Imaging findings in extracranial (giant cell) temporal arteritis

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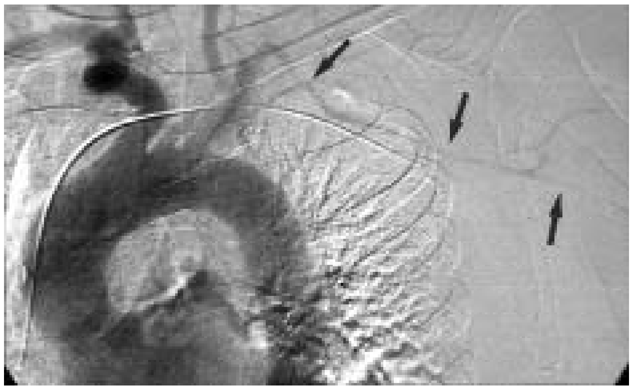
ABSTRACT
Patients with extracranial giant cell arteritis (GCA) present with occlusive arterial lesions that may be detected with multiple imaging modalities: arteriography, intravenous angiography (IV-DSA), CT scanning, and magnetic resonance angiography (MRA).
The lesions often present with a typical arteriographic pattern of bilateral stenoses or occlusions with a smooth, tapered appearance in the subclavian, axillary and proximal brachial arteries. A few patients have aneurysmal lesions. Less commonly involvement may be found in the femoral arteries and their branches. Angiographic study of 65 patients (56 women, 9 men; average age, 65) revealed involvement of the upper extremities in 61 patients, and lower extremities in 13, while 9 had both areas affected.
Detection of these lesions requires a diagnostic modality that depicts the vessel lumen such as: angiographic techniques, CT scanning with reconstructed images, and MRA. However, inflammation of the arterial wall cannot be detected by these means. Standard CT imaging with contrast enhancement, and certain MR sequences as well as ultrasound permit identification of the edema and inflammation of the vessel wall. This is an important marker for active disease.

Imaging findings in extracranial (giant cell) temporal arteritis
Patients with extracranial arterial involvement of Temporal (Giant Cell) Arteritis (GCA) have an opportunity to have their diagnosis confirmed or established by examining these arteries with the imaging modalities of angiography, CT scanning, Ultrasound, and Magnetic Resonance imaging techniques. Each modality offers its unique features to display these arterial lesions. To better understand the imaging findings of the various modalities, an understanding of the vascular pathology at the macroscopic level is an important starting point. The major feature of the arterial lesions is some degree of occlusion. And, rarely, aneurysmal changes may be found (Fig. 1). In the acute state there is also inflammation in the wall of the artery.
Angiography has traditionally been the diagnostic modality used, but with increasing reliance upon non-invasive procedures, there has been increased utilization of ultrasound, CT and MR. Each imaging modality has its own capacity to display one or more of these features.

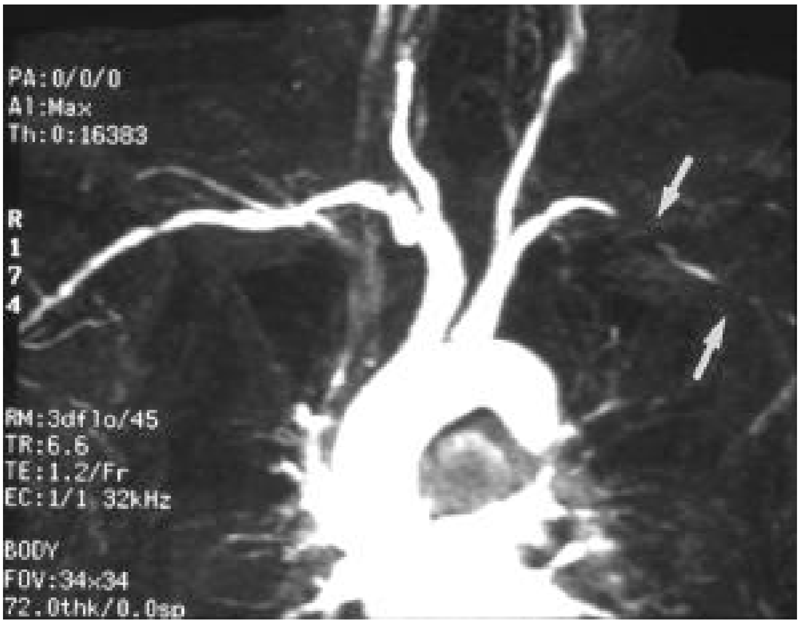
Angiography
Angiography may be performed in one of two ways: intravenous, or intra-arterial. The intravenous method provides

