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CLINICAL VIGNETTE

Use of Temporal Artery Ultrasound for Diagnosis of Giant Cell Arteritis

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Case

A 70-year-old male with hypertension, diabetes, and hyperlipidemia, presented with two-weeks of headache. He described the headache as pressure-like and associated with a dull ache throughout the entire head. He noticed severe pain and tenderness in the area of the temples. He denied any vision changes or jaw claudication. The patient did endorse some psychosocial stressors prior to developing these symptoms. He noticed some improvement in the headache with taking aspirin/paracetamol/caffeine (Excedrin). Past medical history includes anxiety and 15 pack-year history of tobacco use. Physical exam revealed elevated blood pressure of 153/71 mmHg and heart rate of 94/min. He was afebrile and his respiratory rate and oxygenation were normal. Patient had tenderness in the temples bilaterally, greater on the right side compared to the left. Bilateral temporal artery pulsations were normal. Cardiovascular and lung exams were normal. Pupils were reactive and extraocular muscles were intact. Labs were obtained and revealed a sedimentation rate of 60 mm/hr (normal < 12mm/hr) and C-reactive protein of 6.3 mg/dL (normal <0.8mg/dL). Based on these findings, his primary care doctor started prednisone 60 mg daily and referred to Rheumatology for Giant Cell Arteritis (GCA). The primary care doctor also ordered a color doppler ultrasound of the bilateral temporal arteries. Ultrasound was done the next day and showed thickening of the right parietal and left frontal temporal artery rami, which the radiologist noted may be supportive of possible temporal arteritis.

The patient was seen by Rheumatology and advised to continue prednisone 60mg daily and was scheduled for a temporal artery biopsy to confirm his diagnosis. Patient did notice improvement in symptoms with steroids. The sedimentation rate decreased to 11 mm/hr (normal <=12mm/hr) and C-reactive protein went down to 0.3 mg/dL (normal <0.8mg/dL). Temporal artery biopsy was completed and showed non-specific intimal hyperplasia but no evidence of active or healed arteritis. Patient was advised to taper prednisone given the lack of vasculitis seen on biopsy. A CT angiogram (CTA) of neck, chest, abdomen and pelvis was scheduled to evaluate for possible extra-cranial disease. It showed no evidence of active or chronic arteritis. The CTA neck showed moderate scattered densely calcified atherosclerotic plaques with varying degrees of narrowing. Severe narrowing was noted at the left internal carotid artery origin and right V1 and V2 segments. A 5 x 3 x 7 mm penetrating atherosclerotic ulcer was seen at the left carotid bulb. The CTA chest, abdomen, and pelvis showed calcified and noncalcified atherosclerotic plaques throughout the thoracic and abdominal aorta and branch vessels and calcified plaques at the origin of the bilateral renal arteries with mild narrowing of the right main renal artery and moderate narrowing of the left main renal artery. There was also calcified coronary atherosclerosis involving the left anterior descending and the left circumflex artery and aneurysmal dilatation of the ascending thoracic aorta measuring 4.0 cm. Given the CTA findings, the patient was referred to a neurovascular specialist and a cardiologist. He was able to taper off prednisone without return of any symptoms. After consultation he started rosuvastatin 40 mg daily and continued losartan. An echocardiogram was scheduled to monitor the dilated ascending aorta.

Two weeks after tapering off steroids, he started to develop scalp tenderness. He denied any headache, vision changes, jaw claudication, fatigue, chest pain or shortness of breath. With concern for possible recurrence of the presumptive diagnosis of GCA, inflammatory markers were rechecked. The sedimentation rate was 33 mm/hr (normal <=12mm/hr) and the c-reactive protein was 5.4 mg/dL (normal <0.8mg/dL). 60 mg prednisone was restarted.

