

**Figure 1.** Cortical activation during motor tasks in the patient. Only the cortex is represented, and the peak of activation in the coordinates were as follows: **A:** left hand,  $x = 38$  mm,  $y = -22$  mm,  $z = 65$  mm; **B:** left elbow,  $x = 42$  mm,  $y = -26$  mm,  $z = 65$  mm; **C:** right elbow,  $x = -33$  mm,  $y = -18$  mm,  $z = 70$  mm. (R, right hemisphere; L, left hemisphere).

extraplexus nerve transfer techniques, the areas of activation mainly being located in the contralateral cortex<sup>(2-8)</sup>.

This study has some limitations. We presented the patient data only in comparison with those of a single control participant, rather than with a group of control, and both data sets were acquired at only one time point. In addition, the patient did not undergo a pre-operative fMRI scan.

**REFERENCES**

1. Oberlin C, Béal D, Leechavengvongs S, et al. Nerve transfer to biceps muscle using a part of ulnar nerve for C5-C6 avulsion of the brachial plexus: anatomical study and report of four cases. *J Hand Surg Am.* 1994;19:232-7.
2. Malessy MJ, van der Kamp W, Thomeer RT, et al. Cortical excitability of the biceps muscle after intercostal-to-musculocutaneous nerve transfer. *Neurosurgery.* 1998;42:787-95.
3. Iwase Y, Mashiko T, Ochiai N, et al. Postoperative changes on functional mapping of the motor cortex in patients with brachial plexus injury: comparative study of magnetoencephalography and functional magnetic resonance imaging. *J Orthop Sci.* 2001;6:397-402.
4. Malessy MJ, Bakker D, Dekker AJ, et al. Functional magnetic resonance

- imaging and control over the biceps muscle after intercostal-musculocutaneous nerve transfer. *J Neurosurg.* 2003;98:261-8.
5. Beaulieu JY, Blustajn J, Teboul F, et al. Cerebral plasticity in crossed C7 grafts of the brachial plexus: an fMRI study. *Microsurgery.* 2006;26:303-10.
6. Sokki AM, Bhat DI, Devi BI. Cortical reorganization following neurotization: a diffusion tensor imaging and functional magnetic resonance imaging study. *Neurosurgery.* 2012;70:1305-11.
7. Hua XY, Liu B, Qiu YQ, et al. Long-term ongoing cortical remodeling after contralateral C-7 nerve transfer. *J Neurosurg.* 2013;118:725-9.
8. Dimou S, Biggs M, Tonkin M, et al. Motor cortex neuroplasticity following brachial plexus transfer. *Front Hum Neurosci.* 2013;7:500.

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**Hughes-Stovin syndrome: an unusual cause of pulmonary artery aneurysms**

Dear Editor,

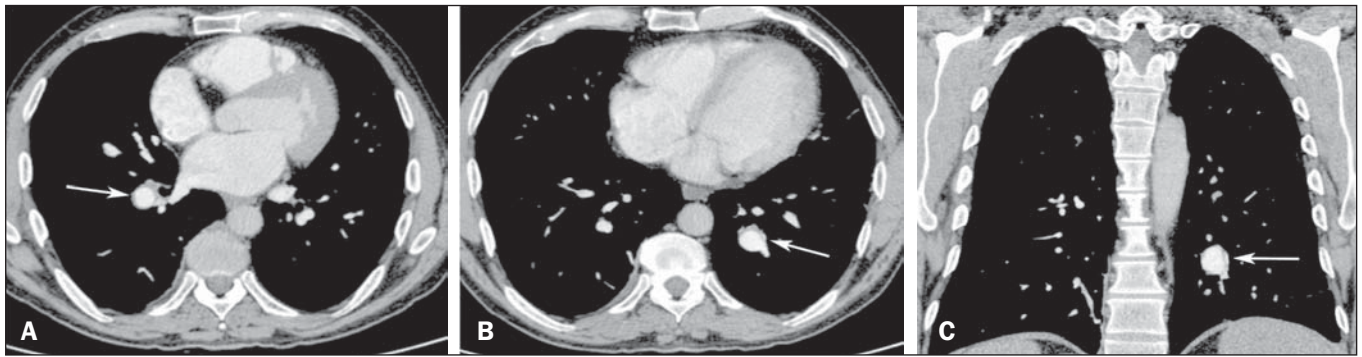
A 43-year-old male presented with a two-month history of persistent cough and fever, associated with recurrent episodes of superficial thrombophlebitis and venous thrombosis of the lower limbs. Physical examination revealed no evidence of oral or genital ulcers. Ancillary tests showed negative blood culture; no thrombophilia or neoplasia; negative serology; mild normocytic, normochromic anemia; elevated C-reactive protein; and elevated erythrocyte sedimentation rate. Contrast-enhanced computed tomography identified aneurysms in branches of the pulmonary arteries (Figure 1). The final diagnosis was Hughes-Stovin syndrome.