Fig. 1. GCA in a 67-year-old female. Aortic arch injection shows bilateral subclavian and axillary artery lesions of stenosis and early aneurysm formation. In the subclavian artery segments, the disease is present distal to the vertebral artery origins.

(a)



(b)



(c)

Fig. 2. GCA in a 73-year-old female. (**a**) Digital subtraction arteriogram in right anterior oblique projection obtained by injecting contrast material into the right atrium from a catheter (curved arrow) placed from the left arm. Note the irregularity of the lumen with a slight narrowing of the right axillary artery (top arrow). There is a higher grade narrowing as the axillary artery approaches the brachial artery (bottom arrow). (**b**) Left anterior oblique projection showing the high-grade stenosis of the left subclavian and axillary artery segments continuing into the proximal brachial (arrows). (**c**) Magnetic resonance angiography with gadolinium injection in AP projection shows the bilateral subclavian and axillary artery stenotic disease. Note that on the left side (arrows) the high-grade stenotic lesions of the subclavian and axillary arteries appear to be occlusions on the MRA image.

Fig. 3. GCA in a 65-year-old female. Intravenous digital subtraction arteriogram demonstrates axillary artery involvement. The degree of narrowing is mild. There is minimal narrowing at the right subclavian-axillary junction (top arrow) and at the axillary-brachial artery junction (bottom arrow).

less spatial and contrast resolution of the arteries than the intra-arterial technique but it is adequate for diagnosing relatively advanced states of the extracranial GCA (Figs. 2 and 3). The technique is simpler and less invasive than puncturing the femoral artery and advancing a catheter into the ascending aorta. Either a large bore needle is placed into the antecubital vein or a small catheter is used from the same site and advanced into the right atrium where the injection is made. The angiographic filming records the circulation of the contrast material after passage through the lungs. The digital filming is obtained over the aortic arch and the brachiocephalic branches. The proximal brachial arteries must be included in the field to be certain to detect disease involvement at this important location. Often the most advanced occlusive findings occur at these peripheral zones.

The location and geometric appearance of the occlusive arterial disease process in GCA is a very important feature in recognizing this disease and distinguishing it from many other pathologic entities (1, 2). The most frequently involved arterial segments are the proximal upper extremity branches, but it does not extend to the primary aortic branches as detected by arteriography. The disease process manifests bilateral involvement, but it is rarely symmetric in appearance. Stenoses and occlusions are the important findings in the medium size arteries, but ectatic areas may occasionally be found. Stenotic lesions, however, are the dominant feature. Their appearance and distribution are critical in making the diagnosis. They are smoothly tapered in appearance, both at the leading and trailing ends. And when occlusion is present, this tapered appearance persists. The involved artery may predominantly be a long zone that maintains a uniformly long, smooth stenosis. Stenotic areas, short or long, may have severe caliber reduction to the point that a near occlusion exists. Another specific feature is that zones of caliber reduction often have an alternating pattern with intervening segments showing near normal diameter. The location of the lesions is usually limited to the upper extremities and involves the distal subclavian, any portion of the axillary, and the proximal most portions of the brachial artery segments. More distal segments may be involved, but only if the more proximal areas show disease. However, it is not rare to observe similar angiographic findings in the small branches of the subclavian and axillary arteries. In cases with occlusions or severe stenoses, bridging collateral arteries are always present, but in the presence of recent onset of disease, they may not be prominent. Therefore, unless an adequate volume of contrast material is injected and the filming is prolonged, the distal refilled segments may not be observed.

