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Research Article

A Diagnostic Challenge in The Emergency Department: Dysarthria-Clunky Hand Syndrome.

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Abstract:

Background: Small vessel vascular disease can present as different entities including lacunar infarctions. Dysarthria-clumsy hand syndrome is the rarest classic lacunar syndrome, this syndrome is described as the combination of dysarthria with clumsiness of the hand, manifested as slowing in fine manipulation, difficulty in writing, along with hesitant ataxia in the finger-nose test that differed from the cerebellar type.

Clinical case: A 59-year-old female attended the continuous admission service due to acute neurological deterioration characterized by dysarthria and right hemiparesis, performing a neurological physical examination where moderate dysarthria was evident, slight deviation of the left labial commissure, decreased strength in the right upper extremity, latero drive of the march to the right, in angiotomographic study it was reported absence of flow in left perforating artery, Diagnosis of lacunar ischemic vascular event, dysarthria-clumsy hand syndrome.

Conclusions: Dysarthria-clumsy hand syndrome is a rare syndrome with a good prognosis, however, as first contact physicians we must be prepared for the different variants of cerebrovascular disease that may present to the emergency area, never delaying the diagnosis with a view to early treatment to limit the damage.

Keywords: Cerebral vascular event, ataxia, dysarthria, clumsy hand syndrome, lacunar infarction.

Background:

Small vessel vascular disease can present as different entities, including lacunar infarcts, with high blood pressure being one of the main associated risk factors, although atherothrombotic lesions may be present. (1)

Dysarthria-clunky hand syndrome was described by the Canadian neurologist Charles Miller Fisher in 1964 and is the rarest classic lacunar syndrome. This syndrome is described as the combination of dysarthria with clumsiness of the hand, which manifests as slowing down in manipulation. fine, difficulty in writing, together with wavering ataxia in the finger-nose test that differed from the cerebellar type. The most frequent anatomical locations associated with this syndrome are the internal capsule (40%), pons (17%), and corona radiata (8.6%). (2)

Clinical case:

A 59-year-old female presented to the continuous admission service due to acute neurological deterioration characterized by dysarthria and hemiparesis, with the following significant history: no diabetes mellitus or arterial hypertension, 2 surgical cesarean sections without complications, allergies denied, drug addiction denied. Prophylaxis against SARS COV 2, 3 doses (2 Astra Zeneca). Her current condition began suddenly at 1:30 a.m. when she got up to go to the bathroom. She presented incoordination when walking with lateropulsion to the right, dysarthria with altered prosody, and right hemiparesis. Upon her arrival at the emergency department, a neurological examination was performed: Glasgow Coma Scale 15, mental functions oriented in the 3 spheres, speech with moderate dysarthria, praxia with weakness, the rest without alterations. Cranial nerves: NC l: not assessed. NC ll: visual acuity 20/20. CN III-IV-VI: Conjugate primary gaze, normal eyelid opening, 4mm symmetrical pupils, eye movements: without alterations, bilateral photomotor and consensual reflex without alterations, cephalic oculi without alterations. NC V: bilateral corneal and

nasociliary reflex sensitivity present, masseter-temporal muscle strength and tone preserved. NC VII: facial symmetry at rest, however, with gesticulation, slight effacement of the right nasal groove and deviation of the labial commissure to the left, preserving expression in the upper 1/3. NC VIII, IX, X XI and XII: without alterations. Strength: decreased tone in the right thoracic limb with preserved trophism in all 4 extremities. Strength in different upper muscle groups: right 4/5, left 5/5. Pelvic limbs without alterations 5/5. Muscle stretch reflexes: without alterations. Pathological reflexes: bilateral negative, Sensitivity: without alterations. Cerebellum: Romberg does not tolerate decreasing the support base. without presence of nystagmus, dysarthria, fingertip maneuver shows discrete right hypometry, although dysdiadochokinesia is not observed. Gait: When sitting with truncal ataxia, requiring right support, he walks with a wide base of support, decreased arm movements, short and uncertain steps with lateral pulsion to the right, he does not perform heel-toe or tandem. Abnormal movements: Not evident, Atavistic signs: absent, Meningeal signs: negative. Autonomous: Without alterations.