After two days of steroids, the patient developed acute onset of shortness of breath and chest pain. He also noted increased fatigue and a mild cough for two days. He was admitted and found to have rising elevated troponin of 91 ng/L (normal <15ng/L) and an elevated pro-B-type natriuretic peptide (BNP) of 10148 pg/mL (normal 0-900 pg/mL). EKG did not show ST segment elevation. Cardiology performed a left heart catheterization which revealed 3-vessel coronary artery disease. His elevated c-reactive protein was thought to be related to cardiovascular disease rather than GCA. Given the multivessel cardiovascular disease, the patient underwent coronary artery bypass grafting (CABG).

Discussion

This case illustrates the complexities in diagnosing and monitoring disease activity in GCA. It also demonstrates the utility of color duplex ultrasonography in the diagnosis of GCA.

GCA is the most common systemic vasculitis in adults over age 50.[1] The exact pathogenesis is unknown but there is a higher incidence in populations of Northern European descent.[2] It predominately affects large- and medium-sized blood vessels,

including the aorta and its major branches. The temporal artery is the most common cranial artery affected.[2] GCA causes inflammation of the entire artery wall in a segmental and focal distribution with or without multinuclear giant cell granulomas.[3] The inflammation can cause thickening of the arteries which can lead to vascular remodeling and occlusion with risk of sudden irreversible blindness and stroke.[2] Early diagnosis and treatment is essential to prevent these permanent changes.

Symptoms of GCA can be heterogenous depending on the vessels affected. Patients can present with many different symptoms including new-onset headache, polymyalgia, jaw claudication, scalp tenderness, fatigue, and visual disturbances in patients with cranial vessel involvement.[3] Limb claudication, fever, weight loss and other non-specific symptoms can be seen in those with extra-cranial large vessel involvement.[3] Diagnosis can be difficult due to non-specific symptoms and heterogenous phenotypes of the disease.

The 1990 American College of Rheumatology (ACR) classification criteria are helpful to diagnose GCA. The criteria include age >50, new onset headache, high sedimentation rate (ESR), abnormal temporal artery on palpation, and temporal artery biopsy with changes consistent with GCA.[4] Temporal artery biopsy has been and remains the gold standard for diagnosis of GCA. It is important to note that a negative temporal artery biopsy does not exclude the diagnosis of GCA. Due to the segmented pattern of inflammation in GCA and effects of steroid use while awaiting biopsy, the false negative rate is 5-13%.[5] The biopsy is an invasive procedure and although it is low risk, surgical risks like infection, bleeding and nerve damage are still present.[6]

Recent studies have demonstrated the utility of non-invasive vascular imaging modalities in GCA. Particularly, color doppler vascular ultrasound has been shown to be useful in the rapid diagnosis of GCA with temporal artery involvement.[7] The most specific finding on ultrasound is a halo sign. This is a homogenous, hypoechoic wall thickening of the artery seen in both transverse and longitudinal planes.[8] The European League Against Rheumatism (EULAR) recommends ultrasound of temporal arteries as the first imaging modality in patients with suspected cranial GCA.[8] A positive ultrasound result has 77% sensitivity and 96% specificity.[8] It also allows full length visualization of arterial wall abnormalities which can be helpful given the segmented nature of the disease.[8] There are risks of a false-positive halo sign in patients with other types of vasculitis, infection, or severe arteriosclerosis.[8] Additional studies are needed to evaluate the effects of atherosclerosis on temporal artery wall thickness such as our patient's ultrasound.[2]

Given the high specificity of halo sign on ultrasound, the updated 2022 American College of Rheumatology/EULAR classification criteria for Giant Cell Arteritis include positive halo sign on temporal artery ultrasound as part of the classification criteria.[9] Depending on the clinical setting, a halo sign on ultrasound may replace the need for temporal artery biopsy based on the updated classification criteria.[9]

Conclusion

This vignette demonstrates how ultrasound can be helpful to aid in the diagnosis of GCA. Although our patient did not have the classic halo sign on ultrasound and required further investigation with temporal artery biopsy, ultrasound provided a rapid and non-invasive way of examining the vessels without increased risk of complication. After additional vascular imaging with CTA, it was evident that significant atherosclerotic disease was present and could be confounding the temporal artery wall thickness seen on ultrasound, making the diagnosis of GCA less likely in this patient.

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