

in this case^(1,2). MRI shows an expansile lesion, with a broad dural base, that is homogeneous, with a hypointense signal in T1-weighted sequences and an intermediate to hypointense signal in T2-weighted sequences, with moderate to marked gadolinium enhancement^(1,2,4).

There have been few reports of the behavior of histiocytosis in advanced MRI sequences. In our case, the lesion presented low signal intensity in a diffusion-weighted sequence, which is in accordance with the findings of Miyake et al.⁽⁶⁾, possibly secondary to the low cell content of the lesion. Classically, histiocytosis lesions do not show signs of hyperperfusion, because they are essentially lymphoproliferative disorders without neoangiogenesis. However, Hingwala et al.⁽⁷⁾ reported a case in which there was increased perfusion, with high positivity for CD34 and CD31, which are intrinsic markers of vascularization^(1,7). In the case reported here, there were no signs of increased perfusion.

The main differential diagnoses of LCH are forms of non-Langerhans histiocytosis (Rosai–Dorfman disease, Erdheim–Chester disease, and hemophagocytic syndrome), sarcoidosis, tuberculosis, meningioma, hemangiopericytoma, and solitary fibrous tumor⁽¹⁾. Although there is no substantive consensus on the treatment of LCH, it is based on the location and number of lesions, the main therapeutic options being surgery and chemotherapy with various combinations of interferon, vinblastine, cladribine, and methotrexate.

Although uncommon, LCH should be considered in the differential diagnosis of extra-axial expansile lesions in children. It

should be considered especially for lesions presenting an intermediate to hypointense signal in T2-weighted MRI sequences.

REFERÊNCIAS

- Gabbay LB, Leite CC, Andriola RS, et al. Histiocytosis: a review focusing on neuroimaging findings. *Arq Neuropsiquiatr*. 2014;72:548–58.
- Grois N, Fahrner B, Arceci RJ, et al. Central nervous system disease in Langerhans cell histiocytosis. *J Pediatr*. 2010;156:873–81.
- Pyun JM, Park H, Moon KC, et al. Late-onset Langerhans cell histiocytosis with cerebellar ataxia as an initial symptom. *Case Rep Neurol*. 2016; 8:218–23.
- Le Guennec L, Martin-Duverneuil N, Mokhtari K, et al. Neurohistiocytose langerhansienne. *Presse Med*. 2017;46:79–84.
- Haroche J, Cohen-Aubart F, Rollins BJ, et al. Histiocytoses: emerging neoplasia behind inflammation. *Lancet Oncol*. 2017;18:e113–25.
- Miyake Y, Ito S, Tanaka M, et al. Spontaneous regression of infantile dural-based non-Langerhans cell histiocytosis after surgery: case report. *J Neurosurg Pediatr*. 2015;15:372–9.
- Hingwala D, Neelima R, Kesavadas C, et al. Advanced MRI in Rosai-Dorfman disease: correlation with histopathology. *J Neuroradiol*. 2011; 38:113–7.

Bruno Niemeyer de Freitas Ribeiro¹, Bernardo Carvalho Muniz¹, Edson Marchiori²

1. Instituto Estadual do Cérebro Paulo Niemeyer – Departamento de Radiologia, Rio de Janeiro, RJ, Brazil. 2. Universidade Federal do Rio de Janeiro (UFRJ), Rio de Janeiro, RJ, Brazil. Correspondence: Dr. Bruno Niemeyer de Freitas Ribeiro. Instituto Estadual do Cérebro Paulo Niemeyer – Departamento de Radiologia. Rua do Resende, 156, Centro. Rio de Janeiro, RJ, Brazil, 20231-092. E-mail: bruno.niemeyer@hotmail.com.

<http://dx.doi.org/10.1590/0100-3984.2017.0034>



Heterotaxy syndrome

Dear Editor,

A 53-year-old man presented to our neurology department with a progressive, throbbing headache accompanied by focal neurologic deficits. His known medical history included congenital heart disease with dextrocardia, a repaired ventricular septal defect, and a right ventricular-left atrial fistula which had been surgically corrected as well. He also had a long history of unexplained dyspnea. A computed tomography (CT) scan of the head revealed a brain abscess. The patient was admitted to initiate specific therapeutic interventions. However, after admission, he experienced significant worsening of dyspnea, low peripheral oxygen saturation, and cyanosis of the extremities. A chest X-ray showed dextrocardia, an increased cardiothoracic index, and enlargement of the proximal pulmonary arteries (Figure 1), which raised the hypothesis of pulmonary hypertension. Findings on CT angiography, such as severely enlarged pulmonary arteries and filling defects, mainly within the right pulmonary artery, suggested pulmonary hypertension secondary to pulmonary thromboembolism. However, unusual findings were also noted on CT, namely right-sided mediastinum, bilobed right lung, centrally located liver, polysplenia, and abnormal intestinal rotation (Figure 2), all of which were consistent with heterotaxy syndrome (HS). The patient had significant clinical deterioration and died from neurological complications of brain abscess before any curative interventions could be performed.

