Ruolo dell’ecografia nella diagnosi di vasculite

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Treviso
Figure 1. Types of vessels that are defined as large vessels (A), medium vessels (B), and small vessels (C) in the Chapel Hill Consensus Conference nomenclature system. The kidney is used to exemplify medium and small vessels. Large vessels are the aorta and its major branches and the analogous veins. Medium vessels are the main visceral arteries and veins and their initial branches. Small vessels are intraparenchymal arteries, arterioles, capillaries, venules, and veins.
2012 Revised International Chapel Hill Consensus Conference Nomenclature of Vasculitides


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![Diagram of vasculitis classifications]

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Role of ultrasound in the understanding and management of vasculitis

Wolfgang A. Schmidt

Abstract: Vasculitis is characterized by a circumferential vessel-wall thickening (‘halo’), which can be visualized by modern imaging techniques. In particular, the resolution of ultrasound has increased to 0.1 mm. Ultrasound detects abnormalities that are pathognomonic even in arteries with a diameter below 1 mm. It is particularly helpful in the diagnosis of large-vessel vasculitides, such as classic temporal arteritis, large-vessel giant-cell arteritis (GCA), Takayasu arteritis and idiopathic aortitis. Echocardiography is important for determining cardiac involvement in Takayasu arteritis and also for examining the coronary arteries of children with suspected Kawasaki disease, which is a medium-vessel vasculitis. In small vessel vasculitides ultrasound has only a role for determining the distribution or organ involvement.
2012 Revised International Chapel Hill Consensus Conference Nomenclature of Vasculitides

<table>
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<tr>
<th>CHCC2012 name</th>
<th>CHCC2012 definition</th>
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<tbody>
<tr>
<td>Large vessel vasculitis (LVV)</td>
<td>Vasculitis affecting large arteries more often than other vasculitides. Large arteries are the aorta and its major branches. Any size artery may be affected.</td>
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<tr>
<td>Takayasu arteritis (TAK)</td>
<td>Arteritis, often granulomatous, predominantly affecting the aorta and/or its major branches. Onset usually in patients younger than 50 years.</td>
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<tr>
<td>Giant cell arteritis (GCA)</td>
<td>Arteritis, often granulomatous, usually affecting the aorta and/or its major branches, with a predilection for the branches of the carotid and vertebral arteries. Often involves the temporal artery. Onset usually in patients older than 50 years and often associated with polymyalgia rheumatica.</td>
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Contrast-Enhanced Ultrasound of Carotid Artery Wall in Takayasu Disease

P. Giordana,

Figure 1. Mode-B ultrasound: right common carotid artery circumferential wall thickening suggesting inflammatory arteritis.

Figure 2. Contrast-enhanced ultrasound (6 min post-IV at T0: pretreatment scan. GDM, grey scale median.

Figure 3. Contrast-enhanced ultrasound (6 min post-IV at T1: 3 months post-treatment scan. GDM, grey scale median.

Figure 4. Contrast-enhanced ultrasound (6 min post-IV at T2: 6 months post-treatment scan. GDM, grey scale median.

$T_0$

3 m

6 m
Diagnóstico de la arteritis de Takayasu mediante técnicas no invasivas

Figura 4 Paciente joven diagnosticada de arteritis de Takayasu. Ecografía Doppler abdominal. A) Se observa un riñón derecho de pequeño tamaño con marcada atrofia de su parénquima en relación con flujo renal muy escaso por estenosis crónica de la arteria renal. B y C) Fenómeno de aliasing en la salida de tronco celiaco (B) y de la arteria renal izquierda (C) en relación con aumento de velocidad en éstas como signo indirecto de estenosis. D) Curva de flujo de la arteria renal izquierda donde se detectan velocidades muy elevadas (400 cm/s) que indican la presencia de estenosis.

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Role of ultrasound in the understanding and management of vasculitis

Wolfgang A. Schmidt

The sensitivity of temporal artery duplex ultrasound was 87% with regard to clinical diagnosis, and specificity was 96% in one of the meta-analyses [Karassa et al. 2005]. The presence of a bilateral halo seems to increase the specificity [Arda et al. 2010]. With increasing experience and quality of ultrasound equipment more centres achieve reliable results with ultrasound examination and replace temporal artery biopsy in cases with definitive clinical and ultrasound findings [Schirmer et al. 2011; Porta et al. 2012; 2010].

Figure 1. Colour Doppler ultrasound showing longitudinal (a) and transverse (b) views of a normal temporal artery and in acute temporal arteritis (c, d). The arrows indicate the vasculitic wall swelling.
Role of ultrasound in the understanding and management of vasculitis

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Figure 2. A longitudinal ultrasound image of a normal axillary artery (a) and an axillary artery in large-vessel giant-cell arteritis. The arrows indicate the vasculitic wall swelling with a diameter of 1.8 mm.

