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A world in the head: Brain abscess caused by Paradoxical embolism due to pulmonary arteriovenous fistula as a debut of hereditary hemorrhagic telangiectasia

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ABSTRACT

Learning points:

-Learning of risk factors for the acquisition of cranial abscess and its etiological study.

-Emphasize the paradoxical cause of systemic embolism.

-Study, screening and Treatment on AVM in hereditary hemorrhagic telangiectasia.

Keywords: Brain abscess; Paradoxical embolism; Pulmonary arteriovenous fistula; Rendu-osler-weber syndrome

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CASE DESCRIPTIPTION

66-year-old patient with a history of alcoholism and smoking, chronic iron deficiency anemia treated with oral iron who was admitted for streptococcal brain abscess, during the etiological study we found arteriovenous shunt on echocardiography and pulmonary arteriovenous fistula, undergoing endovascular closure of the same during stay. In the physical examination, perioral teleangiectasis was observed, when meeting the Curaçao criteria, a genetic study of hereditary hemorrhagic teleangiectasia was requested, which was positive.

DISCUSSION

We present a 66-year-old man, with a personal history of emphysematous chronic obstructive pulmonary disease (COPD), a 100-pack-year heavy smoker, and an alcohol drinker of 2-3 standard drink units per day. He came to the emergency department due to an episode of generalized tonic-clonic seizure with a subsequent post-critical period, in a cranial computed tomography (CT) scan with intravenous contrast that shows the presence of a space-occupying lesion of about 17x15mm left occipital, with peripheral contrast uptake and moderate perilesional vasogenic edema (PICTURE 1).

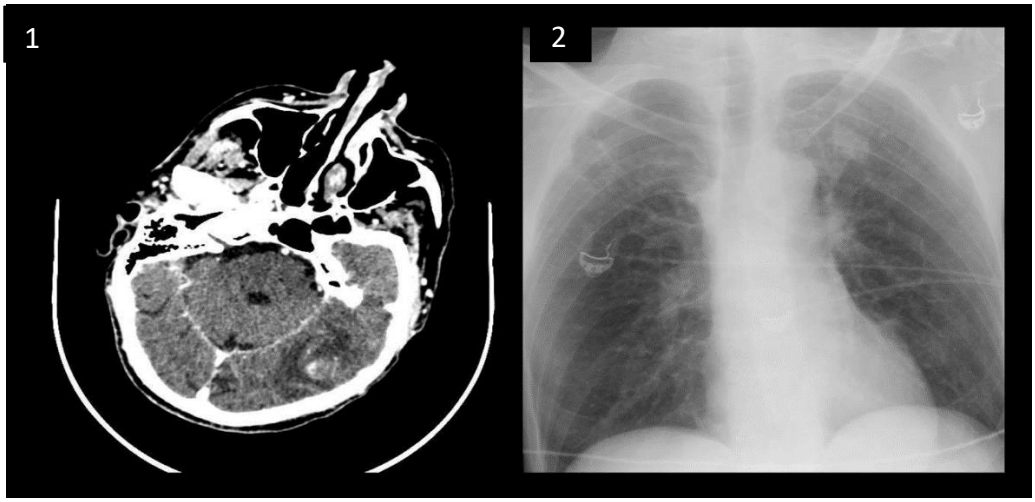


IMAGE 1: Brain CT.

IMAGE 2: Chest X ray.

