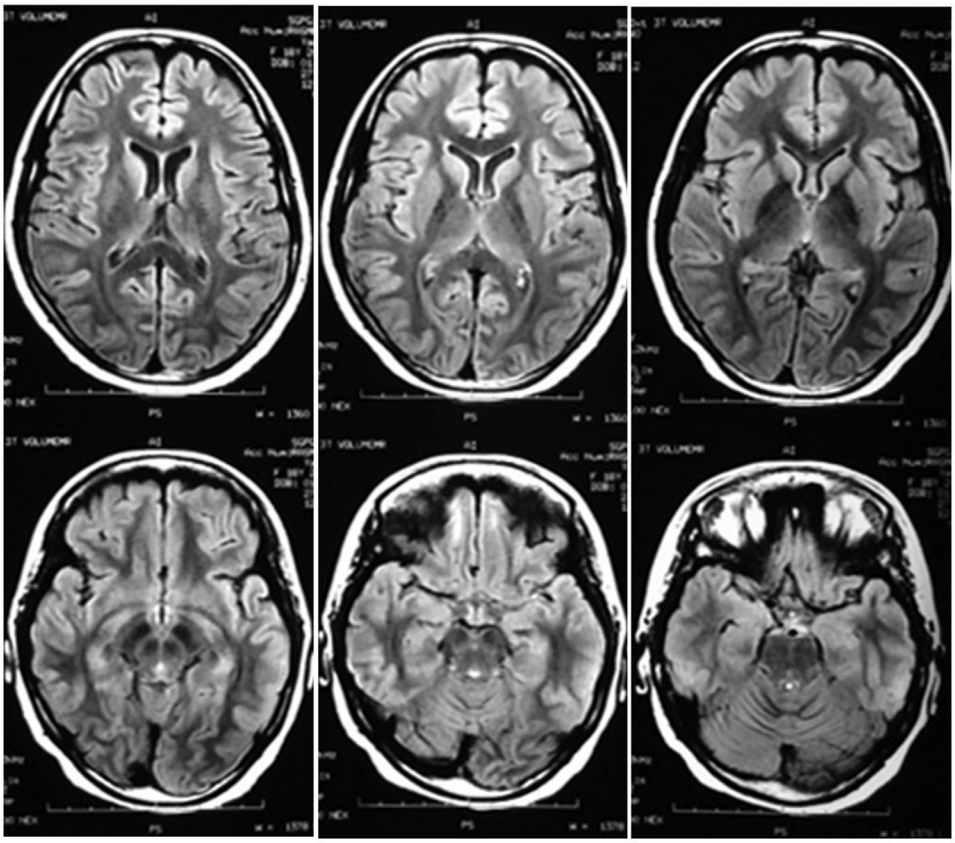

Fig. 1. Magnetic resonance imaging (MRI) of brain showing diffuse mild cerebral atrophy.

was barely palpable on the right side. There was a difference of 18 mm of Hg in the blood pressure recordings taken from her right and left arms (right < left).

Her routine hemogram was normal except for a low hemoglobin level (9.0 gm/dL). Her C-reactive protein and ESR (35 mm/h) were raised. Other laboratory estimations, including cerebrospinal fluid examination (CSF), were within normal limits. CSF manometry revealed a CSF pressure of 135 mm of H_2O.

Magnetic resonance imaging (MRI) of brain was suggestive of diffuse mild cerebral atrophy [Fig. 1]. MR-Angiogram was suggestive of involvement of the arch of aorta, carotid arteries and subclavian arteries [Fig. 2]. Involvement of dural venous sinuses and medium to small cortical veins was also evident in the form of a beaded-string or string-of-pearls due to segmental fibrosis. Additionally, there was involvement of both internal jugular veins (fibrosis of the walls giving rise to formation of stricture in the middle segment) [Fig. 3]. She was, thus, diagnosed with Takayasu arteritis.

The patient responded to the treatment and at 6 weeks her amitriptyline was increased to 25 mg/day; during this period she used clonazepam thrice. At 5 months follow up, she had sustained only 1 episode of headache and was doing all her activities of daily living independently. Her depressive symptoms also resolved completely. During follow up her antitubercular medications were stopped. She was started on prednisolone 20 mg/day, which later tapered off to 10 mg/day. Azathioprine (75 mg/day) was added to the treatment regimen. Follow up MRI was not possible due to financial constraints.

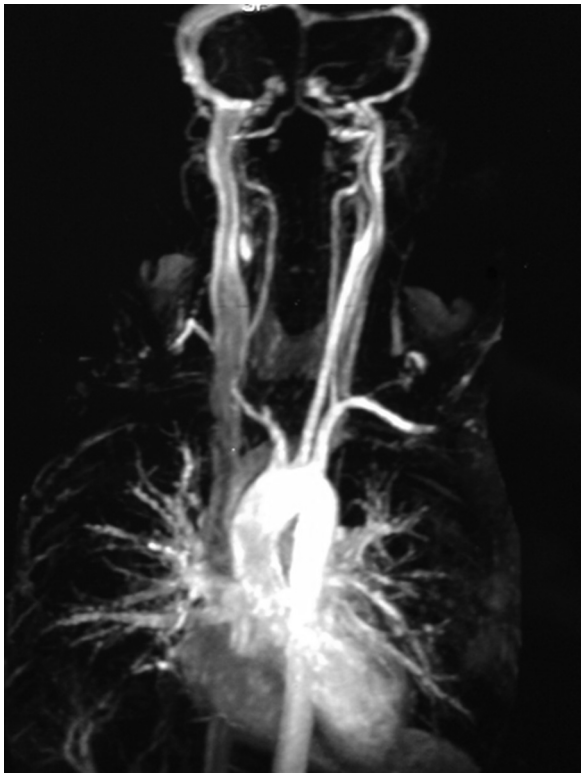

Fig. 2. MR-Angiogram showing involvement of the arch of aorta, carotid arteries, subclavian arteries as well as involvement of both internal jugular veins.

3. Discussion

Magnetic resonance imaging is considered to be the gold standard tool for detection of Takayasu arteritis, both in acute as well as chronic phase. Early diagnosis of Takayasu arteritis can be made by contrast-enhanced MRI and MR angiography (Choe, Kim, Koh, Do, & Lee, 1999). Active inflammation process in Takayasu arteritis is often detected in contrast-enhanced MRI as thickened aortic and carotid artery wall with extreme enhancement (Choe et al., 1999). Disease activity is often detected as contrast enhancement. Pulmonary artery thickening is also