The initial diagnostic suspicion was established as an ischemic cerebral vascular event dependent on the left middle cerebral artery.

To complement the diagnosis, a simple skull tomography was performed, which showed: Hypodensity in the left basal ganglia, possible non-recent lacunar infarction. Diffuse atheromatous disease. Generalized cortico-subcortical atrophy. The rest of the structures without alterations. ASPECT 9 (left lenticular nucleus), see image 1 and 2. Due to a time of evolution greater than 8 hours from the onset of symptoms and tomography with scarce signs of ischemia, subtracting only 1 point on the ASPECT scale, cerebral angiotomography is requested to assess the site of occlusion. vascular and determine if the patient is a candidate for invasive treatment for thrombectomy, reporting in an angiotomography study: arterial routes of the neck are shown to be preserved. The arterial routes of the brain such as internal carotid arteries, middle cerebral arteries, anterior communicating arteries, anterior cerebral arteries, posterior communicating arteries, basilar arteries, posterior and superior anterior cerebellar arteries and both vertebral arteries are shown to be preserved, with adequate flow inside. The vascular reconstruction shows an absence of flow in the left perforating artery, see images 3 and 4.

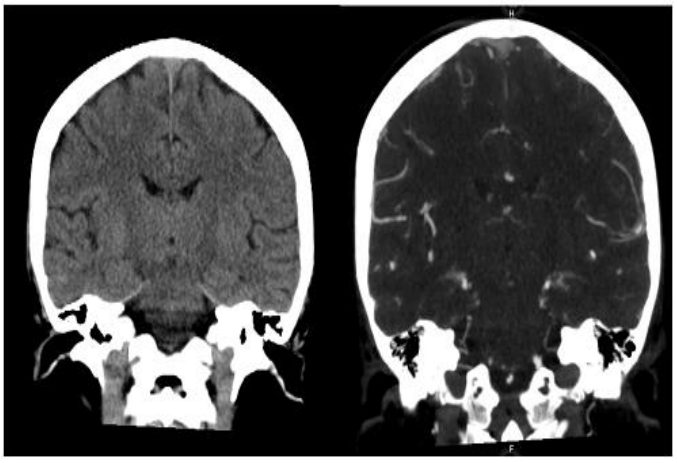

Image-1 and 2 Simple axial section tomography

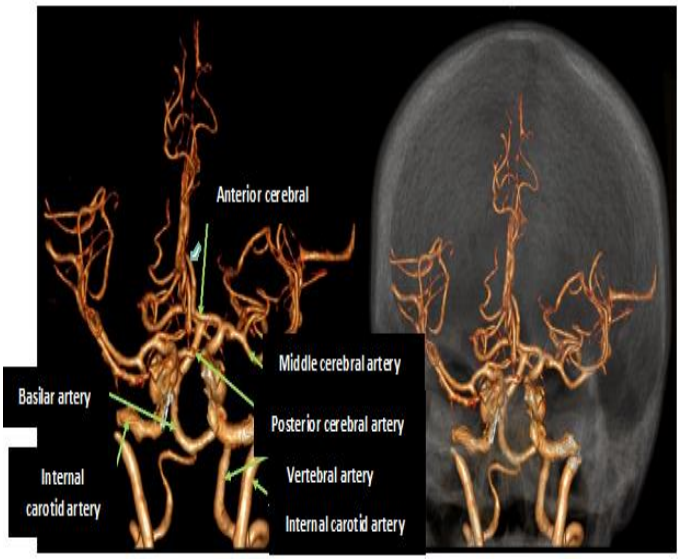

Image-2 Angiotomography with vascular reconstruction showing the absence of flow in the left perforating artery

It presents with "wake-up stroke", attending medical evaluation with more than 8 hours of onset of symptoms, persisting with motor symptoms, so a neurology evaluation was requested due to the suspicion of lacunar infarction syndrome, corroborating the suspicion, establishing the diagnosis of ischemic cerebral vascular disease TOAST 2 lacunar infarction dysarthria-hand (left lenticular artery) NIHSS 6 e-NIHSS 7 (6+1, P-NIHSS 9 (6+3).

