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LETTERS TO THE EDITOR

# Takayasu's arteritis with peripheral nervous system involvement: description of a case and a review of the literature

## Arteritis de Takayasu con afectación del sistema nervioso periférico: descripción de un caso y revisión de la literatura

*Dear Editor:*

We present a 76-year-old female patient reporting a clinical condition lasting for 6 months and comprising pain, weakness and claudication of both upper limbs. A prior electrophysiological study (EPS) showed signs of peripheral nerve involvement (complete severe axonotmesis of the median, ulnar and radial nerves in the right arm and the median nerve in the left arm), because of which the patient had been diagnosed as having peripheral polyneuropathy. Her symptoms worsened and she came to our centre after the appearance of distal necrotic lesions on both hands. Examination on admission revealed an absence of pulses and low blood pressure figures in both upper limbs, an audible murmur on the left clavicular region, cold hands, necrotic lesions in the fingertips and distal paresis of the right arm. The laboratory findings showed a slight normocytic and normochromic anaemia, elevation of GSV (47 mm/h) and reactive C protein (76 mg/dL). An NMR angiogram revealed occlusion-stenosis of both subclavian arteries at the proximal-medial level (fig. 1) and a Doppler study of the supra-aortic trunks ruled out vascular involvement in other territories. A biopsy was effected on the temporal artery without any pathology findings. A diagnosis of Takayasu arteritis (TA) was reached as she met the diagnostic criteria established by the ACR in 1990.[1] In view of the presence of severe ischaemia, percutaneous treatment was performed with implantation of stents in both subclavian arteries, with excellent angiographic results and a speedy clinical improvement (disappearance of the necrotic lesions and recovery of blood pressure and peripheral pulse). Treatment was also started with prednisone (60 mg/day for one month followed by gradual weaning) and clopidogrel (75 mg/day indefinitely). After 7 months she presented a great improvement in her neurological symptoms (with slight persistence of paresis in the right hand), as well as in the follow-up EPS, showing partial axonotmesis of the affected nerves and signs of reinnervation. TA is a vasculitis of unknown aetiology mainly affecting the aorta and its main branches. The initial vascular involvement frequently occurs at the level of the subclavian artery, and when the condition progresses it may also affect other territories (carotid, vertebral, brachiocephalic trunk).[2] The initial phase of vascular involvement usually progresses with coldness, pain and claudication in the limbs. Only in advanced cases such as in the present patient may occlusion of the vessels cause ischaemic ulcers and necrotic lesions. In the event of coronary arteritis and dilatation of the ascending aorta, *angor pectoris* and aortic regurgitation, respectively, will appear. Other less frequent manifestations may involve the respiratory or digestive systems when the pulmonary or mesenteric arteries are affected.[3-5] Finally, neurological symptoms secondary to cerebral ischaemia may appear in cases of carotid and/or vertebral involvement.[6] The neurological manifestations of TA were revised by Wang, with cephalea as the most frequent symptom, followed by major neurological events secondary to ischaemia of the central nervous system (CNS), but no case of involvement of the peripheral nervous system (PNS) has been described.[7] There is also a report of an isolated case of compressive intracranial neuropathy secondary to dilatation of an intracranial artery.[8] Nadeau et al. and Moore et al. reviewed the neurological manifestations of systemic vasculitides, highlighting that PERIPHERAL NERVOUS SYSTEM involvement is relatively common in some of them, and more frequent in polyarteritis nodosa, microscopic polyangiitis, Churg-Strauss syndrome, Wegener's granulomatosis and cryoglobulinaemia. On the contrary, TA was only associated with involvement of the CNS secondary to ischaemia, but no case of involvement of the peripheral nervous system (PNS) was described.[9,10] We carried out a bibliographic search on Medline/PubMed using the following MeSH terminology: [Takayasu Arteritis] and [Medial Neuropathy], [Radial Neuropathy], [Ulnar Neuropathy], [Peripheral Nerves], [Peripheral Nervous System Diseases], [Mononeuropathy], [Polyneuropathy], [Neuritis] without finding any case described. The diagnosis of TA is infrequent in such advanced phases of the illness, as an adequate vascular examination when facing the first symptoms, revealing absence of pulse or reduction in arterial pressure, is usually enough to raise diagnostic suspicions in the initial phases, without the appearance of necrotic lesions of the ischaemic involvement of the peripheral nerves, as happened in the present case. To our knowledge this is the first case with involvement of the PNS described in the literature of TA.

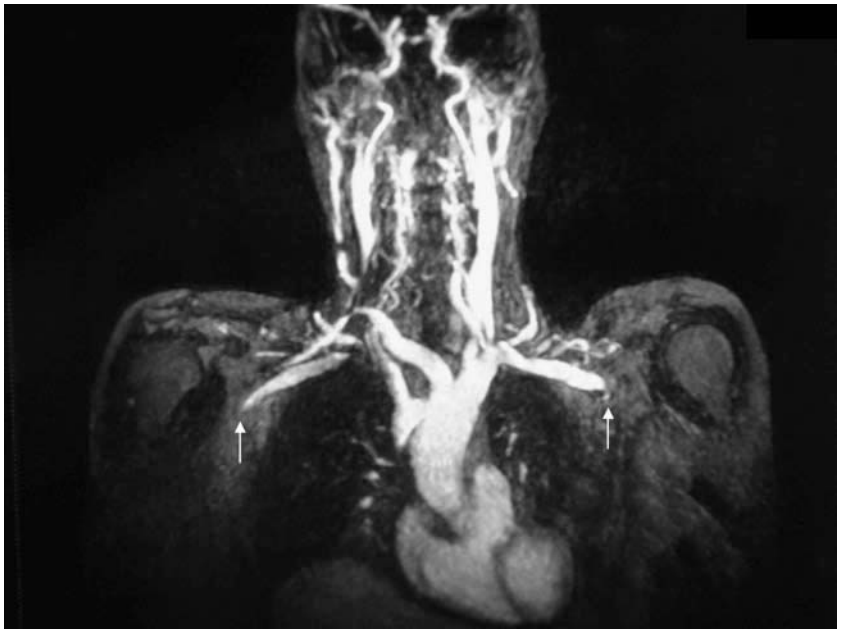

**Figure 1** NMR angiogram of supra-aortic trunks: bilateral occlusion-stenosis of subclavian arteries (white arrows).

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