GIANT CELL ARTERITIS

G.C. Colin, M. Dupont¹

We report the case of a giant cell arteritis (GCA) in an 81-year-old man who presented with headaches and a decreased general condition associated with an important inflammatory syndrome in laboratory analysis responding to corticotherapy. Color-duplex ultrasonography (CDU) features of temporal arteries obtained with a high frequency probe permits us to underline the typical radiological presentation of this disease.

Key-word: Arteritis.

Case report

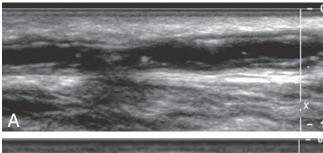
An 81-year-old man was admitted in emergency room for decreased general condition for three weeks with anorexia, loss of weight and insomnia. Blood tests revealed an important inflammatory syndrome with increased C-reactive protein (32 mg/dl). Because the patient was complaining of heavy headaches, a CDU of temporal arteries with a high frequency linear probe (17 MHz) was performed.

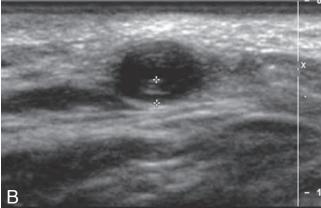
Longitudinal (Fig. 1A) and axial views (Fig. 1B) in B-mode of temporal artery revealed an important hypoechoic circumferential wall thickening (1.2 mm, Fig. 1B). The thickening starts at the origin of the temporal artery in contrast with a spared external carotid artery where thin wall is seen (Fig. 1C).

Color Doppler imaging revealed multifocal areas of aliasing (Fig. 2A) with acceleration in spectral Doppler, the peak systolic velocity was 90 cm/s (Fig. 2B) in contrast with 50 cm/s in the external carotid artery. No artery occlusion was seen but an impaired waveform downstream from these areas with low velocities (32 cm/s), monophasic appearance and diastolic positive component has been observed (Fig. 2C). Combined with the clinical presentation, these US results were suggestive of GCA. The patient received corticotherapy which produced a quick resolution of the inflammatory syndrome and the symptoms.

Discussion

GCA (Horton's disease) is the most common primary systemic vasculitis and typically affects patients older than 50 years (1, 2). GCA is composed by two entities: temporal arteritis and polymyalgia rheumatica. Fever of unknown origin, weight loss and asthenia are often present. Temporal arteritis affects





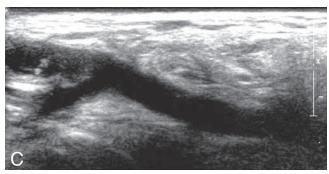
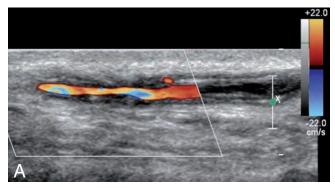
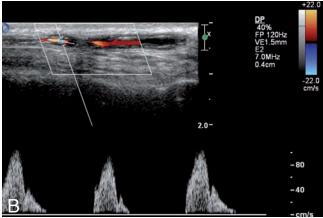


Fig. 1. — Temporal artery in B-mode with a 17 MHz linear probe. Circumferential hypoechoic thickened wall of frontal ramus in longitudinal (A) and axial (B) view. The thickening begins at the origin of temporal artery (white arrowhead) in contrast with the end of external carotid artery (*) where thin wall is seen (C).

the large vessels and particularly the supra-aortic ones. Clinical presentation includes temporal or occipital headaches, jaw claudication and facial pain. The main complication is ophthalmic involvement (anterior ischemic optic neuropathy, central retina artery occlusion) with loss of vision

GCA is often a clinical diagnosis. The American College of Rheumatology (ACR) published in 1990 a five point scoring system for the diagnosis of GCA that appears to be per-





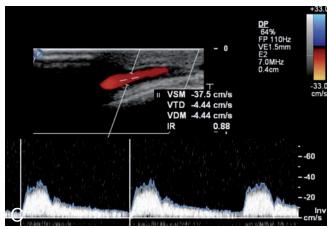


Fig. 2. — Multifocal areas of aliasing in Doppler imaging (A) with acceleration in spectral Doppler (B). These areas represent multiple stenosis. On (C) the post-stenotic arterial waveform in the distal segment of frontal ramus of temporal artery.