Due to the time of evolution, she was not a candidate for the use of thrombolytics, nor was she a candidate for thrombectomy as it was a small vessel disease. Management is given with antiplatelet therapy (acetylsalicylic acid 150 mg PO every 24 hours) and statin (Atorvastatin 80 mg PO every 24 hours) and an MRI is requested.

Discussion:

Infarctions and lacunar syndromes (small vessel disease)

Lacunar infarction is a type of ischemic stroke characterized by a small vascular lesion (<15 mm) and represents up to 25% of ischemic stroke. Lacunar infarcts, together with leukoaraiosis, dilation of perivascular spaces, cerebral microbleeds and cerebral atrophy, are the usual anatomical manifestations of cerebral small vessel disease. (11) It is attributed to an occlusion of one of the perforating arteries of a large cerebral artery. (3) The perforator territory of the middle cerebral artery, lenticulostriate branches, is usually the most affected, characterized by the absence of collaterality. The most frequent locations of lacunar infarcts in order are the nuclei: putamen, globus pallidus, pons, thalamus, caudate, internal capsule and corona radiata. (2) See image 5.

Lacunar stroke shares risk factors with other stroke subtypes, such as age, gender, smoking, hypertension, and diabetes. Among these risk factors, hypertension is the most common in patients with lacunar stroke (68%), followed by diabetes (30%).

Pathophysiology

Lipohyalinosis is defined as concentric hyaline thickening of small cerebral vessels leading to occlusion of small penetrating arteries and is one of the first and most common described and

pathologically proven mechanisms of lacunar strokes. Lipohyalinosis is thought to arise from hypertension-related hypertrophy and fibrinoid degeneration of vessel walls, as well as subintimal foam cells obliterating the lumen of small penetrating arteries, leading to small subcortical infarcts. (3) See image below.

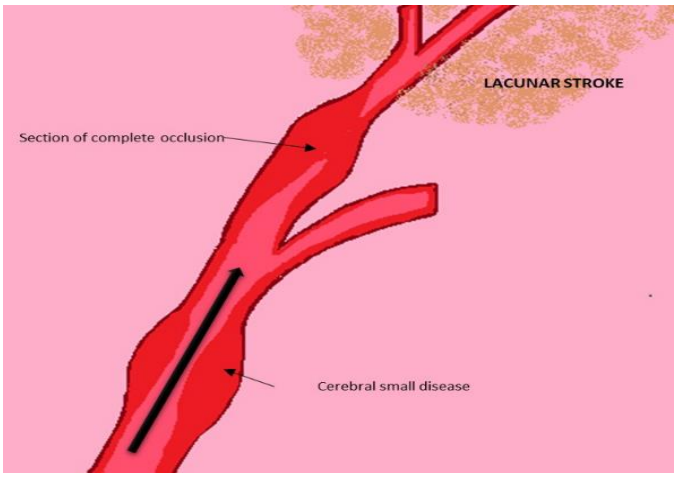

Image-6. From a perforating artery with small vessel disease secondary to atheromatous disease, although some lacunar infarcts may be secondary to thromboembolism. Small vessel disease causes endothelial dysfunction, failure of self-regulation, thickening of the vessel with a decrease in lumen lumen without reaching complete occlusion. (4) Illustration author: Orejel Feria Tabatta

Endothelial dysfunction reflects a shift toward vasoconstriction, a procoagulant, proinflammatory, and proliferative state. (5) This leads to failure in autoregulation, causing the vessel to be unable to maintain perfusion. (4)

Lacunar infarction is the mechanism responsible in 94.4% of cases of dysarthria-clumsy hand.

