CONCISE REPORT

Colour duplex sonography of finger arteries in vasculitis and in systemic sclerosis

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Ann Rheum Dis 2006;**65**:265–267. doi: 10.1136/ard.2005.039149

Case reports: Three patients—two with Wegener's granulomatosis and one with an overlap syndrome of rheumatoid vasculitis, systemic lupus erythematosus, and antiphospholipid syndrome—are described. All patients experienced a sudden onset of Raynaud's phenomenon or acrocyanosis when they had a flare of their disease.

Discussion: Ultrasonography (US) showed dark (hypoechoic) arteries without colour signals, resembling the US pattern of embolism. In contrast, US in patients with systemic sclerosis is entirely different, delineating a smaller artery lumen, reduced pulsation, and thickened, slightly hyperechoic artery walls.

Both in Wegener's granulomatosis and in rheumatoid vasculitis, arteritis usually occurs in small vessels, but it has been shown that larger arteries also, like the temporal arteries and even the carotid arteries, may be affected.^{1–3} Ultrasonography (US) is widely available also among rheumatologists. It has been used to diagnose temporal arteritis, large vessel giant cell arteritis, and Takayasu's arteritis.^{4–6} Although modern high resolution US equipment can easily delineate the finger arteries, few data have been published on this subject. US correlates highly with angiography to detect obliterations of finger arteries.⁷

CASE REPORTS

Case 1

A 36 year old white woman presented with a 4 week history of daily intermittent fever up to 39°C, malaise, arthralgia of hands, feet, toes, and fingers, and new Raynaud's phenomenon. Furthermore, she had a 3 year history of intermittent mild arthritis of hands and fingers, rheumatic nodules, pleural effusions, and mild skin vasculitis. Histology of a lesion in the lower leg displayed vasculitis with lymphocytic and granulocytic inflammatory infiltrates and complete vessel destruction. The patient had a 22 year history of smoking and mild hypercholesterolaemia. She had not had an abortion or thrombosis.

Clinical assessment did not show any swollen joints. The patient had slight urticaria at both arms. All fingers had subungueal splinter haemorrhages (fig 1A). A small rheumatic nodule occurred distal to the right olecranon.

Her erythrocyte sedimentation rate (ESR) was 80 mm/1st h and the C reactive protein (CRP) was 8 mg/l (normal <5.1). Moderate anaemia (6.4 mmol/l; normal 7.6–9.5), mild thrombocytosis (473×10^9 platelets/l), moderate leucocytosis (15.4×10^9 platelets/l), and mild hypergammaglobulinaemia with an IgG of 20 g/l were other indicators of inflammation. Anti-cyclic citrullinated peptide (CCP) antibodies and IgM rheumatoid factor were greatly increased (CCP >100 IU/l, normal ≤5; rheumatoid factor 735 kIU, normal <20). The antinuclear antibody titre was 1/2560 (normal <1/80) with fine-speckled staining pattern but without any further

possible differentiation. IgM anticardiolipin antibodies were repeatedly highly positive. Anti-smooth muscle antibodies were slightly positive (1/40). Anti-dsDNA antibodies and complement were normal.

Radiography of the hands and forefeet was normal, and no erosions were detected. Musculoskeletal US showed moderate synovitis of the right wrist and an erosion of the right 5th metatarsophalangeal (MTP) joint. US of the finger arteries with a 14–8 MHz linear probe (Esaote Technos MPX, Esaote SpA, Genoa, Italy) after a water bath of both hands for 5 minutes at 37°C, delineated hypoechoic (dark) finger arteries without colour signals of three digital arteries distal to the metacarpophalangeal (MCP) joints: right first and third ulnar (fig 1B) and left third radial digital arteries. This ultrasound image is entirely different from that of a healthy person (fig 1C) and the finger arteries of a person with systemic sclerosis (fig 1D). The subclavian, axillary, brachial, radial, ulnar, and palmar arteries were normal. This is the typical US finding of acute thrombosis or embolism. Transoesophageal echocardiography did not show valvular vegetations. Chest radiography demonstrated a small pleural effusion. A Schirmer test and glandular US did not show any signs of Sjögren's syndrome. We gave the patient glucocorticoids, cyclophosphamide, and low dose aspirin. The clinical symptoms quickly improved, but the finger artery occlusions remained.

