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Diagnosis of non-atherosclerotic carotid disease

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Summary Non-atherosclerotic carotid disease in an uncommon group of angiographic defects. It includes the entities: Takayasu's arteritis, giant cell arteritis, fibromuscular disease, moyamoya syndrome, arterial dissection and extracranial carotid aneurysms. Due to advance in imaging techniques, they are being increasingly identified. Growing awareness of diverse clinical picture along with advances in imaging technologies enables early diagnosis. Although catheter angiography is a gold standard in diagnosing most of these diseases, neurosonological tests serve as an excellent screening tool, and are suitable for monitoring. Brain MR and MRA are sometimes essential for confirmation of the diagnosis. Mortality rates are low and functional outcome is generally good if the disease is diagnosed early.

Takayasu's arteritis

Takayasu's arteritis is a granulomatous arteritis affecting the aorta and its branches [1]. Its incidence is estimated at 2.6 cases per million per year, more common in Southeast Asia. It is more prevalent in young woman (9 females:1 male). It has three stages. During the systemic stage symptoms and signs of an active inflammatory illness dominate, like e.g. malaise, fever, night sweats, arthralgia, weight loss, anemia and elevated erythrocyte sedimentation rate. The systemic phase is succeeded by the vascular inflammatory stage, when stenosis, aneurysms, and vascular pain (carotidynia) tend to occur. During this phase patients begin to develop symptoms caused by the narrowing of affected arteries. Symptoms are caused by the narrowing of affected arteries like stroke, transitory ischemic attack (TIA), claudication, dizziness, headache, visual symptoms and hypertension as a result of stenosed renal arteries. This stage sometimes overlaps with the systemic stage. At the end a burned-out stage develops when fibrosis sets in, and this stage is usually associated with remission. According to the American College of Rheumatology [2] the criteria for assessing the diagnosis are: angiographic criteria displaying narrowing or occlusion of the entire aorta, its primary branches, or large arteries in the proximal upper or lower extremities. These changes are not due to arteriosclerosis, fibromuscular dysplasia, or similar causes; changes are usually focal or segmental; the lesions can include stenosis, occlusion, or aneurysms. Angiogram is a gold standard, but sonography assesses both vessel anatomy and luminal status in accessible areas and can detect early vessel wall alterations before lumen changes on angiography [3—6]. Its advantage is limited cost, short time required, and there is no radiation. Due to noninvasiveness, it is suitable for monitoring. Direct or indirect signs can be visualized. Color Doppler flow imaging enables visualization of the mural thickening of the common carotid arteries (Fig. 1), hypoechoic in the early, vascular inflammatory stage [7]. With the development of fibrosis, pronounced echogenicity of the lesions develop in the burned-out stage. Due to inflammation, stenosis occurs. If advanced stenosis affects the brachiocephalic trunk or origin of the left common carotid artery, changed hemodynamic spectra like

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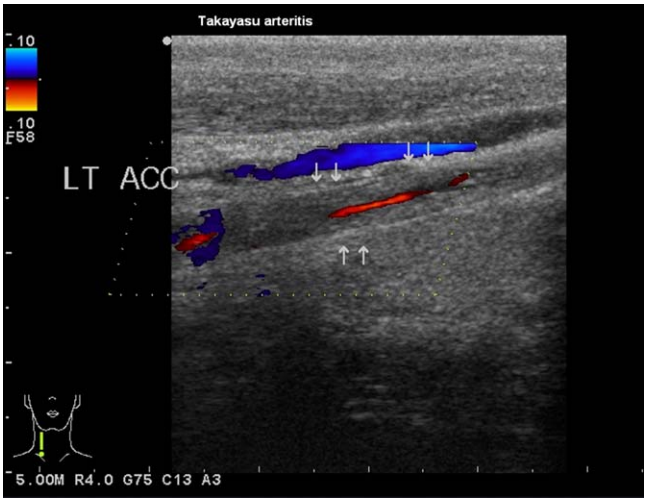

Figure 1 Takayasu arteritis showing advanced mural thickening of the common carotid artery to the level of tight stenosis.