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<table>
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<tr>
<th>Medium vessel vasculitis (MVV)</th>
<th>Vasculitis predominantly affecting medium arteries defined as the main visceral arteries and their branches. Any size artery may be affected. Inflammatory aneurysms and stenoses are common.</th>
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<tbody>
<tr>
<td>Polyarteritis nodosa (PAN)</td>
<td>Necrotizing arteritis of medium or small arteries without glomerulonephritis or vasculitis in arterioles, capillaries, or venules, and not associated with antineutrophil cytoplasmic antibodies (ANCAs).</td>
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<tr>
<td>Kawasaki disease (KD)</td>
<td>Arteritis associated with the mucocutaneous lymph node syndrome and predominantly affecting medium and small arteries. Coronary arteries are often involved. Aorta and large arteries may be involved. Usually occurs in infants and young children.</td>
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</table>
Polyarteritis nodosa with multiple aneurysms and renal arteriovenous fistula successfully diagnosed by colour Doppler sonography

HongYan Wang

Fig. 1  a In 2004, a 3D CTA showed an aneurysm of the celiac trunk and showed that the right renal artery was normal. b In 2004, ultrasound showed a proximal celiac trunk stenosis and distal celiac trunk aneurysm in which swirling flow was detected.
Polyarteritis nodosa with multiple aneurysms and renal arteriovenous fistula successfully diagnosed by colour Doppler sonography

HongYan Wang

Fig. 2 The sonography and angiogram of the arteriovenous fistula in 2009. a Sonography showed the dilated distal right main renal artery, which had a diameter of 1.3 cm. The proximal right main renal artery was normal, with a diameter of 0.6 cm. b An arterial waveform pattern with a very high peak systolic velocity (399 cm/s) and a very low resistant index (0.3) in the proximal right renal artery.

Polyarteritis nodosa with multiple aneurysms and renal arteriovenous fistula successfully diagnosed by colour Doppler sonography

Hong Yan Wang

2012 Revised International Chapel Hill Consensus Conference Nomenclature of Vasculitides

**Immune Complex Small Vessel Vasculitis**
- Cryoglobulinemic Vasculitis
- IgA Vasculitis (Henoch-Schönlein)
- Hypocomplementemic Urticarial Vasculitis (Anti-C1q Vasculitis)

**Anti-GBM Disease**

**ANCA-Associated Small Vessel Vasculitis**
- Microscopic Polyangiitis
- Granulomatosis with Polyangiitis (Wegener’s)
- Eosinophilic Granulomatosis with Polyangiitis (Churg-Strauss)
In small vessel vasculitis imaging findings are not specific. At US large echogenic kidneys without specific doppler abnormalities are early findings. Increased echogenicity of the kidneys, although non specific, is the most common early sign of glomerulonephritis. In chronic renal failure, the kidneys appear scarred and shrunken.
Intrarenal Arterial Doppler Sonography in Patients with Nonobstructive Renal Disease: Correlation of Resistive Index with Biopsy Findings

Joel F. Platt

Fig. 1.—Renal Doppler sonogram of patient with acute tubular necrosis. Patient had a rapid rise in creatinine level from 1.4 to 3.1 mg/dl in 2 weeks. Standard real-time sonography of kidneys was normal. Doppler signal from intrarenal artery shows reduced end diastolic flow resulting in an elevated resistive index of 0.78. Renal biopsy was consistent with acute tubular necrosis.

Fig. 2.—Renal Doppler sonogram of patient with vasculopathy. Creatinine level became elevated after a cardiac transplant. Standard real-time sonography was normal. Doppler signal obtained from intrarenal artery shows a significant reduction in end diastolic flow compared with peak systolic flow, resulting in an elevated resistive index of 0.83. Biopsy revealed changes consistent with cyclosporine vasculopathy.

Fig. 3.—Renal Doppler sonogram of patient with glomerular disease. Patient had lupus nephritis and renal biopsy results were consistent with an active proliferative-type glomerulonephritis. Doppler signal from intrarenal artery in this patient with pure glomerular disease shows good diastolic flow, resulting in a normal resistive index of 0.55.

AJR 154:1223–1227, June 1990
Renal parenchymal diseases: Is characterization feasible with ultrasound?

Emilio Quaia  
Michele Bertolotto

Fig. 5a–d Polyarteritis nodosa. On US, kidneys reveal a clear increase in renal parenchymal echogenicity, with a poor cortico-medullary differentiation and b some hypoechoic cortical areas (arrow) which can be expression of localized cortical oedema (see text). c Reduced renal parenchymal vascularization is documented on CD, whereas d a clear increase in RI values (0.82) is revealed by duplex Doppler

Fig. 6a–d Wegener granulomatosis. a On US, kidneys reveal a clear increase in renal parenchymal echogenicity with a low cortico-medullary differentiation, whereas on b CD a clear reduction of renal parenchymal perfusion is documented. c, d Duplex Doppler reveals diffusely increased RIs values (0.75)
Tubulointerstitial nephritis and uveitis (TINU syndrome)
Wegener’s granulomatosis
Granulomatosis with polyangiitis (GPA)
A rare presentation of renal Wegener granulomatosis in a child