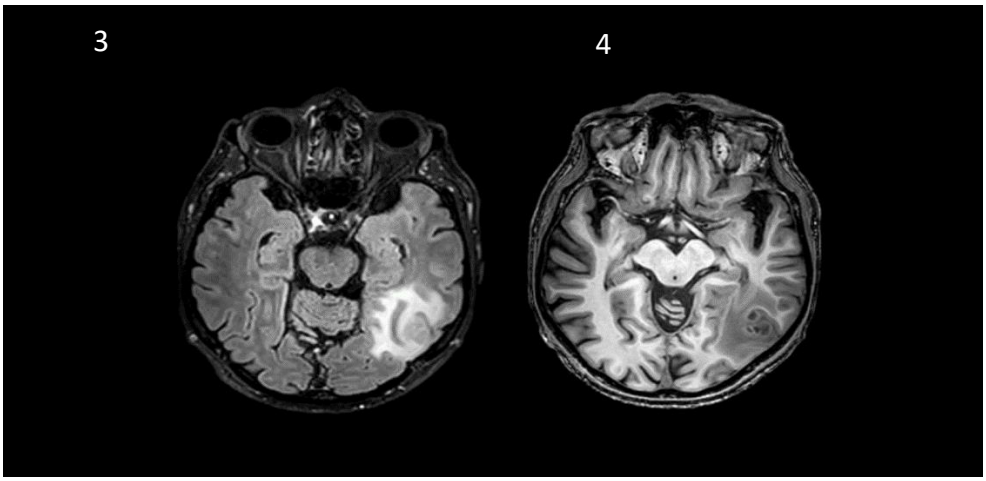
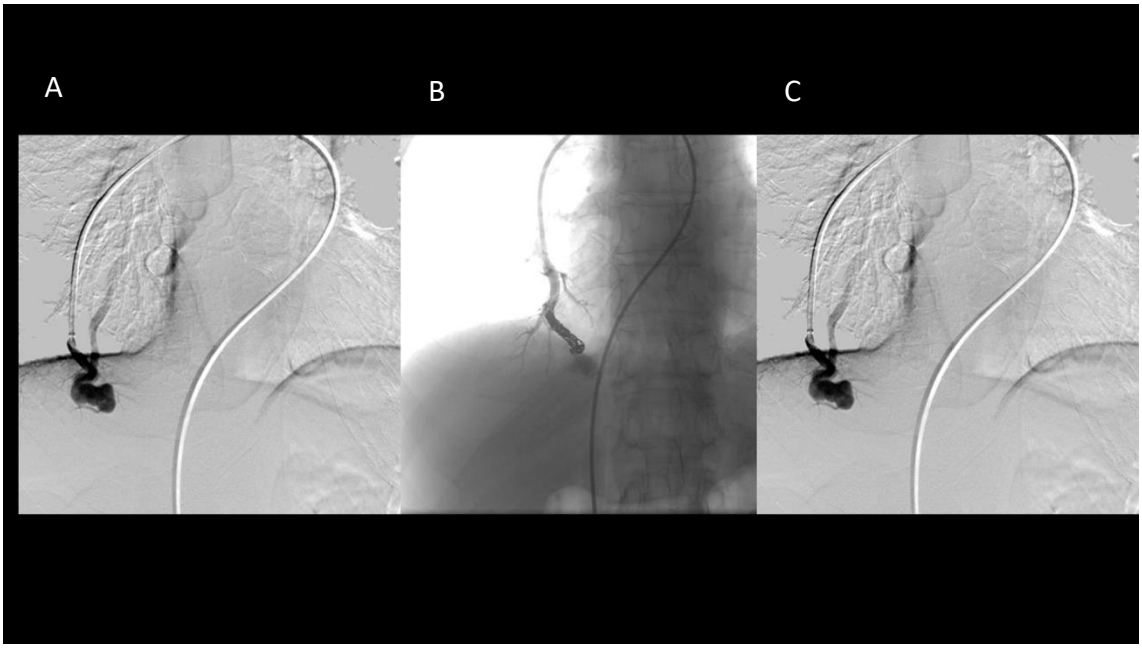


IMAGE 3 – 4 : Cranial MRI

Analytically, total hemoglobin of 12.5g / dL and normal corpuscular volume, absence of leukocytosis and C-reactive protein of 2.2 mg / dL stand out. Anteroposterior chest x-ray was performed without pathological findings (image 2).

The Cranial magnetic resonance imaging (MRI) shows significant diffusion restriction, polylobulated peripheral ring enhancement, and vasogenic edema, all of which are compatible with an abscess as the first possibility (PICTURES 3, 4).