forming with high sensitivity and specificity (3). However, temporal artery biopsy is considered as the gold standard to confirm the diagnosis but is invasive. In this context, the role of CDU in GCA diagnosis was first underlined by Schmidt et al. in 1997 (4). The characteristic grayscale US finding of temporal arteritis is a homogenous and diffusely thickened hypoechoic arterial wall. This presentation is called « halo sign » or « macaroni sign ». The halo must be circumferential and must be demonstrated in two planes (4). It is thought

that this halo is a sign of edema in the artery wall, with bad correspondence with histologic analysis where cell infiltration is seen (4). The lumen of the artery is narrowed with areas of stenosis with aliasing in color Doppler mode and higher velocities in spectral mode; occlusion of the artery is also possible (5). The performance of CDU for the diagnosis of temporal arteritis is still under debate and varies in the different studies. Sensitivity of 71% to 81% and specificity of 72 to 99% were reported for the halo sign compared to

temporal biopsy (4, 5, 6, 7, 8, 9). Combined with presence of stenosis or occlusion, sensitivity may be higher (5). The halo sign is not specific of GCA because it was described in a large range of arteritis (polyarteritis nodosa, Henoch-Schonlein, Churg-Strauss, Takayashu disease), but the clinical presentation is often different. The treatment is corticotherapy with quick biologic response (diminishing of CRP) expected. Useful of CDU after corticotherapy is also under debate: some authors reported rapidly decrease of sensitivity of CDU under corticotherapy (9) and other authors reported that halo sign may persist after treatment with excellent clinical and biological response (7).

In conclusion, CDU is a performing and non-invasive first-line examination that permits, with clinical and biological findings, a relatively accurate diagnosis of GCA. Temporal biopsy remains the gold standard and should be reserved for negative scan or with positive scan with clinical discordance.

References

- Becourt-Verlomme C., Barouky R., Alexandre C., Gonthier R., Laurent H., Vital Durand D., Rousset H.: Symptômes inauguraux de la maladie de Horton sur une série de 260 patients. Rev Méd Interne, 2001, 22: 631-637.
- Lockhart M.E., Robbin M.L.: Case 58: giant cell arteritis. Radiology, 2003, 227: 512-515.
- Davies C., Frost B., Eshan O., McLain A.D., Shandall A.: Temporal artery biopsy...Who needs one? Postgrad Med J, 2006, 82: 476-478.
 Schmidt W.A., Kraft H., Vorpahl K., Völker L., Gromnica-Ihle E.: Color du-
- Šchmidt W.A., Kraft H., Vorpahl K., Völker L., Gromnica-Ihle E.: Color duplex ultrasonography in the diagnosis of temporal arteritis. N Engl J Med. 1997, 337: 1336-1342.
- Schmidt W.A.: Doppler ultrasonography in the diagnosis of giant cell arteritis. Clin Exp Rheumatol, 2000, 18: S40-S44.
- Ball E.L., Walsh S.R., Tang T.Y., Gohil R., Clarke J.M.: Role of ultrasonography in the diagnosis of temporal arteritis. *Br J Surg*, 2010, 97: 1765-1771.
 Pérez López J., Solans Laqué R.,
- 7. Perez Lopez J., Solans Laque R., Bosch Gil JA., Molina Cateriano C., Huguet Redecilla P., Vilardell Tarrés M.: Colour-duplex ultrasonography of the temporal and ophthalmic arteries in the diagnosis and follow-up of giant cell arteritis. Clin Exp Rheumatol, 2009, 27: S77-82.
- Habib H.M., Essa A.A., Hassan A.A.: Color duplex ultrasonography of temporal arteries: role in diagnosis and follow-up of suspected cases of temporal arteritis. Clin Rheumatol, 2012, 31: 231-237.
- Hauenstein C., Reinhard M., Geiger J., Markl M., Hetzel A., Treszl A., Vaith P., Bley T.A.: Effects of early corticosteroid treatment on magnetic resonance imaging and ultrasonography findings in giant cell arteritis. Rheumatology, 2012, 51: 1999-2003.