Clinical manifestations:

The classic clinical expression associated with this lacunar cerebral infarction are the 5 syndromes described as pure motor hemiparesis, pure hemisensory syndrome, sensory-motor syndrome, dysarthria-clumsy hand syndrome and hemiparesis-hemiataxia. (6)

Dysarthria-clumsy hand syndrome affects the anterior limb of the internal capsule in 40% of cases, but can affect the pons. It has been reported that up to 94.3% of patients show weakness in the contralateral limbs and 17.1% ataxia. cerebellar type, and it is not uncommon to find associated facial asymmetry or dysphagia. (2). Most cases resolve spontaneously within ten days to a month. (12).

Pure motor involvement is the most common and usually affects the posterior arm of the internal capsule, centrum semiovale or pons. Sensory involvement in the form of complete hemihypoesthesia or involvement of the mouth, hand and foot (orocheiropodal), is usually the most common, including the thalamus or brain stem.

The proportionality of the degree of involvement of the paresis or similar sensory alteration in the upper and lower extremities suggests a deep or lacunar infarction compared to the disproportionate involvement in the upper extremities in cases of non-lacunar or cortical infarcts.

Hemiparesis-hemiataxia associates ataxia disproportionate to the side of the paresis and is related to infarcts in the pons or cerebellar peduncles or posterior arm of the internal capsule.

Non-focal symptoms such as mood alterations or extrapyramidal disorders such as akathisia, dystonia, ballismus, may occur in some lacunar infarcts located in the basal ganglia or portion of the limbic system.

Diagnosis:

Skull tomography is the simplest test for diagnosis, but less sensitive because in the acute phase it is not always visible given the small size of the lesions. Small oval, elongated or irregular contralateral hypodense areas can be observed and usually occur in the internal capsule or pons.

The vascular study of large vessels with ultrasound of the supra-aortic trunks or angio/MRI or angio-CT is essential in lacunar infarction to demonstrate the absence of ipsilateral disease and to make a diagnosis of exclusion in the event that we do not have cranial magnetic resonance imaging with sequence diffusion.

Cranial magnetic resonance imaging is highly sensitive for demonstrating both acute lacunar infarcts and those in the subacute or chronic stage. Its ability to discern small lacunar infarcts surpasses skull tomography as a diagnostic test for etiological study and semiological correlation.

The MRI shows hyperintensity in the diffusion map (DWI), FLAIR and T2, while the T1 sequence shows hypointense.

Presence of lesions < 3 mm, with a thin hyperintense halo on T2 or FLAIR, located mainly in the putamen, usually indicates small dilated vascular spaces or Virchow-Robin spaces; On the contrary, lesions with a thick and irregular halo (gliosis) in the thalamus or internal capsule usually indicate lacunar strokes. However, the diffusion sequence (DWI) is the most specific and sensitive for the acute diagnosis of lacunar infarction, along with the apparent attenuation coefficient sequence that defines the ischemic nature of the lesion found in the DWI, see image-7.

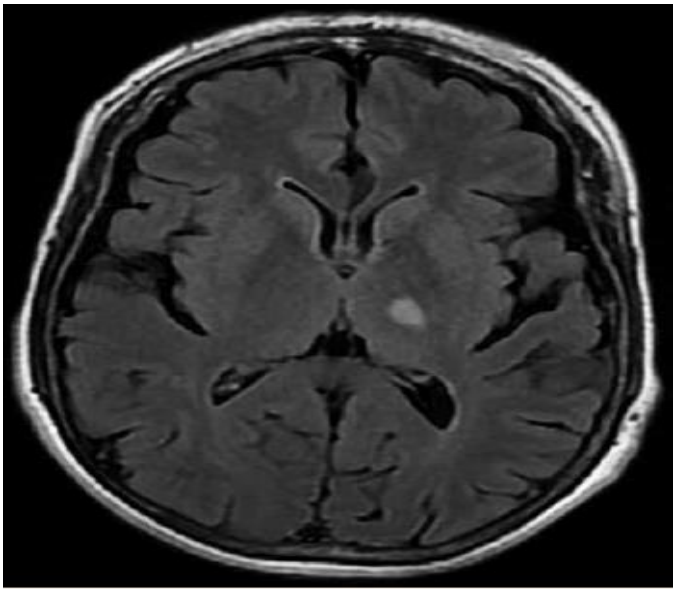

Image-7 Magnetic resonance imaging, DWI/FLAIR sequence, axial section, confirming lacunar infarction that encompasses the genu of the internal capsule and thalamus on the left side. (2)

