The “Macaroni Sign” of Takayasu’s Arteritis

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Takayasu’s arteritis is a large-vessel vasculitis occurring mainly in adolescent girls and young women. Ultrasound of the carotid and subclavian arteries can aid in early diagnosis of the disease.

A 19-year-old woman presented with a 4-month history of fatigue, weight loss, and thrombocytosis. Examination showed a difference of 40 mm Hg in systolic blood pressure between the 2 arms, together with a decreased pulse in the left. Noteworthy laboratory results included erythrocyte sedimentation rate 100 mm/h and C-reactive protein 50 mg/l (normal < 5). Color Doppler ultrasonography of the upper-limb extracranial arteries showed homogeneous mid-echoic circumferential wall-thickening of the common and internal carotid arteries of both arms (Figure 1A) and the vertebral (Figure 1B) and axillary arteries and the proximal part of the brachial artery of the left side (Figure 1C). Intracranial ultrasound was normal. The examination was performed using an AT5500 device with a 12-5 MHz linear probe for extracranial vessels and a 2-4 MHz array probe for intracranial vessels. Flow was detected using the power Doppler mode.

Positron emission tomography (PET) showed inflamed areas on the common carotid artery.

This uncommon ultrasonographic finding is typical of Takayasu’s arteritis and has been designated the “macaroni

sign”^{1,2}. This finding is brighter than the typical “dark halo” sign of giant cell arteritis. Current ultrasound technology allows visualization of large vessels and discrete vessel-wall alterations, and also distinguishes inflamed from atherosclerotic wall lesions. In Takayasu’s arteritis, angiography can show important luminal stenoses in many vessels including small arteries. However, this technique cannot visualize the vessel wall and may not reveal mild wall lesions without stenosis. PET shows inflamed arterial areas but cannot show information on vessels with diameter < 4 mm and details of the vessel wall.

Thus, ultrasound represents an adequate method for diagnosis of Takayasu’s arteritis in young patients with chronic inflammation of unknown origin³.

REFERENCES

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Figure 1. Color duplex ultrasound showing the “macaroni sign” (homogeneous, midechoic, and circumferential wall-thickening together with stenosis) at the common and internal carotid (A), vertebral (B), and brachial (C) arteries. Figure 1A represents a more acute phase of disease and Figure 1C a more chronic state. The stenosis was hemodynamically more relevant in Figure 1C (peak systolic flow velocity 226 cm/s).