The femoral arteries and their branches may also be involved with angiographic evidence of GCA, although they are not as frequently involved as the upper extremity arteries (Figs. 4, 5) (3). The lower extremity arterial segments show similar angiographic changes as those found in the aortic arch branches. The small branches of the deep femoral arteries are often found to be extensively involved. In contrast, it is unusual for the small branches of the subclavian and axillary arteries to be severely affected.

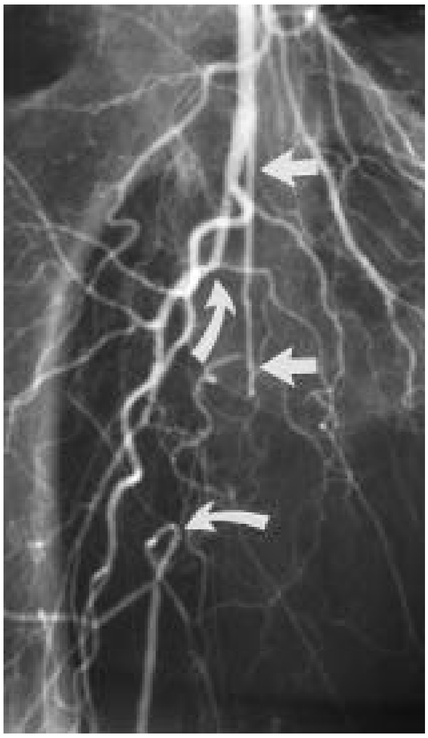


Fig. 4. GCA in a 63-year-old female. Right femoral arteriogram shows occlusion of a long segment of the deep femoral artery (curved arrows). There is high-grade diffuse stenosis of the proximal super ficial femoral artery (arrows) which is occluded distally.

Fig. 5. GCA, 64-year-old male. Left femoral arteriogram shows high-grade, smooth, tapered stenosis involving the distal popliteal artery (arrows). The stenosis tapers at the lower margin of involvement into the posterior tibial and peroneal arteries (curved arrows).

Table I. Arterial segments.

	Subclavian R	Subclavian L	Axillary R	Axillary L	Brachial R	Brachial L
Stenosis	35	40	34	35	21	26
Occlusion	8	6	4	5	7	5

Arterial ectasia was present in 3 patients.
Note: 9 of these patients also had lower extremity angiograms.

In this series of 65 patients with angiograms available for review, 56 were women and 9 were men. The average age was 65. The upper extremities were studied in 61 patients and the lower extremities in 13 (Table I). In 9 patients, both the upper and lower extremities were studied by angiography. The majority of the patients had stenotic disease, followed in incidence by occlusions and only 3 patients had areas of ectasia. In the femoral artery distribution, all 13 patients had some degrees of stenoses and in 8 patients occlusions were also found. The disease was detected in the tibial arteries in 3 patients. The true incidence of lower extremity arterial involvement was not determined by this review because of the nature of the referral pattern and the practice guidelines for angiographic procedures. Disease may have been present at a subclinical level but would have been undetected because the request for angiography of the lower extremities was not made. In such cases, the angiogram was limited to the symptomatic areas, e.g., the upper extremities. The same is true for those 4 patients in whom only the lower extremity angiogram was requested; perhaps upper extremity involvement also would have been detected had angiography included that area.

Aortic lesions found by arteriography are the least common of all other arterial segments in the acute setting. Diffuse aneurysmal alteration of the thoracic aorta may be found as well as a focal, false aneurysm formation secondary to granulomatous destruction of the aortic wall. A better diagnostic method to detect aneurysmal disease is CT or MR scanning. An additional benefit of these imaging methods is their ability to detect the direct finding of inflammation of the aortic wall, an indication that the disease is acute.

