Case report

An unusual clinical manifestation of Takayasu’s arteritis: Spinal cord compression

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Abstract

Takayasu’s arteritis is a chronic inflammatory vasculitis, involving mainly the aorta and its main branches and the pulmonary arteries, with characteristic of stenotic and occasionally dilated lesions. Neurologic manifestations of Takayasu’s arteritis range from simple headache to catastrophic neurologic impairments, including visual loss, stroke and transient ischemic attack. However, spinal cord compression has never been described as a complication of Takayasu’s arteritis. We describe a case of Takayasu’s arteritis complicated by spinal cord compression due to thoracolumbar inflammatory epiduritis.

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1. Case report

A 55-year-old woman was referred because of new onset of weakness of both low extremities and voiding difficulty for 2 weeks. Two months ago, she was admitted because of lower abdominal pain and fever. Abdominal computed tomography (CT) (Fig. 1a) revealed periaortic soft tissue attenuation with enhancement in long segmental abdominal aorta from renal hilum to aortic bifurcation level. She was diagnosed as chronic periaortitis and treated with high dose steroid (prednisolone, 1 mg/kg). Thereafter, the abdominal pain was improved and fever was subsided and she was discharged on day 18 of admission. And she was treated with tapered dose of steroid (prednisolone, 40 mg) before 2nd admission. She had no history of pulmonary tuberculosis, but a history of diabetes mellitus and hypertension for 6 months. Her blood pressure 140/90 mmHg, pulse rate 96/min, respiratory rate 20/min, and body temperature was 36.2 °C. A neurologic examination showed newly
developed grade 0/5 paraplegia below T8 area with decreased pinprick sensation on both sides. Her initial laboratory findings showed: white blood cell (WBC) 11,580/mm³, hemoglobin 10.8 g/dl, platelet 468,000/mm³, ESR 76 mm/h, and CRP 5.94 mg/dl (normal 0.02–0.80 mg/dl). Urine analysis revealed no RBC, WBC and casts. Antinuclear antibody, anti-double stranded DNA antibody, antineutrophil cytoplasmic antibody and viral hepatitis markers were all negative. The serum venereal disease research laboratory test (VDRL) was weakly positive. For evaluation of paraplegia, spine magnetic resonance imaging (MRI) (Fig. 1b) was taken and demonstrated ill-defined and well-enhanced inflammation from T7 to L1 level. She underwent emergency operation of laminectomy from T7 to T12 level and partial removal of dura mater for evaluation and treatment of cord compressive symptoms. Operative findings were thickening of dura mater and granulation tissues of thoracic and lumbar spinal cord. Microscopic examination of the lesion revealed areas of granulomatous pattern of inflammation with giant cells, involving blood vessel and adjacent soft tissue (Fig. S1; see the supplementary material associated with this article online). The histopathological findings were consistent with granulomatous vasculitis such as Takayasu’s arteritis or giant cell arteritis. For evaluation of aortitis, chest and abdominal CT (Fig. 1c,d) were taken and showed focal wall thickening of proximal right innominate and left subclavian arteries, diffuse wall thickening of aortic arch and focal stenosis of right proximal renal artery. Takayasu’s arteritis was considered based on the CT of chest and abdomen and histological findings. She was treated with high dose of corticosteroid (prednisolone, 50 mg a day orally) and intravenous cyclophosphamide therapy (1 g). As for the weakly positive VDRL, fluorescent treponemal antibody absorption test (FTA-ABS) was performed and showed two positive. Immunoglobulin M of FTA-ABS was negative. Subsequently, history suggested that the patient had a primary syphilitic lesion 30 years ago and was treated. However, lumbar puncture was taken to rule out neurosyphilis and showed a clear, colorless, acellular cerebrospinal fluid. VDRL and FTA-ABS of the cerebrospinal fluid showed negative. She was diagnosed as treated or latent syphilis and treated with intravenous penicillin. Her neurologic symptoms were slightly improved, evidenced by motor grade 1/5 and she was discharged on day 35 of admission. She was treated with monthly pulse cyclophosphamide. After 5th cyclophosphamide therapy, the steroid dose was slowly tapered with azathioprine.

Fig. 1. Imaging findings. (a) Abdomen computed tomography revealed eccentric, circumferential wall thickening of abdominal aorta. Abdominal aortic wall thickening was moderately enhanced. (b) Gadolinium enhanced T1-weighted magnetic resonance imaging revealed ill-defined and well-enhancing epidural lesion from T7 to L1 level with dural and leptomeningeal enhancement. Abnormal enhancement of paraspinal muscle from T11 to T12 level was also seen. (c) Chest computed tomography angiography showing focal wall thickening of proximal right innominate artery and left subclavian artery with mild luminal narrowing. Diffuse wall thickening of aortic arch was also noted. (d) Abdominal computed tomography angiography revealed focal stenosis at right proximal renal artery.
(50 mg a day orally). At 2-yr follow-up, neurologic examination showed much improvement with motor power of grade 4/5 on both lower extremities and laboratory test showed; ESR 54 mm/h, and CRP 2.07 mg/dl with low dose oral steroid (prednisolone 2.5 mg/day) and azathioprine.