Case 2

A 45 year old white man had a 2 year history of Wegener's granulomatosis. Necrotising glomerulonephritis and severe polyneuropathy were treated with glucocorticoids and cyclophosphamide. Ten months before re-admittance, azathioprine was given instead of cyclophosphamide. After dose reduction from 200 to 100 mg/day a flare occurred with malaise, arthralgia, mild bloody nasal discharge, mild skin vasculitis, and severe acrocyanosis of most fingers (fig 2A). The patient had arterial hypertension and type 2 diabetes mellitus. He was a non-smoker, and his cholesterol was normal.

His ESR was 52 mm/1st h and CRP was 48 mg/l. The patient had slight anaemia (7.6 mmol/l). The cANCA titre was 1/320 (normal <1/20) with positive anti-proteinase 3 antibodies (35.8 mU/l, normal <7). There was no pulmonary, cardiac, or gastrointestinal involvement. Antiphospholipid antibodies were normal.

US of the finger arteries with the same ultrasound equipment after a water bath of both hands for 5 minutes at 37°C showed the same findings as in the first patient in 17/20 finger arteries. The finger arteries were obliterated distal to an area around the PIP joints (figs 2B and C). Only the

Abbreviations: CCP, cyclic citrullinated peptide; CRP, C reactive protein; ESR, erythrocyte sedimentation rate; MCP, metacarpophalangeal; MTP, metatarsophalangeal; PIP, proximal interphalangeal; US, ultrasonography

Figure 1 US in patient No 1. (A, B), in a healthy subject (C), and in a patient with systemic sclerosis (D). (A) Splinter haemorrhages of the fingers of case 1 (↑). (B) Colour duplex US image with longitudinal view of the occluded ulnar digital artery of the third right finger (↓) proximal to the proximal interphalangeal (PIP) joint in case 1. The colour represents minor flow of a collateral vessel. (C) Colour duplex US image of a normal ulnar digital artery of the author's third finger at the PIP joint. Red colour represents blood flow towards the US probe. Blue colour represents blood flow away from the probe. (D) Right radial digital artery at the second PIP joint of a patient with systemic sclerosis. The lumen is narrow (0.2 mm in contrast with 0.8 mm in fig 1C) with occlusion and collateral flow (↑). The artery wall is slightly hyperechoic and thickened.

perfusion of the right third ulnar digital artery and both first radial digital arteries was normal. The subclavian, axillary, brachial, radial, ulnar, and palmar arteries were normal. We gave the patient high dose heparin, iloprost, aspirin, high dose prednisolone, and again, cyclophosphamide. After 6 weeks, most finger arteries were reperfused.

Case 3

A 41 year old white woman had Wegener's granulomatosis. At presentation, she had had bloody nasal discharge for 2 years, inner ear granuloma, weight loss of 13 kg within 6 months, dry cough for 2 months, paraesthesia and acrocyanosis of the fingers for 1 month. The clinical image of the acrocyanosis was similar to that of patient No 2 (fig 2A).

Her ESR was 28 mm/1st h and CRP was 25 mg/l. She had a thrombocytosis of 568×10^9 platelets/l. The cANCA titre was 1/640 with positive anti-proteinase 3 antibodies (52.2 mU/l).

Radiography and computed tomography disclosed pulmonary infiltrates. Computed tomography and magnetic resonance imaging displayed destructive sinusitis. Sinus histology showed granulomatous vasculitis. Furthermore, the patient had severe polyneuropathy. No signs of renal, cardiac, or gastrointestinal disease or any risk factors for cardiovascular disease were seen.

Duplex US of the finger arteries with the same US equipment after a water bath of both hands for 5 minutes at 37°C showed the same findings as in the first two patients in 18/20 finger arteries, except for the left first radial and right fourth ulnar digital arteries (fig 2D). Occlusions occurred slightly more proximal in most arteries than in case 2: between the MCP and PIP joints. Furthermore, all palmar metacarpal arteries, except for the one of the left fourth and fifth fingers, were occluded with collateral flow. The radial and ulnar arteries, including the palmar arch and the proximal arm arteries, were normal.

