and 19 years in men and 28, 33, and 34 years in women, respectively.[4]

Clinical suspicion of this disease is important since cardiac symptoms can be treated.[5] A study of 50 patients with hypertrophic myocardiopathy and negative results on a genetic screening for 9 sarcomeric genes revealed 2 cases with Danon disease, accounting for 1% of the 197 patients with a clinical diagnosis of hypertrophic cardiomyopathy.[6]

HyperCKaemia is present with mental disability in dystrophinopathies, especially in Duchenne muscular dystrophy and Danon disease; a slight increase in CK may be observed in congenital myotonic dystrophy. However, other causes must be considered; given the high prevalence of mental disability, there may be a casual association with some other myopathy.

We highlight the importance of studying significant persistent hyperCKaemia, and note that a muscle biopsy should be performed when there is no diagnosis. We also underscore the need for cardiological studies of the different myopathies which may present with myocardiopathy, and follow-up on myopathies in families with myocardiopathies and no established aetiologial diagnosis.

References

1. Danon MJ, Oh SJ, DiMauro S, Manaligod JR, Eastwood A, Naidu S, et al. Lysosomal glycogen storage disease with normal acid maltase. Neurology. 1981;31:51—7.
2. Nishino I, Fu J, Tanji K, Yamada T, Shimojo S, Koori T, et al. Primary LAMP-2 deficiency causes X-linked vacuolar cardiomyopathy and myopathy (Danon disease). Nature. 2000;406:906—10.
3. Sugie K, Noguchi S, Kozuka Y, Arikawa-Hirasawa E, Tanaka M, Yan C, et al. Autophagic vacuoles with sarcolemmal features delineate Danon disease and related myopathies. J Neuropathol Exp Neurol. 2005;64:513—22.
4. Boucek D, Jinkovic J, Taylor M. Natural history of Danon disease. Genet Med. 2011;13:563—8.
5. D'souza RS, Levandowski C, Slavov D, Graw SL, Allen LA, Adler E, et al. Danon disease: clinical features, evaluation, and management. Circ Heart Fail. 2014;7:843—9.
6. Charron P, Villard E, Sebillon P, Laforêt P, Maisonobe T, Duboscq-Bidot L, et al. Danon's disease as a cause of hypertrophic cardiomyopathy: a systematic survey. Heart. 2004;90:842—6.

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Diabetes and motor impairments[☆]

Diabetes y alteraciones motoras

Dear Editor,

We present the case of a 88-year-old Spanish woman with a history of arterial hypertension treated with amiloride/hydrochlorothiazide who arrived at the emergency department with decreased level of consciousness, agitation, and disorientation in time. Her homecare service had detected elevated serum glucose levels (>500 mg/dL); the patient was given regular insulin and transported to the emergency department. In the previous 2 months, she had presented polydipsia and polyuria, and her consumption of solid food had decreased. She was afebrile and had no other symptoms. Physical examination revealed normal vital signs, mild dehydration, and no oedema of the lower limbs. The cardiopulmonary and abdominal exploration showed no anomalies. The neurological examination revealed disorientation in time with preserved orientation in place and person. The emergency department's complementary tests included an electrocardiogram and chest radiography, which yielded normal results; a blood biochemistry study (glucose 661 mg/dL, creatinine 1.88 mg/dL, urea 64 mg/dL); a complete blood count (no leukocytosis or neutrophilia); a urine test (no ketone bodies, high presence of bacteria in urine sediment); and a venous blood gas analysis, which revealed metabolic acidosis (pH 7.27, HCO_3 18.8 mmol/L). Our patient received the first dose of antibiotics (amoxicillin/clavulanic acid) and continuous regular insulin infusion to normalise her glucose levels, which resulted in orientation in time, place, and person, and a normal level of hydration. She was subsequently admitted to the internal medicine department with the following diagnosis: simple hyperglycaemic decompensation in a patient with undiagnosed diabetes mellitus, nonketotic metabolic acidosis, acute renal failure of probable prerenal origin, urinary tract infection, and acute confusional state, which had been resolved. After admission to the internal medicine department, our patient experienced good clinical progress and biochemical normalisation. Basal insulin therapy achieved good glucose control. During hospitalisation, the

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patient began to experience choreic movements on the right side of her body; movements were initially mild but worsened over time. A cranial CT revealed no significant alterations. An MRI scan ordered by the neurology department revealed no lesions to the basal ganglia. Since the hemiballismus and choreic movements affecting the right side of the body were probably due to sustained severe hyperglycaemia, we initiated treatment with haloperidol, which achieved a good clinical response. Haloperidol was discontinued during hospitalisation after choreic movements had resolved due to normalisation of glucose levels. Our patient required no haloperidol after discharge.