Riet D’Hauwe

Fig. 1 Renal ultrasound. a A nodular mass (arrows) is seen in the upper pole of the right kidney. It has a heterogeneous hyporeflective center (asterisk), and a more echogeneous thick rim (dashed arrow). b With colour Doppler, the mass (arrows) appears avascular.
Case report

Granulomatous renal pseudotumor in Wegener’s granulomatosis: imaging findings in one case

G. Verswijvel, I. Eeens, T. Maelen, R. Oyen

The patient was treated with cyclophosphamide and corticosteroids. C-ANCA titer and serum creatinine improved and his clinical and subjective condition returned to normal. Follow-up was done sonographically; 6 months after treatment, the lesion disappeared completely and the kidney returned to normal.

Discussion

Wegener’s granulomatosis is a rare clinicopathological entity of unknown etiology. It is characterized by vasculitis, necrotizing granulomatous lesions which are usually located in the upper and lower respiratory tracts, and glomerulonephritis. It occurs most commonly in the
Cas clinique
Présentation inhabituelle d’une granulomatose de Wegener
Unusual presentation of Wegener’s granulomatosis
S. Ketari Jamoussi

ABSTRACT

We report an unusual case of Wegener’s granulomatosis revealed by spleen infarction and complicated by reno-vascular hypertension. A 33-year-old man with a history of spleen infarction and cerebral venous thrombosis was admitted for malignant hypertension, renal failure and nephritic syndrome. On Doppler renal ultrasonography, ostial stenosis of the right renal artery was evidenced. Right kidney was non functional on scintigraphy and the patient underwent a right nephrectomy. Granulomatous vasculitis of the right renal artery was found but anti-neutrophilic cytoplasmic antibodies were absent. The diagnosis of Wegener’s granulomatosis was suspected. The patient was treated with cyclophosphamide, corticosteroids and plasma exchanges, and renal function markedly improved.

L’échographie abdominale montrait un rein droit diminué de taille avec un rein gauche hypertrophié. L’échographie doppler des artères rénales était en faveur d’une thrombose de l’artère rénale droite. La scintigraphie rénale montrait un rein droit non fonctionnel et un rein gauche de perfusion et de captation diminuées. Un diagnostic d’HTA maligne d’origine rénoverveineuse était posé. En raison de l’échec d’un traitement anti-hypertenseur quadruple, la décision d’une néphrectomie droite était posée.

La Revue de médecine interne 31 (2010) e16–e18
Use of Imaging Studies in the Diagnosis of Vasculitis

Wolfgang A. Schmidt.

Figure 1. Imaging studies of a 37-year-old man with newly diagnosed and untreated active Wegener’s granulomatosis with involvement of the carotid bifurcation and the internal carotid artery. A, Angiography of the left carotid artery shows narrowing of the proximal internal carotid artery. Kinking and coiling of the artery occurs further distally. B, The T1-weighted magnetic resonance image of the left carotid bifurcation shows a perivascular infiltrate. The ultrasound image in a longitudinal (C) and transverse view (D) delineates narrowing of the artery, hyperechoic (bright) wall thickening, and a perivascular infiltrate [5].
La Malattia di Goodpasture: l’esperienza di un singolo centro

Mauro Dugo¹, Alessandra Pasi¹, Pier Luigi Gatti¹, Stefania Mastro Simone¹, Licia Laurino², Giuseppina Barbero³,
Fabrizio Farretti⁶, Cesarina Facchini³⁶, Maria Cristina Marasca¹


### Variable vessel vasculitis (VVV)

Vasculitis with no predominant type of vessel involved that can affect vessels of any size (small, medium, and large) and type (arteries, veins, and capillaries).

### Behçet’s disease (BD)

Vasculitis occurring in patients with Behçet’s disease that can affect arteries or veins. Behçet’s disease is characterized by recurrent oral and/or genital aphthous ulcers accompanied by cutaneous, ocular, articular, gastrointestinal, and/or central nervous system inflammatory lesions. Small vessel vasculitis, thromboangiitis, thrombosis, arteritis, and arterial aneurysms may occur.

### Cogan’s syndrome (CS)

Vasculitis occurring in patients with Cogan’s syndrome. Cogan’s syndrome characterized by ocular inflammatory lesions, including interstitial keratitis, uveitis, and episcleritis, and inner ear disease, including sensorineural hearing loss and vestibular dysfunction. Vasculitic manifestations may include arteritis (affecting small, medium, or large arteries), aortitis, aortic aneurysms, and aortic and mitral valvulitis.
Pseudoaneurysm of the Renal Interlobar Artery in Behçet’s Disease

H. OZKURT

Fig. 1. Abdominal ultrasonography showing a right subcapsular renal hematoma.

Fig. 3. Angiography showing the pseudoaneurysm (arrow) arising from the interlobar artery in the upper pole of the right kidney.

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