Long standing polyneuropathy as a form of presentation of primary systemic amyloidosis

Polineuropatía de larga evolución como forma de inicio de amiloidosis sistémica primaria

Dear Editor:

Systemic amyloidosis is a rare disease caused by deposition of a fibrillar protein in immunoglobulin light chains (kappa or lambda) on different tissues. Symptoms depend on the organs affected by the deposits, but may include myocardiopathy, liver disease, kidney failure, lingual hypertrophy, dermatitis, gastro-oesophageal reflux disorder, and polyneuropathy.

A 59-year-old male patient was examined due to sensory deficit of the hands and feet associated with very painful paraesthesia which had been developing gradually over the course of a year. Neurological examination showed cortical functions and cranial nerve pairs to be normal. The motor system revealed preserved tone with slight motor weakness in dorsiflexion of the feet and extension of the fingers and intrinsic hand muscles. All other muscle groups were normal. Deep muscle reflexes were hypotonic, except for the Achilles reflex which was absent. There was neither myotrophy nor fasciculations. Decreased superficial and deep sensitivity in a stocking-and-glove pattern were observed. Autonomic changes in hands and feet were also observed. The coordination was normal. Heel-to-toe gait was somewhat abnormal. The general examination did not reveal relevant changes.

We ran a haemogram and complete biochemical analyses; glucose and antinuclear antibodies, proteinogram, tumour markers, and onconeural antibodies were all within normal ranges. The lumbar puncture revealed no relevant biochemical or cytological changes. Serological tests for Borrelia, Brucella, syphilis, HIV, and neurotropic viruses were negative. The thoracic and abdominal CT showed no alterations. The electromyogram showed decreased amplitude of sensory potentials at the sural and superficial peroneal nerve levels, and to a lesser extent, in median and cubital nerves. Sensory nerve conduction velocities and distal latencies were within normal limits. There was a slight decrease in motor evoked potential amplitudes at the level of both common peroneal nerves, with normal motor potentials at the median and cubital levels. Data were compatible with predominantly sensory axonal polyneuropathy. Biopsies of the sural nerve on 2 different occasions showed no vasculitic changes or amyloid deposits.

The patient was treated with different drug groups such as antiepileptic agents (carbamazepine, topiramate, gabapentin, oxcarbazepine, pregabalin, and clonazepam); antidepressants (amitriptyline, venlafaxine); morphine derivatives, and a cycle of intravenous gamma globulin without symptoms improving.

After 4 years of follow-up, a proteinogram with immunoelectrophoresis showed a monoclonal spike of light lambda chains. The patient refused consent for a bone marrow aspiration. The haematology department diagnosed him with probable monoclonal gammopathy of undetermined significance, and the patient stopped attending haematology check-ups.

The course of the patient’s disease from symptom onset was insidious, but progressive. On the clinical level, sensory polyneuropathy worsened, producing more resistant neuropathic pain, with the autonomic disorder progressing at the same time. This deterioration was detected using several electromyograms performed after symptom onset. Repeated complementary studies did not show any other possible causes of polyneuropathy.

After more than 12 years of follow-up with no extraneurological symptoms, he suffered a major wasting disorder associated with heart failure secondary to atrial fibrillation and hypertrophic myocardopathy, dyspnea, detached right retina, and multiple lacunar infarcts that were probably cardioembolic in origin. He died a few days later of septic shock. Complementary analyses performed during this hospitalisation revealed a monoclonal band (M-spike) in the gamma-globulin region; immunofixation showed lambda light chains. A biopsy of the duodenum and of abdominal cutaneous fat showed eosinophilic material in the vascular walls of the submucous membrane with positive Congo red and thioflavin stains.

