

# Macaroni sign: a clue to Takayasu arteritis diagnosis

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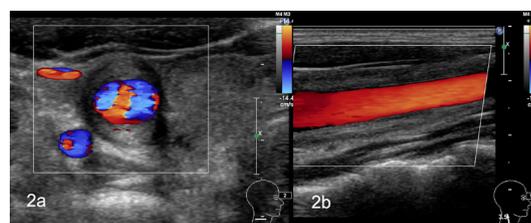
## DESCRIPTION

Takayasu arteritis (TKA) is a chronic systemic inflammatory autoimmune disease characterised by inflammation of the aorta and major branch vessels, possibly leading to several symptoms, including fever, weight loss, joint pain and fatigue.<sup>1</sup> Carotid artery (CA) pain or carotidynia is a significant TKA symptom resulting from carotid involvement and subclavian arterial vascular lesions.<sup>2</sup> CA lesions in TKA, known as the 'macaroni sign', are characterised by global thickening of the common CA wall and lumen narrowing, and can be observed using carotid ultrasound.<sup>3</sup>

Herein, we report a TKA case diagnosed based on the presence of the 'macaroni sign' on carotid ultrasound.

A female adolescent patient, without relevant medical history, visited the hospital with complaints of fever and neck pain that started 90 days before her visit. At admission, her body temperature, blood pressure and heart rate were 36.8°C, 112/60 mm Hg on her right arm (left, 84/60 mm Hg) and 76 beats/min, respectively. The pulse in her left radial artery was not palpable. Physical examination revealed spontaneous pain and tenderness on both sides of the patient's neck. Other physical abnormalities were absent.

Blood test results demonstrated an elevated inflammatory response C-reactive protein, 5.2 mg/dL (normal range: less than 0.3 mg/L); erythrocyte sedimentation rate (ESR), 95 mm/hour). We suspected TKA and performed a carotid ultrasound, which revealed marked thickening of the intima-media of the bilateral common CAs and a positive macaroni sign (figure 1, figure 2). Detailed evaluation of the systemic vessels was conducted, and the patient was diagnosed with TKA type IIb. The patient was administered oral steroids at a dose of



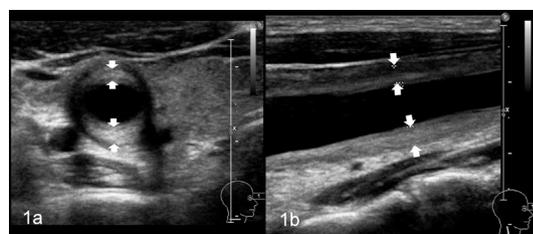
**Figure 2** Colour Doppler images. (a) Transverse image. (b) Longitudinal image.

0.8 mg/kg and thereafter showed marked improvement with continuing treatment.

The clinical manifestations of TKA vary depending on the vascular lesions. TKA diagnosis is often delayed in relatively young women (median, 17.5 months) and becomes extremely complex in older women.<sup>4</sup> The keys to TKA diagnosis are CA involvement and carotidynia (57% and 81% of cases, respectively),<sup>1</sup> which must be considered in young women experiencing neck pain, to avoid a misdiagnosis. TKA can be diagnosed with B-mode ultrasonography of the common CA, to detect the presence of the 'macaroni sign', which is a characteristic TKA feature.

The intima-media normally thickens with age. Generally, an intima-media thickness of  $\geq 1.0$  mm is considered an abnormal thickening and a positive finding. A sensitivity of 81%, specificity of 90%, positive likelihood ratio of 8.1 and negative likelihood ratio of 0.21 add to its accuracy.<sup>5</sup> Details of other carotid pathologies diagnosed using carotid ultrasound are summarised in table 1.

Although MRI is usually recommended for the definitive diagnosis of TKA,<sup>6</sup> it may not be easily accessible for all cases. However, TKA diagnosis based on the 'macaroni sign' identified by carotid ultrasound is a low-cost, simple and non-invasive



**Figure 1** 'Macaroni sign' in the right common carotid artery detected by carotid ultrasound. We used linear probes with a B-mode frequency of 9 MHz and image depth of 30 mm. Long, homogeneous, isoechoic, circumferential wall thickening of the common carotid arteries is demonstrated. (a) Transverse image (maximum, 3.1 mm; normal range,  $<0.6$  mm). (b) Longitudinal image (maximum, 3.1 mm; normal range,  $<0.6$  mm).

**Table 1** Differential diagnosis of global thickening of the common carotid artery wall and lumen narrowing

|                          |   |
|--------------------------|---|
| Takayasu arteritis (TKA) | <ul style="list-style-type: none"> <li>▶ Intima-media complex thickening</li> <li>▶ Multilayered</li> <li>▶ Circumferential (early stage may be localised)</li> <li>▶ Mostly occurs within 40 years of age</li> </ul> |
| Arteriosclerosis         | <ul style="list-style-type: none"> <li>▶ Intima-media complex thickening localised plaques</li> <li>▶ Mostly occurs over the age of 50 years</li> </ul>   |
| Giant cell arteritis     | <ul style="list-style-type: none"> <li>▶ Intima-media complex thickening</li> <li>▶ Same as TKA but quite rare</li> <li>▶ Mostly occurs over the age of 50 years</li> </ul>   |



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method that can be easily performed during initial hospital visits. In addition, a clear correlation between MRI and CT angiography findings and disease activity or progression has not been found for TKA yet. Therefore, methods using contrast-enhanced ultrasound in combination with ESR are considered.<sup>5</sup>

In middle-aged and older patients with TKA, stringent distinction is difficult because of the presence of concomitant atherosclerotic lesions. However, temporal arteries are not affected in TKA, an important factor in differentiation between TKA and giant cell arteritis.

### Learning points

- ▶ In younger women with neck pain, the possibility of carotidynia due to Takayasu arteritis (TKA) should be considered.
- ▶ The 'macaroni sign' is a useful diagnostic feature of TKA in carotid ultrasound.
- ▶ Carotid ultrasound can help prevent delays in TKA diagnosis in settings with limited resources.

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Case reports provide a valuable learning resource for the scientific community and can indicate areas of interest for future research. They should not be used in isolation to guide treatment choices or public health policy.

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