Neurología. 2017;32(2):120—135

SOCIEDAD ESPAÑOLA DE NEUROLOGIA

NEUROLOGÍA

www.elsevier.es/neurologia

LETTERS TO THE EDITOR

Positron emission tomography/computed tomography with 18-fluorodeoxyglucose: A technique for assessing vasculitis of the central nervous system secondary to giant cell arteritis☆,☆☆

Tomografía por emisión de positrones/tomografía computarizada con 18-fluorodeoxiglucosa en la evaluación de vasculitis del sistema nervioso central secundarias a arteritis de células gigantes

Dear Editor:

Giant cell arteritis (GCA) is an immuno-mediated systemic type of vasculitis affecting medium- and large-calibre blood vessels. Its histopathology is characterised by an initial inflammatory process which may lead to vascular occlusion. GCA is the most frequent type of vasculitis in adults. Typical clinical manifestations include headache, jaw claudication, and polymyalgia rheumatica. Stroke is a less frequent form of presentation.

Magnetic resonance angiography (MRA) and arteriography may display non-specific alterations in blood vessels. Biopsy of the temporal artery continues to be the gold standard for diagnosis. According to recent studies, positron emission tomography (PET) may be a useful diagnostic tool for inflammatory diseases such as GCA.[1,2]

We present the cases of 2 patients with symptoms of CNS vasculitis; in these cases, a vascular study with ^{18}F-FDG PET/CT helped diagnose GCA.

Patient 1

Our first patient, a 67-year-old woman, was admitted to the neurology department due to a one-week history of progressive right hemiparesis. She had no history of headache, jaw claudication, fever, or systemic symptoms. Basic laboratory tests, an immunology study, and a CSF test yielded normal results. She displayed an ESR of 43 mm/h. The ECG revealed atrial fibrillation, which had previously been documented. An echo-Doppler study of the supra-aortic trunks (SATs) revealed non-significant stenosis. A brain MRI scan disclosed multiple ischaemic lesions at different stages. The arteriography only revealed non-specific alterations, including stenosis and irregularities in the intracavernous segment of both intracranial carotid arteries, and irregularities in both vertebral arteries. A PET/CT scan displayed increased ^{18}F-FDG uptake along the subclavian and axillary arteries, which suggested GCA (Fig. 1). Biopsy of the temporal artery confirmed this diagnosis. The patient received corticosteroid treatment, achieving progressive improvement of symptoms.

Patient 2

Our second patient was a 63-year-old woman with vascular risk factors and a 2-year history of symptoms of intermittent claudication in the lower limbs. She was admitted to the neurology department due to sudden-onset right brachial monoparesis. A brain MRI scan revealed an ischaemic lesion in the left hemisphere. Six months later, she developed symptoms compatible with right-hemisphere stroke. An additional MRI scan showed acute and subacute ischaemic lesions in both hemispheres. An arteriography of the SATs and intracranial blood vessels revealed distal occlusion of the intracranial portion of the right internal carotid artery, preocclusive stenosis of the left internal carotid artery, and stenosis of the right vertebral and left subclavian arteries. The arteriography of the aorta revealed no abnormalities. A biopsy of the temporal artery showed no vasculitic changes. The ^{18}F-FDG PET/CT study displayed diffuse hypermetabolism in the thoracic aorta, supra-aortic vessels, and abdominal aorta, which was compatible with early-stage GCA (Fig. 2). Symptoms improved with corticosteroid treatment.

The 2 cases presented here illustrate the applicability of a non-invasive technique, in this case ^{18}F-FDG PET/CT, for diagnosing GCA, especially in patients with CNS vasculitis and exclusively neurological symptoms.

☆ Please cite this article as: Mañé Martínez MA, Lopez-Cancio Martínez E, Caresia-Aróztegui AP, Vidaller Palacín A, Martínez-Yélamos S, Escudero D. Tomografía por emisión de positrones/tomografía computarizada con 18-fluorodeoxiglucosa en la evaluación de vasculitis del sistema nervioso central secundarias a arteritis de células gigantes. Neurología. 2017;32:120—122.

☆☆ Partially presented in poster format at the 60th Annual Meeting of the Spanish Society of Neurology. Barcelona, November 2008.

