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Spontaneous convexity subarachnoid haemorrhage: Clinical series of 3 patients with associated cerebral amyloid angiopathy☆

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Cheiro-oral symptoms

Abstract
Introduction: Convexity subarachnoid haemorrhage (cSAH) is a rare type of spontaneous, non-traumatic, and nonaneurysmal SAH characterised by blood collections in one or more cortical sulci in the convexity of the brain; the aetiology varies. We report a clinical case series of 3 patients with cSAH associated with probable cerebral amyloid angiopathy (CAA) who presented with focal sensory seizures and responded well to corticosteroid treatment.
Patients: Case 1 was a 67-year-old man reporting right-sided paroxysmal sensory episodes with Jacksonian progression, cheiro-oral symptoms, and motor dysphasia. Case 2 was a 79-year-old man reporting left-sided paroxysmal episodes with cheiro-oral signs and dysarthria. Case 3 was a 71-year-old woman also reporting recurrent left cheiro-oral signs and dysarthria. None of the patients had headache or clinical dementia. Aneurysms were ruled out using MR angiography.
Results: Brain CT scan detected an isolated hyperintensity in a sulcus of the frontal convexity; brain gradient echo T2-weighted MRI sequences showed meningeal haemosiderosis and microbleeds. However, no atrophy was identified in medial temporal lobes including the hippocampal formation. All patients had low levels of beta-amyloid in CSF, low values on the Hulstaert index and high levels of phosphorylated tau protein. Patients were initially treated with prednisone and levetiracetam, but symptoms recurred in 2 patients after prednisone was discontinued.

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Conclusions: We present a series of 3 patients with cSAH associated with CAA, characterised by a stereotypical syndrome responding well to corticoid treatment; there were no cases of headache or clinical dementia.

PALABRAS CLAVE
Hemorragia subaracnoidea;
Hemosiderosis meníngea;
Microsangrados;
Apolipoproteína E;
Péptido beta-amiloide;
Síntomas queiroorales

Hemorragia subaracnoidea espontánea de la convexidad cerebral: una serie clínica de 3 pacientes asociada con angiopatía amiloide cerebral

Resumen
Introducción: La hemorragia subaracnoidea de la convexidad cerebral (HSAc) consiste en la presencia de un sangrado espontáneo, no aneurismático ni traumático, localizado en los surcos de la convexidad cerebral, cuya etiología es muy variada. Presentamos una serie de 3 casos de HSAc con probable angiopatía amiloidea cerebral (AAC) con clínica sensitiva recurrente que respondió al tratamiento con corticoides.
Pacientes: Caso 1: varón de 67 años que presenta episodios paroxísticos sensitivos en el hemicuerpo derecho con progresión jacksoniana, episodios sensitivos queiroorales con disfasia motora. Caso 2: varón de 79 años, con trastorno paroxístico sensitivo-motor queirooral izquierdo y disartria. Caso 3: mujer de 71 años, con trastorno paroxístico sensitivo-motor queirooral izquierdo y disartria. Ningún paciente tuvo cefalea ni deterioro cognitivo. Se descartó la presencia de malformaciones aneurismáticas con una angio-RM cerebral.
Resultados: La tomografía computarizada craneal mostró una hiperdensidad aislada en un surco de la convexidad frontal y la RM encefálica en la secuencia de eco-gradiente mostró depósitos de hemosiderina en dichos surcos y lesiones sugestivas de microsangrados. La RM no mostró atrofia de hipocampos ni temporal medial. En el líquido cefalorraquídeo todos los pacientes tuvieron un descenso del péptido beta-amiloide, valores bajos del índice de Hulstaert y aumento de la proteína tau fosforilada. Todos los pacientes se trataron inicialmente con prednisona y levetiracetam pero los síntomas recurrieron en dos pacientes tras la suspensión de la prednisona.
Conclusiones: Presentamos a 3 pacientes con HSAc asociada a AAC, caracterizados por una clínica estereotipada, con ausencia de cefalea y de demencia clínica, con buena respuesta al tratamiento corticoideo.

Introduction

Cerebral subarachnoid haemorrhage (cSAH) refers to subarachnoid bleeding in a sulcus (or several adjacent sulci) of the convexity of the brain that does not extend to the interhemispheric fissures, basal cisterns, or ventricles.[1–5] Different authors have stated that age at presentation of haemorrhage is related to the multiple aetiologies of cSAH: cerebral vasoconstriction syndromes are more frequent among patients younger than 60 whereas cerebral amyloid angiopathy (CAA) is the main cause of bleeding in patients aged 60 and older.[1–5] The clinical presentation of cSAH is also linked to aetiology: patients with cerebral vasospasm usually experience thunderclap headache whereas those with CAA more frequently experience transient episodes of TIA-like or aura-like focal neurological dysfunction.[1–6]

We present a series of 3 patients with cSAH associated with probable CAA and assess the potential role of the amyloid-beta and tau protein pathways in the pathogenesis of the disease using CSF tests. Our patients displayed typical neurological symptoms, which initially responded very well to corticosteroids. However, final outcomes were poor due to their episodes of recurrent intracranial bleeding.

Patients

Case 1: 67-year-old man with a history of hypercholesterolaemia, diagnosed with TIA in 2005 and treated with an antiplatelet drug (clopidogrel). He was seen by the neurology department due to multiple episodes of neurological dysfunction: a tingling sensation in the mouth area affecting the right side of the tongue, as well as in the fingertips of the right hand and spreading towards the forearm or shoulder. Tingling was accompanied by episodes of speech impairment lasting 10 to 15 minutes and presenting up to 8 times per day. On some occasions, sensory symptoms extended over the right side of his ribcage and down to his right