Idiopathic and vascular diseases of the thorax have been the subject of recent publications in the radiology literature of Brazil<sup>(1-6)</sup>. Hughes-Stovin syndrome is a rare condition, characterized by the combination of multiple pulmonary artery aneurysms and peripheral venous thrombosis, that mainly affects males (80-90% of cases) between the second and fourth decades of life<sup>(7-11)</sup>. Although the lesions often affect arteries and veins simultaneously

(in 68% of cases), isolated arterial or venous impairments are reported at frequencies of 25% and 7%, respectively<sup>(9,11)</sup>.

In its typical presentation, Hughes-Stovin syndrome occurs in three stages<sup>(7-9,11)</sup>; in the first stage, there are signs and symptoms of thrombophlebitis; the second stage includes the formation and expansion of pulmonary artery aneurysms; and the third stage is characterized by aneurysmal rupture with massive hemoptysis, progressing to death. The formation of pulmonary aneurysms has been attributed to the weakening of the vessel walls by an inflammatory process. Other hypotheses proposed to explain these changes include septic embolism and angiodysplasia of the bronchial arteries<sup>(8,9,11)</sup>. Aneurysms can be single or multiple, unilateral or bilateral, and can even arise at other sites (in the iliac, femoral, popliteal, carotid, or hepatic arteries), although with a lower risk of rupture<sup>(9-11)</sup>.

Some authors consider Hughes-Stovin syndrome an incomplete form of Behcet's disease, due to the similarity between the two in terms of the clinical, radiological, and pathological findings<sup>(7-11)</sup>. Therefore, Behcet's disease, which typically affects young males, is the main differential diagnosis<sup>(11)</sup>. The major (and mandatory) criterion for a diagnosis of Behcet's disease is oral ulcers that recur at least three times within 12 months, which should be



**Figure 1.** Contrast-enhanced computed tomography of the chest, with axial slices (A,B) and coronal slices (C), showing aneurysms in branches of the pulmonary arteries (arrows).

accompanied by at least two of the minor criteria (not necessarily simultaneously), including recurrent genital ulcers, ocular lesions, skin lesions, and a positive pathergy test<sup>(12)</sup>, none of which were observed in our patient. Other causes of pulmonary artery aneurysms are trauma, infection, pulmonary hypertension, and Marfan syndrome<sup>(8–11)</sup>.

There is no standard treatment for Hughes-Stovin syndrome, the most widely used treatment option being immunosuppression therapy involving a combination of glucocorticoids and cyclophosphamide, which has the potential to stabilize aneurysms or even promote regression in some cases<sup>(11)</sup>. The use of anticoagulants is controversial because of the risk of fatal hemoptysis, being allowed only in selected cases and provided jointly administered with immunosuppression therapy<sup>(7–11)</sup>. Other possible treatments include surgical resection and arterial embolization, which are used in most cases in which there is massive hemoptysis<sup>(11)</sup>.