HS is a rare condition that occurs in approximately 1 in 10,000 live births⁽¹⁾. Patients with HS present with organ arrangement variations other than the typical asymmetry expected in normal anatomy (situs solitus) or its exact mirror image (situs inversus)⁽²⁾. Normal visceral arrangement depends on a series of intricate processes that take place during early mesoderm develop-

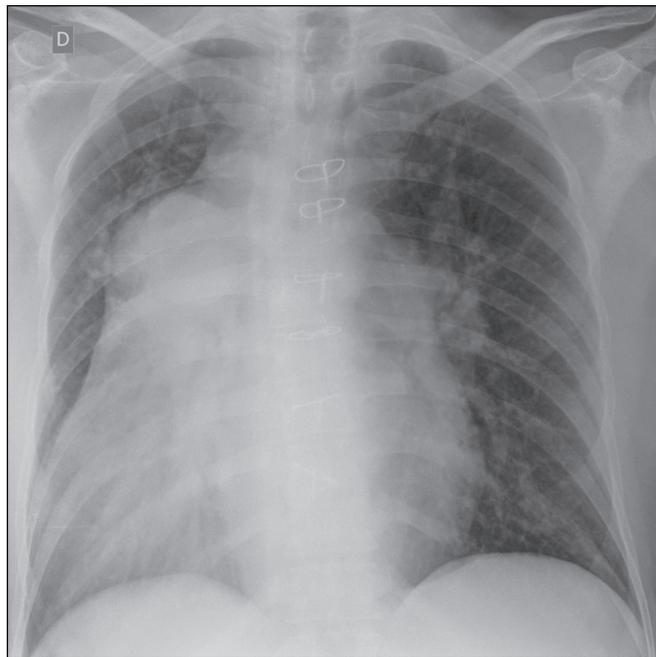


Figure 1. Chest X-ray showing dextrocardia, a widened mediastinum, and enlarged pulmonary arteries suggestive of pulmonary hypertension.

ment, such as adequate expression and leftward flow of growth signals^(2,3). Impairment in any of these factors during organogenesis may lead to abnormal organ positioning and HS.

Patients with HS have historically been classified as having either asplenia (right isomerism) or polysplenia (left isomerism) syndromes⁽²⁻⁴⁾: congenital spleen absence and duplication of right-sided structures characterize the asplenia syndrome, whereas

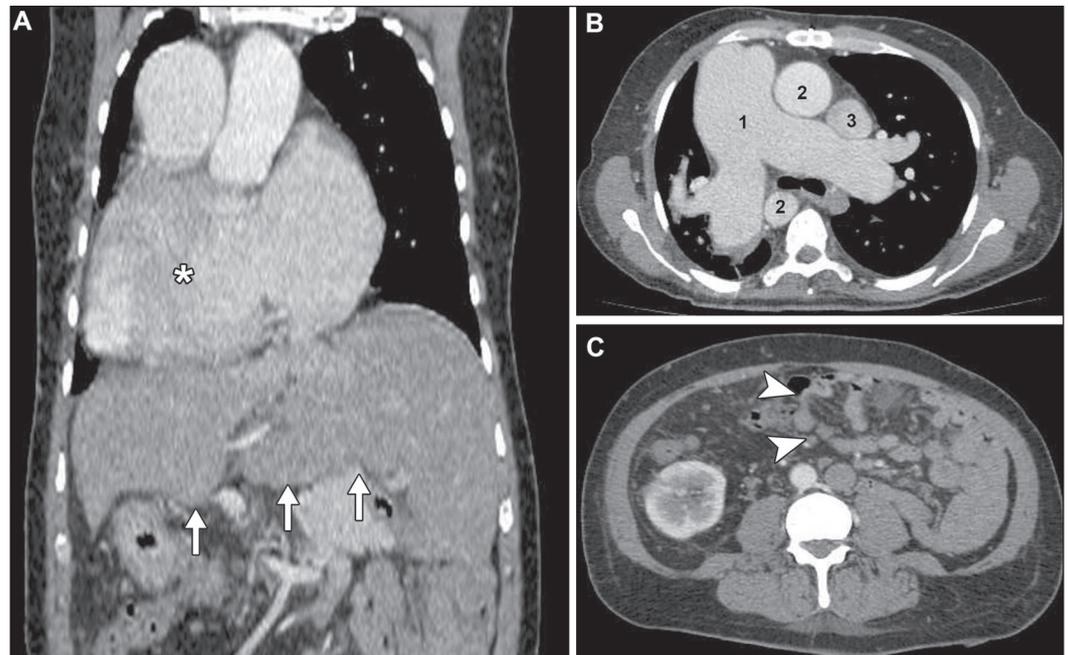


Figure 2. Contrast-enhanced CT scan. **A:** Coronal thoracoabdominal CT scan showing dextrocardia (asterisk) and a centrally located liver (arrows). **B:** Axial chest CT demonstrating enlargement of the right-sided pulmonary trunk, which measured 5.8 cm in its largest diameter (1), right-sided aorta (2) and left-sided superior vena cava (3). **C:** Axial abdominal CT showing abnormal intestinal rotation (arrowheads)—the entire small bowel is positioned to the left of the midline.