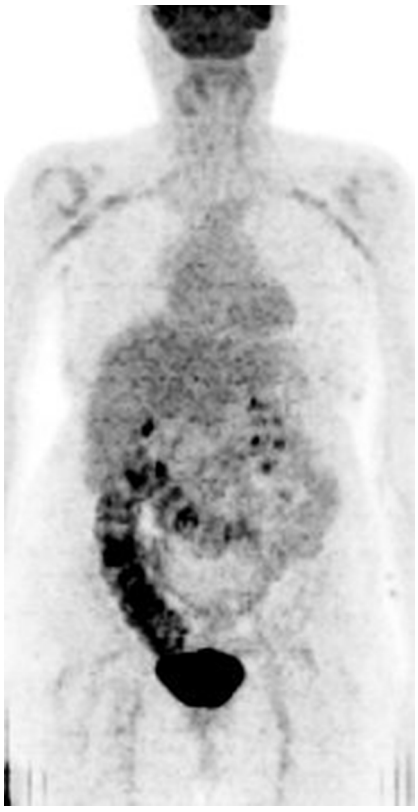

Figure 1 A PET/CT scan displayed increased ^{18}F-FDG uptake along the subclavian and axillary arteries.

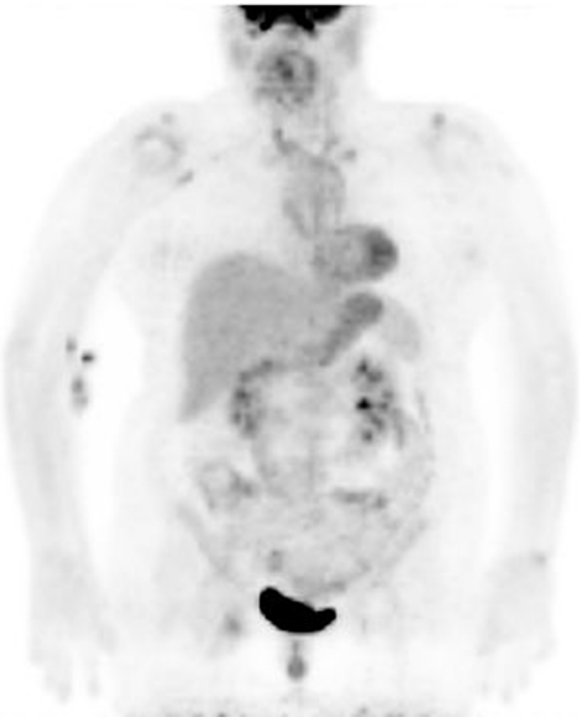

Figure 2 ^{18}F-FDG PET/CT scan showing diffuse hypermetabolism in the thoracic aorta, supra-aortic vessels, and abdominal aorta.

Among non-invasive techniques, echo-Doppler of the SATs may complement diagnosis of large-vessel vasculitis. Unlike ^{18}F-FDG PET/CT, it cannot be used to assess the extent of the disease, but rather serves to examine the temporal and supra-aortic arteries exclusively.[1] In any case, when the disease affects the temporal arteries exclusively, we may find positive results in echo-Doppler of the SAT and false negative results in ^{18}F-FDG PET/CT.[3]

Brain MRI may show vascular alterations when structural lesions are already present; these findings are frequently non-specific. Arteriography of the SATs and intracranial blood vessels, an invasive technique, may confirm intracranial stenosis and display extracranial involvement for patients in advanced stages. However, findings of intracranial stenosis in arteriography studies may be non-specific, since features are similar in primary and secondary vasculitis of the CNA, and even in atherosclerosis.

^{18}F-FDG PET/CT can detect areas of inflammation in the arterial wall of vessels measuring more than 5 mm in diameter in early stages, even before structural changes occur. Furthermore, in the 2 cases presented here, ^{18}F-FDG PET/CT revealed unusual findings, such as diffuse uptake along the arteries typically affected in GCA, which was helpful for differentiating atherosclerosis from arteritis.[4] Likewise, this technique helps determine the spread of the disease in the affected vessels,[5] an important feature for classifying vasculitis, especially when isolated CNS vasculitis is suspected.

In conclusion, ^{18}F-FDG PET/CT has been proved useful in diagnosing large-vessel vasculitis since it enables a more thorough study of the spread and activity of the disease than do ultrasonography, MRI, or arteriography. We therefore suggest using this non-invasive technique in GCA diagnosis and follow-up, especially in initial stages or in cases with atypical manifestations such as CNS vasculitis.