#### REFERENCES

- Batista MN, Barreto MM, Cavaguti RF, et al. Pulmonary artery sarcoma mimicking chronic pulmonary thromboembolism. *Radiol Bras.* 2015;48:333–4.
- Yamanari MGI, Mansur MCD, Kay FU, et al. Bullet embolism of pulmonary artery: a case report. *Radiol Bras.* 2014;47:128–30.
- Pessanha LB, Melo AMF, Braga FS, et al. Acute post-tonsillectomy negative pressure pulmonary edema. *Radiol Bras.* 2015;48:197–8.
- Francisco FAF, Rodrigues RS, Barreto MM, et al. Can chest high-resolution computed tomography findings diagnose pulmonary alveolar microlithiasis? *Radiol Bras.* 2015;48:205–10.
- Koenigkam Santos M. Diagnosis of pulmonary alveolar microlithiasis [Editorial]. *Radiol Bras.* 2015;48(5):ix–x.
- Fernandes GL, Teixeira AA, Ant6n AGS, et al. Churg-Strauss syndrome: a case report. *Radiol Bras.* 2014;47:259–61.
- Cruz VA, Muniz YA, Silva Torres PPT, et al. S6ndrome de Hughes-Stovin. *Rev Bras Reumatol.* 2009;49:747–52.
- Chung MP, Yi CA, Lee HY, et al. Imaging of pulmonary vasculitis. *Radiology.* 2010;255:322–41.
- El Aoud S, Frikha F, Snoussi M, et al. Moderate hemoptysis caused by Hughes-Stovin syndrome. *Clin Pract.* 2014;4:647.
- Silva R, Escobar A, Vega R, et al. S6ndrome Hughes-Stovin: caso cl6nico. *Rev Med Chile.* 2013;141:922–6.
- Khalid U, Saleem T. Hughes-Stovin syndrome. *Orphanet J Rare Dis.* 2011;6:15.
- Belczak SQ, Aun R, Valentim L, et al. Tratamento endovascular de aneurismas da aorta em pacientes com doen7a de Beh7et: relato de dois casos. *J Vasc Bras.* 2010;9:89–94.

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#### Differential diagnosis of anterior sacral meningocele during the evaluation of post-hysterectomy pelvic collections

Dear Editor,

Here, we present the case of a 34-year-old woman who suffered postoperative pain and fever after subtotal abdominal hysterectomy. Conventional radiography of the pelvis showed unilateral sacral curvature (Figure 1A). Computerized tomography (CT) performed on the second postoperative day revealed a dense loculated collection, interspersed with small air bubbles, in the pelvic cavity and a cyst-like formation with hypodense liquid content in the presacral space, communicating with the spinal canal, dislocating the rectum to the right (Figure 1B). A diagnosis of anterior sacral meningocele (ASM) was made, and the surgical team was informed of its coexistence with the postoperative pelvic collections. A new procedure was carried out to drain the collections, care being taken to avoid the sacculation caused by the ASM, which was visible and palpable. Magnetic resonance imaging (MRI) was carried out in order to monitor the postoperative drainage and to characterize the malformation in greater detail (Figure 1C).

Various conditions related to anomalies in central nervous system development have been reported in Brazil<sup>(1–3)</sup>. ASM is a rare form of spinal dysraphism, in which the meningeal sac herniates into the presacral space<sup>(4,5)</sup>. It accounts for approximately 5% of all retrorectal masses and is more prevalent in women<sup>(6)</sup>.

ASM can occur in isolation or in association with other congenital abnormalities, such as urogenital malformations, anorectal malformations, lipoma, teratoma, epidermoid tumor, and dermoid cyst<sup>(7,8)</sup>. Due to its occult nature, it is generally diagnosed in the second or third decades of life. It can be asymptomatic or present with nonspecific symptoms, such as constipation, urological symptoms, and, in rare cases, neurological symptoms<sup>(9)</sup>. The diagnostic investigation can include conventional radiography, ultrasound, CT, and MRI.

With conventional radiography, it is sometimes possible to observe a “scimitar sacrum”, characterized by an unilateral sacral curvature, which is considered pathognomonic for ASM<sup>(10)</sup>. Abdominal ultrasound can show a retrovesical cystic lesion, un-specific to the method<sup>(11)</sup>. CT is an important tool, because it provides detailed information on associated bone alterations and