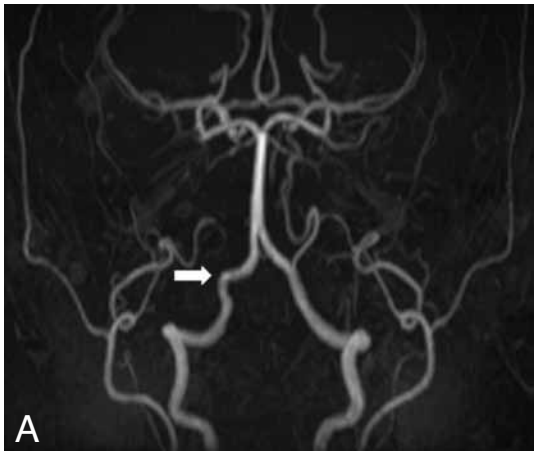
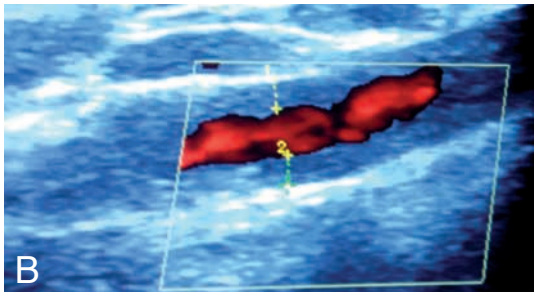
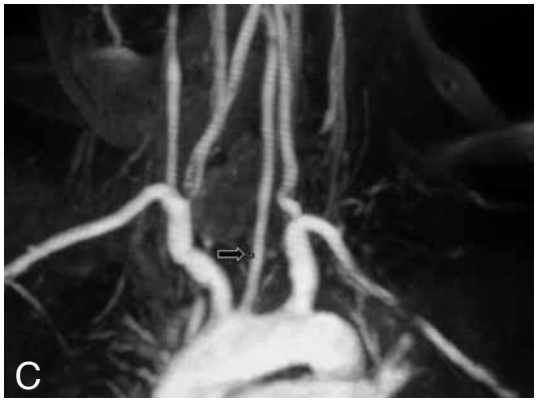
JBR–BTR, 2011, 94: 297.

IMAGES IN CLINICAL RADIOLOGY







The macaroni sign

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A 41-year-old woman was referred to radiology department for the evaluation of sudden paresis in the left side. Brain magnetic resonance imaging (MRI) showed multiple bilateral ischemic regions in the watershed areas. Also, the brain magnetic resonance angiography (MRA) showed bilateral internal carotid artery (ICA) occlusions at the level of the cervical ICA region. Both vertebral arteries were found to be enlarged (arrow) and the retrograde filling of the anterior circulation was observed (Fig. A). For the investigation and the differentiation of the bilateral carotid lesions, the color Doppler ultrasonography was obtained. This study showed homogenous, hypoechoic, circumferential wall thickening of both distal common carotid artery arteries (CCA). The thickening of the arteries wall were like 'Macaroni'. The wall thickness was 0.17 cm (Fig. B). The tapering of both CCA and total oclusion of both ICA were seen. The carotid MRA revealed collateral circulation from the subclavian arteries to the ECA and 50% stenosis of the left CCA (arrow) and 70% stenosis of the right CCA at its origin. Vertebral arteries were also prominant and there were 50% narrowing in the mid portion of the left subclavian artery (Fig. C). The radiological diagnosis of the Takayasu's arteritis (TA) was made and the patient was referred to the rheumatology clinic for further diagnosis and work-up.

Comment

TA is an granulomatous vasculitis of medium and large arteries which is commonly seen in young women. This disease has various clinical morbidities, such as arm claudication, decreased arterial pulses, carotidynia and hypertension as well as constitutional symptoms. It has three stages according to the symptomalogy. First, prepulsenessless period with systemic constitutional inflammatory complaints; second, vascular pain period characterized with carotidynia, and fibrotic period with ischemia and vascular murmurs. Diagnosis is made based on the widely accepted criteria of the American Collage of Rheumatology. The difference of systolic arterial pressure in both arms of more than 10 mm Hg, claudication of one extremity, weak brachial pulse, murmurs on the aorta and brachiocephalic arteries, younger than 40-year old and large artery involvement of the aorta and primary branches are the main diagnostic clues. At least three of these findings must have to be met for the diagnosis of the TA.

TA mostly involves the aortic arch and its branches in about 75% of the cases. The most consistent finding for a diagnosis of Takayasu's arteritis is angiographically determined bilateral occlusion of the subclavian and common carotid arteries. Occlusion of large arteries supplying the cephalic structures, without neurological symptoms, is a characteristic of this disease. In our case, there were bilateral ICA occlusions, both vertebral arteries compensated blood supply to the brain.

Rare ultrasonographic finding, previously described by Maeda et al., as the "macaroni sign", is pathognomonic for TA (1). This finding is secondary to the mononuclear and giant cell infiltration of the media and adventitia of the medium and large sized arteries that leads external elastic lamina degeneration with intimal proliferation.

Color Doppler US is diagnostic for TA. Ultrasonography and angiography are complementary methods for the diagnosis. While angiography shows extended areas of luminal changes, ultrasonography enables the characterization of the wall changes in a limited area.

Reference

1. Maeda H., Handa N., Matsumoto M., Hougaku H., Ogawa S., Oku N., Itoh T., Moriwaki H., Yoneda S., Kimura K., et al.: Carotid lesions detected by B-mode ultrasonography in Takayasu's arteritis: "macaroni sign" as an indicator of the disease. *Ultrasound Med Biol*, 1991, 17: 695-701.

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