Funding

This study has received no funding.

Conflicts of interest

The authors have no conflicts of interest to declare.

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2173-5808/

Innovative and effective immunosuppressive bitherapy for an unusual paraneoplastic opsoclonus-myoclonus-ataxia syndrome of the adult☆,☆☆

Biterapia inmunosupresora efectiva e innovadora en un síndrome opsoclono-mioclono-ataxia paraneoplásico e inusual del adulto

Dear Editor:

Opsoclonus-myoclonus-ataxia syndrome (OMAS) is a movement disorder whose origin is frequently paraneoplastic or parainfectious. It is characterised by acute or subacute rapid, multi-directional saccades associated with truncal ataxia and diffuse myoclonus; to a lesser extent, dysarthria, appendicular ataxia, and/or deterioration in level of consciousness also appear. In 50% of the cases, neurological symptoms manifest before a tumour is diagnosed and neuroimaging studies display no alterations. Adults with paraneoplastic OMAS show partial or no response to immunomodulatory therapy; in these patients, relapses are frequent and may lead to fatal diffuse encephalopathy. Furthermore, some patients experiencing relapses will need to resume treatment. OMAS is an extremely rare and probably immune-mediated disorder. Lack of clinical trials means that no treatment guidelines are available to date.[1—16] We present a case of recurrent OMAS in an immunocompetent adult whose symptoms resolved with IV methylprednisolone and cyclophosphamide pulse therapy (MCPT).

Our patient was a 62-year-old man with a history of severe use of tobacco (94 pack-years) and alcohol (3-4 glasses of whisky/day) who came to the emergency department due to a 2-week history of ataxic gait with alternating bilateral lateropulsion resulting in falls. Since cerebellar involvement was suspected, he was admitted to the neurology department and underwent a complete analysis and a thorough radiological study. During the hospital stay, our patient's state worsened: he exhibited fluctuations in the level of consciousness, incoherent thinking, visual hallucinations, jargon aphasia, dysarthria, psychomotor agitation, multifocal myoclonus, and binocular nystagmus, which was horizontal-rotatory during the first few days and multidirectional thereafter (Appendix A. Additional material [video available online]). A general analysis ruled out a toxic, metabolic, or infectious aetiology; a brain MRI scan, a whole-body PET/CT study (Fig. 1), and testicular ultrasound revealed no signs of tumours. Our patient tested positive for anti-CV2 antibodies (Table 1) and the EEG displayed generalised grade 2 delta activity. Given the clinical suspicion of OMAS and the lack of response to megadose corticosteroid therapy (1 g/day for 5 days) with 2 subsequent cycles of IV immunoglobulins (0.4 g/day for 5 days), we decided to administer MCPT on a monthly basis (Fig. 2) based on the protocols described in paediatric literature; this treatment has been proven effective and significantly less costly than rituximab. Treatment achieved symptom resolution. Unfortunately, our patient experienced a relapse one year later, after reducing the medication. This time, small cell lung cancer (SCLC) was detected and confirmed by anatomical pathology studies (Fig. 3); extrathoracic metastases were also detected (pleura, liver, adrenal glands). Our patient was diagnosed with definite OMAS and died 7 months later during hospitalisation.

Paraneoplastic neurological syndromes (PNS) are extremely rare, with an incidence rate below 1% in patients with cancer. In adults, the types of cancer most frequently associated with PNS are SCLC (anti-Hu or anti-CV2 antibodies) and breast cancer (anti-Ri antibodies). On the other hand, SCLC represents 20% of all malignant lung tumours. This type of tumour, which originates in Kulchitsky cells, is normally central and frequently associated with mediastinal adenopathies. SCLC is very aggressive and spreads to

☆ Please cite this article as: León Ruiz M, Benito-León J, García-Soldevilla MA, Rubio-Pérez L, Parra Santiago A, Lozano García-Caro LA, et al. Biterapia inmunosupresora efectiva e innovadora en un síndrome opsoclono-mioclono-ataxia paraneoplásico e inusual del adulto. Neurología. 2017;32:122—125.

☆☆ This study was presented in poster format at the 66th Annual Meeting of the Spanish Society of Neurology (Valencia, November 2014).