Treatment was similar to that of patient No 2. Partial reperfusion occurred within several weeks.

DISCUSSION

The Chapel-Hill classification distinguishes between small, medium, and large vessel vasculitides.⁸ Wegener's granulomatosis is a primary small vessel vasculitis. Rheumatoid vasculitis is a secondary vasculitis that also usually affects small vessels. Nevertheless, small vessel vasculitides may sometimes affect larger arteries.

Patient No 1 has an overlap syndrome of systemic lupus erythematosus, antiphospholipid syndrome, and nodular rheumatoid arthritis with arthritis, positive rheumatoid factor and CCP antibodies, and a sonographically detectable erosion. US is more sensitive than conventional radiography for detecting small erosions at MCP and MTP joints.⁹ Furthermore, antinuclear antibodies and antiphospholipid antibodies were positive in this patient as further risk factors for vasculitis and artery obliteration.

Cases 2 and 3 were patients with Wegener's granulomatosis with vasculitis of the finger arteries. To our knowledge, digital artery involvement in Wegener's granulomatosis has as yet only been described in two previous cases.^{10,11} In patients with large vessel vasculitis, US delineates a characteristic hypoechoic wall swelling in temporal arteries,⁴ axillary arteries,⁵ and other arteries like the ulnar arteries.¹² We have never detected a hypoechoic wall swelling of the finger arteries in vasculitis. In patients with finger artery vasculitis, acute occlusion occurs as in embolism. The same US image occurs in temporal arteries of 30% of patients with acute temporal arteritis because of inflammatory artery occlusion.¹³ US cannot describe the cause of the acute occlusion of finger arteries. Finger artery embolism in up to 18 different finger arteries without embolism of any other

Figure 2 Case 2 (A–C) and case 3 (D). (A) Cyanotic fingers of patient No 2 at room temperature. (B) Colour duplex US image with longitudinal view of occluded ulnar digital artery of the second right finger proximal to the PIP joint. The coloured area at the left side delineates the perfused vessel, the dark area at the right side shows the occluded artery (¶). The coloured area in the middle represents collateral flow (¶). (C) Transverse view of the same artery. The dark area delineates the occluded artery. The small blue spot represents collateral flow. (D) Left ulnar digital artery of the fourth finger between the PIP and distal interphalangeal joint of patient No 3 with occlusion (¶) and collateral flow (¶) in a longitudinal view.

arteries together with normal transoesophageal echocardiography is highly improbable.

The US image of finger arteries in systemic sclerosis and other connective tissue diseases with finger artery involvement (for example, CREST syndrome, dermatomyositis) is entirely different from that in vasculitis. The finger arteries are very small with decreased pulsation and frequently with stenoses. Stenoses lead to increased blood flow velocities and turbulent flow. Often finger, palmar, and ulnar arteries are chronically occluded.¹⁴ Thus it is impossible to detect any artery structure in the US image. In addition, vascularity in the nail beds is reduced in patients with secondary Raynaud's phenomenon in connective tissue diseases.¹⁵ Figure 1D shows a radial digital artery at the PIP joint with narrow lumen, occlusion, and collateral flow in a 60 year old white woman with a 5 year history of Raynaud's phenomenon, sclerodactyly, pulmonary fibrosis, and oesophagus involvement without pulmonary hypertension. The changes of the finger arteries remained despite treatment with glucocorticoids, cyclophosphamide, and angiotensin converting enzyme inhibitors.

We conclude that vasculitis with occlusion of the finger arteries may occur in Wegener's granulomatosis and in rheumatoid vasculitis, although the small number of cases in this report does not allow us to provide enough information about the general pattern of vascular involvement in these conditions. The US image displays acute artery occlusion. It is completely different from the appearance of finger arteries in systemic sclerosis.

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Conflict of interest: None.

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Accepted 27 June 2005

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