The TOAST classification is based on the pathophysiology of vessel wall degeneration in lacunar infarction, ignoring the possibility of other mechanisms such as embolic and atherothrombotic, although they are much less frequent. The term TOAST brings together other lesions visible in imaging tests such as cranial MRI: dilated perivascular spaces, microbleeds, brain atrophy disproportionate to age, white matter hyperintensity, small subcortical and lacunar infarcts.

TOAST classification for etiology of acute ischemic cerebral vascular event

Classification	TOAST	CCS
1	Atherosclerosis of large vessels	Cardio aortic
2	Small vessel occlusion	Atherosclerosis of the great arteries
3	Cardioembolic	Small vessel occlusion
4	Other etiology	Other etiology
5	Indeterminate	Not determined

TOAST: Trial of Org 10172 in Acute Stroke Registry
CCS: Causative Classification of Stroke System, year 2007

Table-1: Differential diagnosis of lacunar infarct syndromes.

Syndrome	Clinical manifestations	Location of the lesion	Neuroanatomical condition
Pure motor stroke/hemiparesis	Unilateral motor deficit from face, arm, and leg	Internal capsule rear arm	Corticospinal tract
Pure sensory stroke	Unilateral paresthesias and/or hyperesthesia	Thalamus and corpus callosum	Anterolateral corticospinal tract
Ataxic hemiparesis	More severe hemiparesis in the lower extremity and ipsilateral ataxia	Internal capsule and pons	Corticospinal tract and Corticopontocerebellar Pathway
Dysarthria-clumsy hand syndrome	Dysarthria with facial weakness, discrete dysphagia, and clumsiness of the hand	Internal capsule and pons	Corticospinal tract
Mixed sensorimotor stroke	Ipsilateral motor and sensory deficit	Thalamic capsule	Anterolateral corticospinal tract and Corticospinal tract

Author: Eric Miguel Pozos Gutiérrez

Treatment:

Treatment can be divided into two stages, treatment of the acute vascular event and treatment to prevent recurrence of re-infarction. In the acute phase of stroke, the use of thrombolysis should be considered within 4.5 hours of the onset of symptoms; its benefit in small vessel symptoms does not differ from cases of large vessel occlusion as reported in different studies. without a significant increase in bleeding risk.

Treatment with an antiplatelet agent offers a 30% reduction in the risk of recurrence, which is why a global approach to vascular risk factors is important.

The addition of aspirin to clopidogrel in the SPS3 study on prevention of recurrence of lacunar infarctions did not demonstrate efficacy and did show an increase in bleeding, so this indefinite association would not be indicated in the case of small vessel infarctions. In the case of isolated lacunar infarcts, control with statins seems effective with a certain independence of cholesterol levels due to pleiotropic effects.

Cilastozole is a phosphodiesterase III inhibitor that induces mild antiplatelet effects, BP lowering, and triglyceride lowering for the prevention of lacunar stroke. (3)

Since hypertension is the main risk factor associated with small vessel disease, it must be carefully and strictly analyzed after the acute phase of the stroke, both daytime and nighttime, and if necessary through outpatient monitoring. Blood pressure limits of 130/80 mmHg seem optimal, as can be seen from the SPS3 study, in the prevention of new symptomatic or silent lacunar infarctions. Reducing blood pressure will reduce stroke recurrence by almost 30%.