Hemiballismus-hemichorea is a movement disorder characterised by continuous, rapid, violent, involuntary movements of the limbs.[1] It is caused by a lesion in the contralateral basal ganglia (including the striatum [caudate nucleus and putamen], globus pallidus, subthalamic nucleus, and substantia nigra), and more specifically, the contralateral striatum. Lesions in the putamen may be due to a number of causes. Although they are most frequently caused by vascular processes (ischaemic stroke in the subthalamic area), they can also result from injury to other areas including the thalamus, striatum, and cortical and subcortical regions.[2] The second most frequent cause is hyperosmolar hyperglycemic nonketotic syndrome (HHNS), which is characterised by hyperintensities in the contralateral putamen in CT images and MRI signal alterations in the contralateral putamen (hyperintensities in T1 and hypointensities in T2), all of which disappear as symptoms resolve.[3] Other metabolic causes include hyperthyroidism, hypoparathyroidism, pregnancy, hyper- or hyponatraemia, hypomagnesaemia, hypocalcaemia, hypoglycaemia, acute hepatocellular degeneration, and nutritional disorders. Hemiballismus-hemichorea may also be of infectious, autoimmune, or neoplastic aetiology.

The pathogenic mechanisms by which hyperglycaemia causes a lesion in the basal ganglia are not clear. It has been hypothesised that it may be caused by a petechial haemorrhage in the area of the lenticulostriate artery that supplies the striatum[4–6]; however, a biopsy of the putamen found no haemosiderin deposits.[7] Another theory suggests hypofunction of the contralateral putamen, with decreased blood flow and metabolic activity.[8,9] Other researchers have proposed a vascular mechanism due to hyperviscosity secondary to hyperglycaemia.

Hemiballismus-hemichorea secondary to HHNS usually affects elderly patients with diabetes and may constitute the initial presentation of diabetes mellitus, as in our case; its mean age at presentation is 72 years,[10] whereas hemiballismus-hemichorea of vascular origin presents at younger ages.[1] Incidence is similar for both sexes. This disorder seems to be more frequent in Asian people.

In addition to aetiological treatment, patients also receive symptomatic treatment with neuroleptics. Tetrabenazine may prevent late-onset dyskinesia.[2] Antiepileptic drugs (valproic acid, gabapentin, topiramate) may also be useful. Surgical treatment of the thalamus[11] or internal globus pallidus[12,13] is an option when symptoms do not resolve. Patients usually respond well to treatment[14]: symptoms tend to resolve after normalisation of glucose levels and radiological lesions in the putamen disappear after 3 to 11 months[15].

Conclusions

Glucose metabolism should be analysed in patients with sudden-onset movement disorders, especially in the elderly, since symptoms may be reversible in some cases and most patients improve with appropriate metabolic control.

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Conflicts of interest

The authors have no conflicts of interest to declare.

References

1. Dewey RB, Jankovic J. Hemibalism-hemichorea: clinical and pharmacologic findings in 21 patients. Arch Neurol. 1989;46:862—7.
2. Shannon KM. Hemiballismus. Curr Treat Options Neurol. 2005;7:203—10.
3. Wang JH, Wu T, Deng BQ, Zhang YW, Zhang P, Wang ZK. Hemichorea-hemiballismus associated with nonketotic hyperglycemia: a possible role of inflammation. J Neurol Sci. 2009;284:198—202.
4. Altafullah I, Pascual-Leonew A, Duvall K, Anderson DC, Taylor S. Putaminal hemorrhage accompanied by hemichorea-hemiballism. Stroke. 1990;21:1093—4.
5. Broderick JP, Hagen T, Hrott T, Tomsick T. Hyperglycemia and hemorrhagic transformation of cerebral infaction. Stroke. 1995;26:484—7.
6. Chang MH, Chiang HT, Lai PH, Sy CG, Le SJ, Lo YU. Putaminal patechial hemorrhage as the cause of chorea: a neuroimagen study. J Neurol Neurosurg Psychiatry. 1997;63:300—3.
7. Nagai C, Kato T, Katogiri T, Sasaki H. Hyperintense putamen on T1-weighted MR images in a case of chorea with hyperglycemia. Am J Neuroradiol. 1994;16:124—6.
8. Chang MH, Li CJ, Lee SR, Men CY. Non-ketotic hyperglycemic chorea: a SPECT study. J Neurol Neurosurg Psychiatry. 1996;60:628—30.
9. Tamawaki T, Isa K, Watanabe Y. A long-term neuroimageng follow-up study in a case of hemichorea-hemiballism with non-ketotic hyperglycemia. Mov Disord. 2000;15 Suppl. 3:251.
10. Lin JJ, Lin GY, Shih C, Shen WC. Presentation of striatal hyperintensity on T1-weighted MRI in patients with hemiballism-hemichorea caused by non-ketotic hyperglycemia. Report of seven new cases and a review of literature. J Neurol. 2001;248:750—5.
11. Tsubokawa T, Katayama Y, Yamamoto T. Control of persistent hemiballismus by chronic thalamic stimulation. Report of two cases. J Neurosurg. 1995;82:501—5.
12. Slavin KV, Baumann TK, Burchiel KJ. Treatment of hemiballismus with stereotactic pallidotomy. Case report and review of the literature. Neurosurg Focus. 2004;17:E7.
13. Hasegawa H, Mundil N, Samuel M, Jarosz J, Ashkan K. The treatment of persistent vascular hemidystonia-hemiballismus with unilateral GPi deep brain stimulation. Mov Disord. 2009;24:1697—8.
14. Lee BC, Hwang SH, Chang GY. Hemiballismus-hemichorea in older diabetic women: a clinical syndrome with MRI correlation. Neurology. 1999;52:646—8.