Polineuropathy may be caused by a wide variety of processes. In patients with predominantly sensory polyneuropathy, doctors must rule out diabetes mellitus, vitamin B12 deficiency, Sjögren syndrome, HIV, leprosy, paraneoplastic syndrome, and amyloidosis. The prevalence of amyloidosis in sural nerve biopsies is approximately 1% in some series. An amyloid deposit is an amorphous extracellular eosinophilic deposit that shows apple green birefringence after staining with Congo red. Polyneuropathy in primary systemic amyloidosis follows different patterns, but axonal damage predominates over demyelinating damage. It typically presents with an intractable and progressive course; it is painful, affects sensation, and has a significant autonomic component. Diagnosis of primary systemic amyloidosis can often take as long as 2 years from the onset of polyneuropathy; the case we present is extreme. We therefore recommend monitoring over 10 years or more in order to detect amyloidosis if there is a clinical suspicion of that entity. Using abdominal fat aspiration to detect amyloid may be more sensitive than using rectal biopsies, and may therefore be carried out in cases in which the latter procedure gives a negative result. On the other hand, there may be cases of neuropathy in primary systemic amyloidosis in which an increase in immunoglobulins is not detected in serum or in urine. The presence of an autonomic disorder supports a suspected diagnosis of polyneuropathy due to amyloid deposition. Progression of systemic amyloidosis is the cause of death in most of these cases.

References


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Humeral arterial access: An alternative route to the femoral artery in the endovascular treatment of acute stroke

El acceso arterial humeral: una vía alternativa al acceso femoral en el tratamiento endovascular del ictus agudo

Dear Editor:

The femoral artery is the vascular access route most commonly used in cerebral endovascular procedures. Nevertheless, access at this site is impossible in a number of situations, such as atheromatosis of the femoral arteries, elongation of the supra-aortic trunks, and anatomical variations of the aortic arch. When the femoral access cannot be used, endovascular therapy may be delayed or even contraindicated according to some protocols. Here, we describe the case of 2 patients with ischaemic stroke who were able to undergo endovascular treatment by means of a brachial approach only.

Case 1: male patient aged 45 years, diagnosed with basilar artery thrombosis, who arrived at the angiography room 6 h and 20 min after symptom onset. Twenty minutes later, a usable femoral access was obtained. However, even after multiple attempts, the right vertebral artery could not be catheterised due to arterial elongations (the left vertebral artery was hypoplastic). We therefore used a right transbrachial approach and achieved basilar artery recanalisation 33 min after puncture.

Case 2: male patient aged 71 years with a history of peripheral artery disease. He arrived at the angiography room with an ischaemic stroke in the territory of the left sylvian artery due to an M1 proximal occlusion evolving over 4 h and 5 min. Angiography of the aortoiliac axis through the femoral access showed pre-occlusive stenosis of the right iliac artery with occlusion of the left iliac artery. After 4 h and 50 min of onset, we gained access through the right brachial vein and then recanalised the medial cerebral artery in 49 min.

The transfemoral approach (Seldinger technique) is the standard access route for neurovascular surgical procedures, as it allows the use of larger devices and provides better navigation capabilities. The dose of radiation is lower, and the femoral artery has a low thrombotic complication rate. The axillary, humeral and radial puncture sites are all included in the brachial approach. The axillary artery allows use of 8 French introducers. However, the risk of haematoma is higher since haemostatic compression is more difficult at this site. Although the humeral artery allows use of introducers of up to 6 French and offers easy access, the risk of arterial thrombosis is high in prolonged procedures. The radial artery allows use of introducers as large as 6 French, but the risk of arterial thrombosis can be as high as 10%. Over the past few years, we have read published results from isolated series in which transradial or transbrachial approaches were used for diagnostic cerebral arteriography, and for stents in stenosis of the anterior and posterior circulation. Using a radial or brachial access eliminates the risk of retroperitoneal haemorrhage and allows the patient to resume walking early on. This alternative is used when vessel tortuosity makes it difficult or impossible to reach the vertebral artery via the femoral artery. Nevertheless, for purposes of the current endovascular treatment for acute stroke, the smaller diameter of the brachial artery limits the size of the introducer normally used with 7 or 8 French balloon-tipped guide catheters needed for aspiration thrombectomy. It also limits use of devices needing catheters with a larger internal lumen. Other factors, such as lack of experience with this approach and its increased complexity, which may prolong the procedure, also limit the use of brachial access as a first-choice surgical option in acute stroke. Nevertheless, there are no controlled clinical trials comparing the efficacy and safety of the different approaches. The fact that new devices are being designed could change that situation.

Two extensive meta-analyses were recently published which compare transradial and transfemoral access for percutaneous coronary intervention procedures.

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