Table-2: Reinfarction prevention strategies

Treatment	Study reviewed	Considerations	Recommendations
Hypertension control	The secondary prevention of small subcortical strokes (SPS3)	Systolic BP numbers less than 130 mmHg vs 130-149 mmHg	Maintain systolic blood pressure numbers below 130 mmHg

Antiplaquetary treatment	The secondary prevention of small subcortical strokes (SPS3) y estudio CSPS.	Aspirin monotherapy had a bleeding greater than 1.1% per year vs. dual therapy with a 2.1% risk in case of aspirin + clopidogrel. Cilostazol use has a relative risk of 0.41 (95% CI 0.21 to 0.81) for recurrence of a new event.	Based on the guidelines for the prevention of cerebral vascular events, antiplatelet monotherapy is recommended.
Statins use	The Stroke prevention by Aggresive Reduction of Cholesterol Levels stroke (SPARCL)	Atorvastatin 80 mg dose in patients with recent TIA with LDL levels greater than 100 mg/dL.	Atorvastatin 20 mg orally every 24 hours (low doses vs high doses at risk benefit of intracerebral bleeding).
Glycemic control	Post hoc de The secondary prevention of small subcortical strokes (SPS3)	Diabetes mellitus leads to an increased risk of recurrence, as well as higher mortality.	Adequate glycemic control.

Rehabilitation should be considered in case of neurological sequelae, and should include language or cognitive therapy, occupational and physical therapy. The purpose of rehabilitation is to allow the functional independence of people who have suffered a cerebral vascular event. (7)