15. Clark JD, Pahwa R, Koller WC, Morales D. Diabetes mellitus presenting as paroxysmal kinesigenic dystonic choreoathetosis. Mov Disord. 1995;10:353—5.

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Radiological findings of a symptomatic carotid pseudocclusion: ''Guadiana river sign''☆,☆☆

Hallazgos radiológicos de una pseudooclusión carotídea sintomática: signo del Guadiana

Dear Editor,

> ''Your squire Guadiana, lamenting your hard fate, was in like manner metamorphosed into a river that bears his name; yet still so sensible of your disaster, that when he first arose out of the ground, to flow along its surface, and saw the sun in a strange hemisphere, he plunged again into the bowels of the earth; but the natural current forcing a passage up again, he reappears again and again...''[1]
>
> Miguel de Cervantes Saavedra

Pseudocclusion of the internal carotid artery (ICA) is the consequence of a large atheromatous lesion causing high-degree stenosis. This lesion is commonly located in the proximal segment of the ICA or at the level of the carotid bifurcation, giving rise to a filiform flow in the distal portion followed by recovery of its normal lumen.[2]

In 1967, Davies and Sutton were the first to refer to a slowing of the carotid flow in the context of their study of intracranial hypertension.[3] Lippman et al.[4] described an atherosclerotic lesion in the ICA and distinguished it from carotid hypoplasia.

Pseudocclusions may result from carotid dissection, whether traumatic or spontaneous, or else from stenosis secondary to a radiation-induced arteritis, congenital carotid hypoplasia, or a severe diffuse atheromatous lesion extending distally[5] and mimicking a complete occlusion. This erroneous impression frequently arises when the working diagnosis has been determined using a single non-invasive imaging technique which does not enable reliable identification of a critical stenosis or pseudocclusion of the ICA. Combining several non-invasive techniques to study an apparent occlusion increases their diagnostic ability considerably, with no need for such invasive techniques as angiography. However, radiological findings characteristic of a carotid pseudocclusion have classically been defined by angiography, and they are traditionally grouped under the term 'carotid string sign'.

Carotid string sign

The radiological terms 'carotid slim sign' or 'carotid string sign' refer to the angiographic finding of a critical stenosis of approximately 99%, located at the root of the ICA, with preserved patency of the threadlike distal segment.[2]

Many diverse and different findings have been included under the umbrella term 'string sign': from carotid artery disease (mentioned above) to the narrowing, frayed appearance of the terminal ileum in the context of inflammatory bowel disease. In neurology, and especially in the field of stroke, the term 'string sign' is usually associated with a hyperdense artery during ischaemic stroke in the hyperacute stage. Even so, questions remain about the validity of using a catch-all heading under which such different diseases are included.

Guadiana river

The seeming disappearance and distal reappearance of blood flow, which are characteristic of carotid pseudooclusion, are reminiscent of the 'hidden' course of the Guadiana river, the Iberian peninsula's fourth greatest river in terms of length and discharge. It runs east to west from Castille-La Mancha across Andalusia to reach Extremadura, where it finally turns south to mark the Spanish-Portuguese border before flowing into the Atlantic Ocean. The course of the river disappears in the vicinity of the town of Argamasilla de Alba (Castille-La Mancha) to later reappear through natural apertures, such as those located near Villarrubia de los Ojos, gateway to the springs known as the ''Ojos del Guadiana''. The river's underground course is explained by the multiple cracks in the rocks which enable the water to flow through an aquifer system before reaching the surface again.[6]

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