Aortic involvement is more commonly found to be a late feature of GCA. Within a time span of several years, aortic aneurysms or dissections are found in at least 10% of patients with GCA overall, and is not limited to just those patients in the extracranial GCA group (Fig. 6) (4). Although angiography is capable of displaying these pathologic conditions of aneurysm and dissection, the best imaging modalities to evaluate them are CT and MR scanning because of their non-invasive nature and lower cost.

The angiographic findings of GCA are somewhat unique and in the absence of advanced arteriosclerosis the diagnosis is not difficult after initial experience with a few cases and careful correlation with the clinical presentation. However, the similarity of the angiographic findings between GCA and Takayasu's disease can be difficult to distinguish unless global arteriography is performed. An important distinction is that patients with Takayasu's disease virtually always have aortic involvement manifested by some degree of narrowing in most cases and ectasia in about 25% of patients. Of course, the ages of the two patient populations is critical to know. Typically, the onset of GCA occurs decades after that of Takayasuís disease.

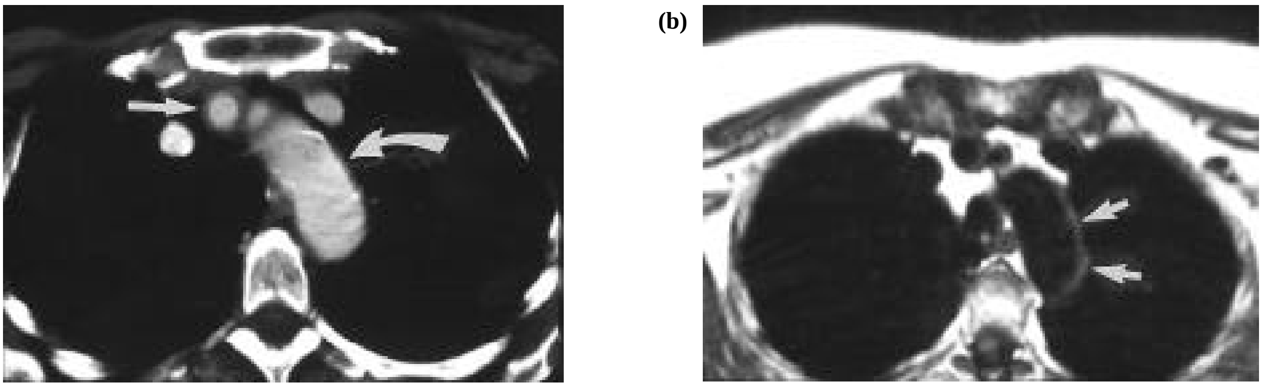
Computed tomographic (CT) scanning can also be used to detect extracranial GCA, but only in certain arterial segments. The limitation is related to the orientation of the acquired images. Arte-

(a)

(b)

Fig. 6. GCA in a 63-year-old female. (**a**) CT scan through the mid-thorax shows a dissecting aneurysm of the descending aorta (arrow). This was a late development occurring many months after the acute phase of her disease process. Also note the dilated pulmonary outflow tract (curved arrow). (**b**) On this sagittal reconstruction of the CT images, the dissection flap is identified (arrows) between the two different densities of contrast enhancement. The low density area is the false lumen.