The patient undergoes a craniotomy with evacuation of the purulent material, the culture samples show growth of Gram-positive cocci being identified as Streptococcus constellatus and Parvimonas micra with a "wild" phenotype. In the etiological search, on physical examination, the presence of abundant malar teleangiectasis, petechiae on the lips, oral and nasal mucosa and tongue, with erythema in the Kiesselbach area without signs of bleeding, was striking. Angio-CT study showed the presence of a pulmonary arteriovenous fistula (AVF) in the medial segment of the right lower lobe. Echocardiography with contrast ultrasound with bubbles shows a severe passage of microbubbles to the left chambers from the fourth beat. The patient underwent embolization of said pulmonary AVF (PICTURES 5-6).



IMAGES: A-C: Catheterization and closure of pulmonary AVM 1

Upon meeting the Curaçao criteria, a genetic study was requested with NGS sequencing of the ACVRL1, BMPR2, ENG, GDF2, SMAD4 genes associated with Hereditary Hemorrhagic Telangiectasia, with the probable pathogenic deletion in the chromosomal region 9q34.11, resulting positive.

The Hereditary Hemorrhagic Teleangiectasis (HHT) or Rendu-Osler-Weber syndrome (ROW) is an autosomal dominant disorder with variable expression and penetrance [4], it involves the blood vessels of different body territories, and can cause arteriovenous malformations (AVMs) in up to 50% of patients with HHT, they can be harbored in different locations, such as the lung, liver or brain, the pulmonary being the most frequent (up to 50% of AVMs) [5]. One of the main complications of HHT is the creation of intrapulmonary AVMs (15-45%) [6]. These AVMs can also trigger major complications such as ischemic strokes due to thrombotic emboli or brain abscesses due to septic embolisms [1, 2]. The prevalence of brain abscesses in patients with pulmonary AVMs reaches 9% [5]. The brain locations that are particularly affected in are the basal ganglia and thalamic region [2] and the identified organisms are commonly isolated in periodontal infections [1] as microaerophilic and anaerobic bacteria [5].

TABLE 1 – Curaçao Criteria (from Shovlin C.L. et al., . Am. J. Med. Genet. 91:66-67, 2000)

1. Epistaxis: spontaneous, recurrent nose bleeds
2. Telangiectases: multiple, at characteristic sites (lips, oral cavity, fingers, nose)
3. Visceral lesions such as gastrointestinal telangiectasia (with or without bleeding), pulmonary arteriovenous malformation(AVM), hepatic AVM, cerebral AVMs, spinal AVM
4. Family history:a first degree relative with HHT according to these criteria
Diagnosis of HHT
Definite: 3 criteria are present
Possible or suspected: 2 criteria are present
Unlikely: <2 criteria are present

The diagnosis of HHT is based on the Curaçao diagnostic criteria (8) [TABLE 1], and the currently available genetic panel, of which endoglin (type 1 HHT), a mutation of the gene that encodes the ALK-1 protein (HHT type 2) and the mutation of the SMAD4 gene (overlap syndrome between juvenile polyposis and HHT).

The approach of AVMs is essential, their elimination with interventional radiology methods and intra-arterial embolization has improved the comorbidity and survival of patients with HHT who carry such malformations. Furthermore, this technique has been shown to significantly reduce the morbidity and mortality associated with brain abscesses and ischemic strokes in patients with HHT [6]. However, AVMs are often too small and / or numerous for embolization and many treated patients are left with residual shunts [5]. There are medical options like tranexamic acid or systemic bevazizumab for epistaxis and gastrointestinal bleeding [7].

This highlights the importance of screening programs for the population carrying HHT disease pathogenic mutations [1]. The need to carry out AVM screening programs in patients previously diagnosed with THH is raised, as well as the screening of the disease itself in first-degree relatives of patients affected by this pathology. This could prevent the development of major complications that represent a significant increase in morbidity and mortality in patients with THH disease.

Although HHT can cause premature death, the overall life expectancy is surprisingly good, further improving with medical treatments.

The high rate of patients not identified as carriers of AVMs at the time of stroke or brain abscess development highlights the importance of screening programs for the population carrying pathogenic mutations of HHT [1]. Therefore, there is a need to carry out AVM screening programs in patients previously diagnosed with ROW or THH disease, as well as screening for the disease itself in first-degree relatives of patients with this pathology. This screening could greatly prevent the development of major complications that represent a significant increase in morbidity and mortality in patients with HHT.

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