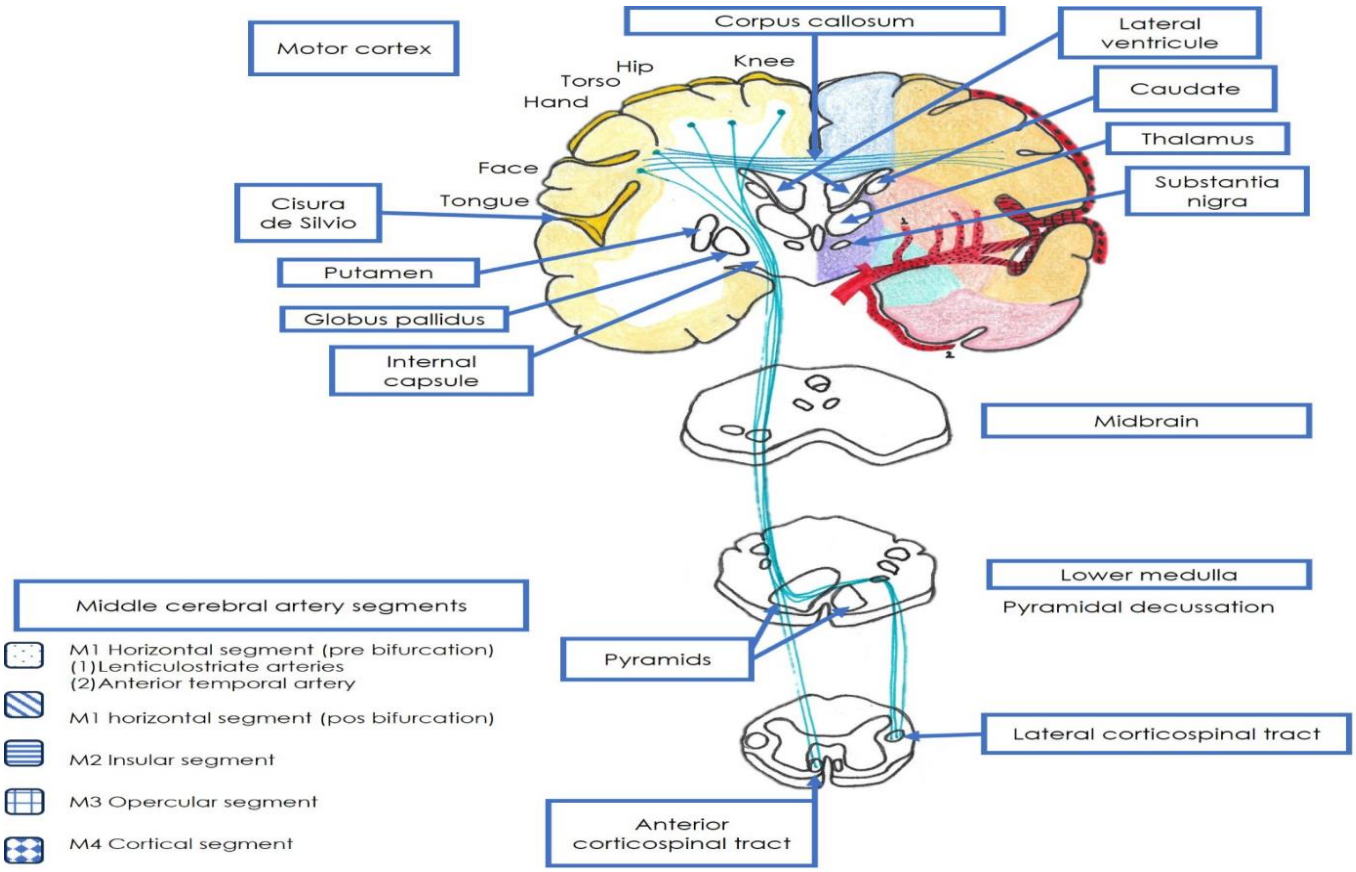

Image-5: In the left half of the image you can see the corticospinal or pyramidal tract, this controls the voluntary movement of skeletal muscles. About a third of the nerve fibers in this tract originate in the primary motor cortex (area 4), another third in the secondary motor cortex (area 6), and the other third in the parietal lobe (areas 3, 1, and 2). , in this diagram we can see how the descending nerve fibers pass through the posterior arm of the internal capsule, this is relevant since the nerve fibers closest to the knee innervate the cervical portions of the body and the more posterior ones the lower limbs. The tract continues through the cerebral peduncles of the midbrain, enters the pons and is divided into bundles by the pontocerebellar transverse fibers. In the medulla oblongata the bundles are grouped to form pyramids. When the tract reaches the junction of the medulla oblongata with the spinal cord, the fibers decussate and enter the lateral cord of the spinal cord to form the lateral corticospinal tract, the fibers that do not decussate are grouped as the anterior corticospinal tract. (8,9) In the right half, the irrigation territories of the arteries are marked with different colors: in purple the territory of the anterior cerebral artery, in green the territory of the anterior choroidal artery, in pink the territory of the posterior cerebral artery, the segments of the middle cerebral artery are also observed, in the case of dysarthria-clumsy hand syndrome it is important to locate the lenticulostriate arteries (marked with the number 1). Illustration author: Orejel Feria Tabatta.

Forecast:

The control of vascular risk factors, especially hypertension, but also diabetes mellitus or cessation of tobacco consumption, are key in the prevention of small vessel vascular disease.
Mortality and complications are low and are usually related to other underlying diseases. More than 90% of patients will present functional independence in the first 6 months.
The recurrence of lacunar infarcts ranges between 10% in the first year and 25% in the following 10 years, where the only factor associated with its poor prognosis is poor control of hypertension.
Dysarthria-clumsy hand syndrome is a lacunar syndrome with an excellent prognosis, characterized by moderate or severe dysarthria and motor clumsiness of the hand in the execution of tasks such as writing, without motor deficit, with a mortality of 0% with 47.5% of patients without neurological sequelae at the time of discharge, with a hospital stay of 11.7 days. (2.10)

Conclusions:

Dysarthria-clumsy hand syndrome is a rare syndrome, with a good prognosis, however, as first contact doctors we must be prepared for the different variants of cerebrovascular disease that can present to the emergency room, never delaying the diagnosis. with a view to early treatment to limit the damage.
Clumsy hand dysarthria is the rarest of all lacunar syndromes. Affecting between 2 and 6% of lacunar strokes. (12)
Prophylactic treatment based on antiplatelet aggregation and high molecular value statins as well as changes in lifestyle are essential to modify risk factors and follow-up in patients who received thrombolysis or who are only in rehabilitation therapy.

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