(a) (b)



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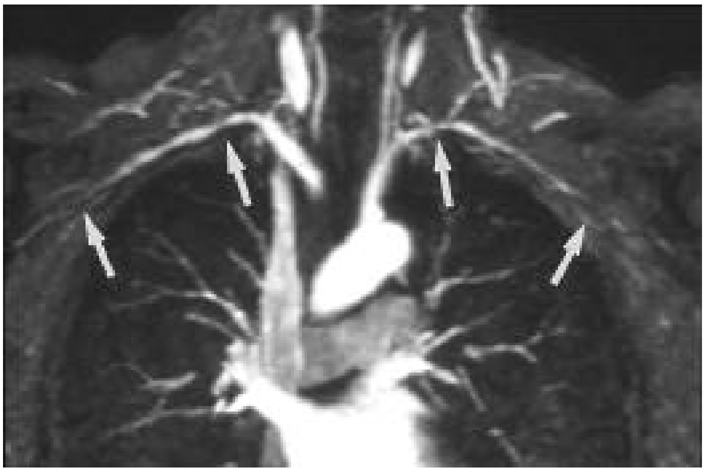

Fig. 7. GCA in a 66-year-old female. (**a**) CT scan through the level of the aortic arch shows slight wall thickening of the left lateral aspect of the aortic arch (curved arrow) and also circumferential wall thickening of the innominate artery (arrow). (**b**) Magnetic resonance image through the same level shows a high signal of intensity of wall thickening along the left lateral aspect of the aorta (arrow). (**c**) Magnetic resonance angiography with gadolinium injection. This frontal projection shows bilateral stenotic disease of the subclavian and axillary arteries (arrows).

Fig. 8. Takayasu's arteritis in a 40-year-old female. Magnetic resonance angiography with gadolinium injection, coronal view. Note on the left side there are three parallel vascular structures (arrows). The artery is obscured by the veins. The left arm was used for the injection of contrast material.

ries that course in the long axis of the imaging are imaged in cross section and can be well seen. However, those that are horizontal to the plane of the section, such as the majority of the subclavian and axillary segments are not adequately identified for diagnostic purposes. To compensate for this shortcoming, multi-dimensional reconstructions can be created to better display anatomic features in other planes of view.

One important capacity of contrast enhanced CT scanning is the ability to detect inflammation in the wall of the artery and of the aorta, which allows the detection of acute disease (Fig. 7). Ultrasound and magnetic resonance imaging can also detect wall thickening, although the specificity of inflammation versus fibrosis may not be possible with ultrasound (5-7).

Ultrasound also has the ability to detect stenoses and to assess velocity changes, which indicate the degree of stenosis. Additionally, wall thickness can be detected (5, 8). In cases of acute inflammation, this is valuable information for the clinician. The limitation of US is with deep tissues and with overlying bone and gas as would occur in the thorax and abdomen. Another major shortcoming of US is in the detection of slight changes in arterial caliber from one segment to another. A more global imaging technique has an advantage here. Also, in detection of femoral artery branch vessel disease, US would not be reliable. For the detection of wall thickness in the scalp arteries, US is excellent.

Magnetic resonance imaging (MRI) offers a broad range of imaging sequences that can be implemented to provide information about the arterial wall, to detect lumen characteristics (either with or without intravenous contrast injection) as well as to indicate velocity changes. Currently, at all sequence choices, the MR images do not have the same high levels of specificity and image resolution as do ultrasound and angiography. But helpful information can be obtained by MR especially in those cases of more advanced disease, although in the thorax when intravenous contrast is injected, the venous route (left or right arm) may obscure the overlying subclavian and axillary artery segments (Fig. 8).

In patients with clinical evidence of upper extremity pulse deficit, presumably the disease is of sufficient magnitude that it would be satisfactorily detected by MRI. However, if early or mild disease involvement is present, then it may not be possible to detect slight caliber alterations that would be easier to appreciate by angiography (9). This would be especially true in some cases of lower extremity disease.

It is important to anticipate that the current status of MR technology undoubtedly will be superseded in the near future. The decision of which imaging modality to use should be based upon the clinical presentation of the patient and upon the level of expertise of the physicians and the imaging equipment available within the medical practice. The goal of utilizing the least invasive procedure is worthwhile in many situations when a thorough medical evaluation has been performed to help direct the correct imaging pathway. Full knowledge of the angiographic findings of extracranial GCA is an important foundation for utilizing the other imaging modalities.

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