the presence of multiple accessory spleens and duplication of left-sided structures illustrate the polysplenia syndrome. Classical findings in HS include cardiac malpositioning, septal defects, bilateral bilobed or trilobed lungs, midline liver, intestinal malrotation, and abnormal spleen development. Intestinal malrotation can lead to gut volvulus and ischemia^(5,6), whereas complete asplenia predisposes to bacterial infections and sepsis^(1,2). Up to 75% of patients with polysplenia have significant cardiac malformations, namely endocardial cushion defects, double-outlet right ventricle, left heart obstruction, and anomalous venous return⁽⁴⁾. The severity of congenital heart disease remains a main determinant of the long-term prognosis of HS patients—even after surgical repair of congenital heart disease, patients are prone to developing arrhythmias, thromboembolism due to right atrium enlargement⁽⁷⁾, and progressive systolic dysfunction⁽³⁾.

In conclusion, HS is a complex syndrome that has remarkable phenotypic variability and is a challenge to manage. Patients with HS are prone to develop potentially life-threatening complications, which should be promptly diagnosed and managed. Therefore, imaging studies are critical in evaluating these patients, because they delineate the spectrum of possible cardiac and extra-cardiac involvement in HS and associated complications.

REFERENCES

1. Teele SA, Jacobs JP, Border WL, et al. Heterotaxy syndrome: proceedings from the 10th International PCICS Meeting. *World J Pediatr Congenit Heart Surg.* 2015;6:616–29.
2. Applegate KE, Goske MJ, Pierce G, et al. Situs revisited: imaging of the heterotaxy syndrome. *Radiographics.* 1999;19:837–52.
3. Shiraishi I, Ichikawa H. Human heterotaxy syndrome – from molecular genetics to clinical features, management, and prognosis. *Circ J.* 2012; 76:2066–75.
4. Ghosh S, Yarmish G, Godelman A, et al. Anomalies of viscerotaxial situs. *AJR Am J Roentgenol.* 2009;193:1107–17.
5. Lee SE, Kim HY, Jung SE, et al. Situs anomalies and gastrointestinal abnormalities. *J Pediatr Surg.* 2006;41:1237–42.
6. Gupta AC, Herts B. Heterotaxia with polysplenia. *J Urol.* 2015;194:801–2.
7. Ferdman B, States L, Gaynor JW, et al. Abnormalities of intestinal rotation in patients with congenital heart disease and the heterotaxy syndrome. *Congenit Heart Dis.* 2007;2:12–8.

Lucas Samuel Perinazzo Pauvels¹, Felipe Welter Langer¹, Daiane dos Santos², Carlos Jesus Pereira Haygert¹

1. Department of Radiology and Imaging Diagnosis, Universidade Federal de Santa Maria (UFSM), Santa Maria, RS, Brazil. Correspondence: Dr. Felipe Welter Langer. Universidade Federal de Santa Maria – Departamento de Radiologia e Diagnóstico por Imagem. Avenida Roraima, 1000, Camobi. Santa Maria, RS, Brazil, 97105-900. E-mail: felipewlanger@gmail.com.

<http://dx.doi.org/10.1590/0100-3984.2017.0063>



Overuse of the hip external rotators: greater trochanter apophysitis in the karate kid

Dear Editor,

A 13-year-old male presented to our institution with an approximately one-month history of pain in both hips that had worsened in the last two weeks, after a soccer match. There was no definitive history of trauma. The patient was a young athlete who practiced soccer and martial arts (karate) regularly. On physical examination, there was tenderness in both hips, with pain that radiated to both thighs and diminished with rest. An X-ray of the pelvis was taken in the emergency department to rule out fractures. The X-ray showed mild irregularity and

sclerosis of both greater trochanters. It was also possible to see small peritrochanteric bone fragments. After a few days, the patient underwent a magnetic resonance imaging scan, which showed insertional tendinopathy and peritendinitis in the obturator internus, gemellus superior, and gemellus inferior muscles (external rotators), bilaterally. There were also irregularities in both greater trochanters, as well as small avulsed cortical fragments with intense bone edema and enhancement (Figure 1A–C). After this initial investigation, clinical and imaging findings suggested bilateral traction apophysitis. Treatment consisted in non-operative management, with good outcome. Clinical follow-up showed good recovery, with complete resolution of the symptoms.