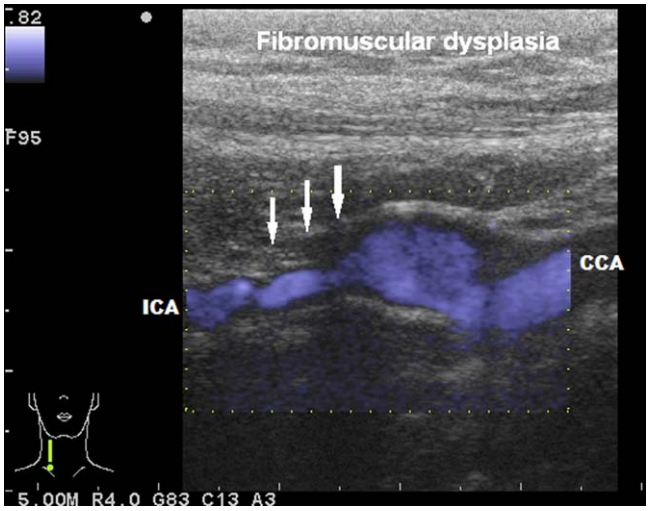

Figure 2 Segmental narrowing and widening or the color coded flow in internal carotid artery showing characteristic ''ring of beads'' appearance in medial type of fibromuscular dysplasia.

dampened flow in carotid artery, distal from the stenosis, can be recorded. If the stenosis affects subclavian artery, changed hemodynamic spectra suggesting subclavian steal syndrome are recorded (Fig. S1 supplementary file). When occlusion of the subclavian artery sets in, in ipsilateral vertebral artery hemodynamic spectra are completely inverse (Fig. S2 supplementary file), and in the contralateral one it is accelerated. Transcranial Doppler of the Willis circle and vertebrobasilar system shows redistribution of the hemodynamics.

Giant cell arteritis — GCA

GCA, is also known as temporal arteritis or cranial arteritis, is the most common form of vasculitis that occurs in adults [8]. Almost all patients who develop GCA are over the age of 50. It is a granulomas arteritis affecting large or medium-sized artery, usually temporal or ophthalmic artery. It has an acute or subacute start. Symptoms are headache, jaw pain, blurred or double vision. If the disease is undiagnosed complications like blindness and, less often, stroke may occur. Standard test for diagnosing GCA is biopsy of the temporal artery. More samples are needed because the inflammation may not occur in all parts of the artery. Prompt treatment with corticosteroids relieves symptoms and prevents loss of vision. Ultrasound finding will show swelling of the arterial wall presenting as a hypoechoic dark halo around the color coded flow in the temporal, ophthalmic artery or external carotid artery [7,9]. The disease is segmental, therefore, its visualization is suitable for localization of the biopsy. Due to noninvasiveness it is suitable for monitoring the disease. During healing regression of the dark halo will be visible parallel with the restitution of the color coded flow.

Fibromuscular dysplasia

Fibromuscular dysplasia (FMD) is a fibrous thickening of the arterial wall, causing segmental narrowing of arteries in the kidneys (in 75% of patients), carotid or vertebral arteries and the arteries of the abdomen [10]. It is an autosomal dominant disorder, affecting up to 5% of the population, in 2/3 the internal carotid artery (ICA), usually the C2 segment. It is usually asymptomatic, but if dissection occurs, it causes aneurysm and occlusion and becomes symptomatic. There are three types of fibromuscular dysplasia: intimal, medial, and subadventitial (perimedial) of the arterial wall. These three types of FMD are not easily differentiated by findings on angiography. The medial type of FMD is by far the most common (about 80—85%) and it is classically diagnosed on the basis of a ''string of beads'' appearance on angiography. This appearance is explained by the presence of luminal stenosis alternating with aneurysmal dilatation. Classically, the intimal form of FMD is associated with smooth focal stenoses on angiography. Type 1 is the most common form. In 6—12% of patients with arterial fibroplasia, a long tubular stenosis may be seen. This form is most commonly seen with the intimal form. The unusual form (seen in 4—6% of patients) is characterized by involvement of only one side of an artery. Such involvement leads to diverticularizations of the arterial wall. These lesions may be difficult to distinguish from atherosclerotic ulceration and pseudoaneurysm. Ultrasound findings correlate with the angiographic findings, and may show segmental narrowing and widening or the color coded flow in carotid or vertebral arteries, with the characteristic string of beads appearance in medial type of FMD, long tubular stenosis, usually distally from a widened carotid bulb in intimal type of FMD, or irregular local widening of the arterial wall in subadventitial type of FMD. Fig. 2 shows ''string of beads'' appearance in medial type, and Fig. S3 supplementary file shows occlusion of the internal carotid artery after the carotid bulb as a result of dissection in intimal type (Fig. S3 supplementary file).

Moyamoya disease

Moyamoya disease is an inherited genetic abnormality causing intimal thickening in the walls of the terminal portions of the internal carotid vessels bilaterally and stenosis