2. Discussion

Neurologic manifestations of Takayasu’s arteritis range from simple headache to catastrophic neurologic impairments, including visual loss, stroke and transient ischemic attack [2—4]. The incidence of neurologic manifestations has previously been reported to be variable. If nonspecific symptoms such as headache and vertigo are included, the frequency rises to 90% [2]. In 1992, Wang [5] reported 38 cases of Takayasu’s arteritis and neurologic manifestation, and major neurologic events occurred in 52.7% patients in their group, including transient ischemic attack, cerebral infarction, hypertensive encephalopathy, lacunar infarct, seizure, paraplegia, watershed infarct, cerebral hemorrhage, moyamoya phenomenon and confusion in order frequency. Headache was the most common symptom of neurologic manifestations (55%). Kim et al. [3] reported 42% of frequency of neurologic manifestations, and suggested that the reason for the low frequency was exclusion of headache that is nonspecific and dizziness that does not lead to significant restriction of physical activity. In their study, dizziness was the most common neurologic manifestation (56%). These neurologic manifestations result mainly from decreased blood flow caused by steno-occlusive lesions and/or shifting of the blood flow. Other factors are hypertension and thromboembolism, leading to stroke. Spinal cord compression has never been described as a complication of Takayasu’s arteritis. In the literature, paraplegia among neurologic manifestation of Takayasu’s arteritis was described [5,6]. However, one case was spastic paraplegia due to cerebrovascular problem, and other case was not exactly described.

The differential diagnoses include other causes of large vessel vasculitis: infectious aortitis (syphilis and tuberculosis), noninfectious aortitis (lupus, rheumatoid arthritis, spondyloarthopathies, Behcet’s disease, Kawasaki disease and giant cell arteritis) and developmental abnormalities (coarctation of the aorta and Marfan’s syndrome) [7]. Most of these causes have specific features which make it possible to diagnose, however, syphilis has remained an important differential and possible etiologic factor in our case. Syphilitic aortitis tends to affect an older age group with calcification, sparing the descending thoracic aorta, and stenoses are not a feature. Furthermore, syphilitic aortitis without aneurysm is uncommon. Syphilitic aortitis involves the proximal aorta, and only occasionally extends below the renal arteries, probably because of the rich vascular and lymphatic circulation which is limited to the thoracic aorta. Also, classic histologic descriptions of the syphilitic aneurysms are medial necrosis and perivascular infiltrates, adventitial fibrosis, and patchy medial and adventitial lymphocytic infiltrations [8,9]. In our case, although our patient was older than 40 years at disease onset, mainly aortic lesions were stenoses without aneurysm, and her serologic status was treated syphilis. Furthermore, the histopathological findings of the lesion were consistent with granulomatous vasculitis.

Systemic vasculitis can present as variable compilations of symptoms and signs, which are sometimes difficult to diagnose, or even misleading and may present as a tumorlike lesion although not usual. In one report, seventy-nine cases of vasculitis presenting as a tumorlike lesion were found: the most common vasculitis categories with tumorlike presentation were Wegener’s granulomatosis (28 cases) and giant cell arteritis (17 cases), the most common location of a tumorlike lesion was the breast (22%), followed by central nervous system lesions (16%) [10]. In another reports, only two cases of Takayasu’s arteritis were described, and patients with symptoms caused by an expanding mediastinal mass and pituitary mass were presented [11,12]. Granulomatous vasculitis of tumorlike lesion associated with spinal cord compression has been reported in sarcoidosis, giant cell arteritis and Wegener’s granulomatosis with typical clinical or laboratory feature [13,14]. In our case, the patient had no involvement of nose, sinus, larynx, lung, kidney, lymph node or temporal artery, and arterial involvement was compatible with Takayasu’s arteritis. And the development of spinal cord compression nearly coincided with the appearance of symptoms and signs in large arteries. Also it is now known that Takayasu’s arteritis may affect small vessels as well although epidural vessel involvement has not been reported [15]. So we concluded that epidural granulomatous vasculitis of our case was probably developed from inflammation of Takayasu’s arteritis.

Our first attempt to control the activity of our patient’s with a high dose steroid therapy was unsuccessful because neurologic involvement was newly developed. Therefore, we started monthly intravenous cyclophosphamide therapy with combination of high dose steroid. After 5th cyclophosphamide and maintenance therapy of azathioprine, her neurologic symptom was much improved: from motor grade 0/5 to 4/5. Cyclophosphamide and azathioprine were both an effective in this case. Supplementary material

Supplementary material (Fig. S1) associated with this article can be found at http://www.sciencedirect.com, at doi: 10.1016/j.jbspin.2008.09.007.

References