Respiratory Medicine Case Reports 19 (2016) 74–76

Contents lists available at ScienceDirect

Respiratory Medicine Case Reports

journal homepage: www.elsevier.com/locate/rmcr

Case report

Paravertebral pseudotumor in patient with hereditary spherocytosis

Lêda Maria Rabelo [a, *], Vítor Lopes Galvão Vieira [b], Gustavo Rengel dos Santos [b], Giselle Heimbecker [a], Dante Luiz Escuissato [b]

[a] Department of Pneumology, Clinics Hospital, Federal University of Paraná, Brazil
[b] Department of Radiology and Diagnostic Imaging, Clinics Hospital, Federal University of Paraná, Brazil

ARTICLE INFO

Article history:
Received 16 June 2016
Accepted 18 July 2016

Keywords:
Extramedullary hematopoiesis
Hereditary spherocytosis
CT scan
MRI

ABSTRACT

As a rare complication in chronic anemic states, the extramedullary hematopoiesis may provide diagnostic and therapeutic challenge. Caused by the insufficiency of the bone marrow with reactivation of quiescent erythropoietic sites, this condition may vary its presentation as a simple radiologic finding to a spontaneous massive haemothorax. In this paper, we report the case of a 61-years-old female patient with hereditary spherocitosys and paravertebral masses, focusing on clinical and radiological findings in CT and MRI to conclude the tumors etiology and provide adequate care.

1. Introduction

extramedullary hematopoiesis is a rare complication that occurs in anemic states. This is due to a compensatory physiological phenomenon that occurs due to bone marrow failure and their inability to meet the circulatory demands [1]. It may occur in the course of hematological disorders such as thalassemia, hereditary spherocytosis, myelofibrosis, polycythemia vera, leukemia, lymphoma, and another great variety of conditions [2]. The extramedullary hematopoietic tissues maintain the ability to produce red blood cells but they are not able to support this function for long periods, suffering fatty degeneration after long period of time [3].

2. Case report

Patient female, 61 years old, without respiratory symptoms, carrying hereditary spherocytosis. Investigation has shown thoracic masses on chest radiograph. She is a former smoker of 45 packs per year, and stopped smoking twenty years ago. Physical examination showed pale mucous membranes and painless splenomegaly. There was no fever, chest abnormalities, and lymphadenopathy. Laboratory findings showed hemoglobin 6,3g/dl, hematocrit 20,1%; white cell count 6.600/mm3 (neutrophils 65%, 30% lymphocytes, 4% eosinophils); platelets 147.000 cells/mm3; AST: 8UI/l; ALT: 6UI/l, direct bilirubin: 0,55mg/dl; total bilirubin: 2,63mg/dl; albumin: 3,3g/dl; PT/INR: 20,3s/1,44. Chest CT scan showed bilateral paraspinal tumors (Fig. 1). The masses had heterogeneous aspect, with calcifications and fatty attenuation foci, and mild iodinate contrast enhancement. Mild pleural effusion was present. Magnetic resonance imaging showed heterogeneous paraspinal masses, mainly hypointense on T2 and T1-weighted sequences, without restriction of water on diffusion weighted imaging, and mild enhancement of the paramagnetic contrast medium (Figs. 2 and 3). The diagnosis of extramedullary hematopoiesis was considered based on clinical and imaging findings.

3. Discussion

It is well established in medical literature the relationship between extramedullary hematopoiesis and anemic states, mainly in haemoglobinopathies, such as the hereditary spherocytosis. This is probably due to several factors, but the basic mechanism is the bone marrow insufficiency with reactivations of other quiescent sites of erythropoiesis. It may occur in various organs such as the spleen, liver, lymph nodes, thymus, heart, breast, prostate, kidneys, adrenal glands, pleura, retroperitoneal tissue, skin, peripheral nerves, brain, and spinal canal [2].

The incidence of extramedullary hematopoiesis in non-transfusion-dependent patients can reach up to 20% compared to patients with multiple transfusions, conditions in which the incidence remains below 1%, since the demand of bone marrow will be lower. The paravertebral location occurs in 11%–15% of cases and

* Corresponding author.
E-mail address: ledamrabelo@terra.com.br (L.M. Rabelo).

http://dx.doi.org/10.1016/j.rmcr.2016.07.011

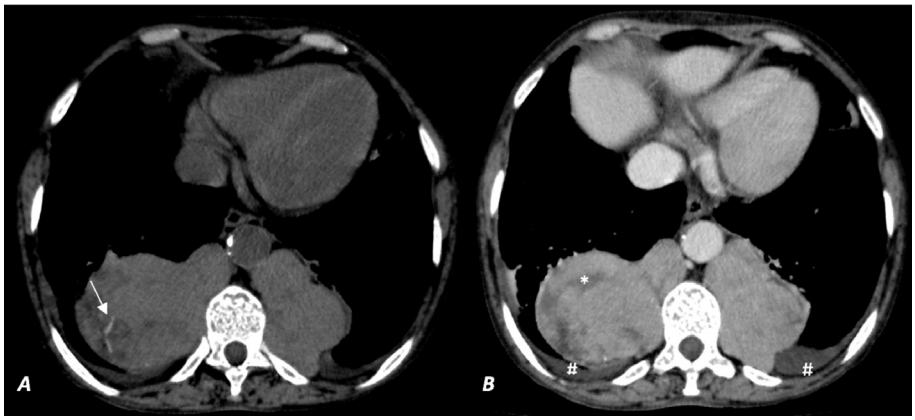

Fig. 1. CT scan. ***A*** (pre-contrast) and ***B*** (post-contrast) shows lobulated paravertebral masses with mild enhancement, linear calcification foci (arrow) and areas of fat attenuation within (*). A small pleural effusion (#) can be seen in both sides.

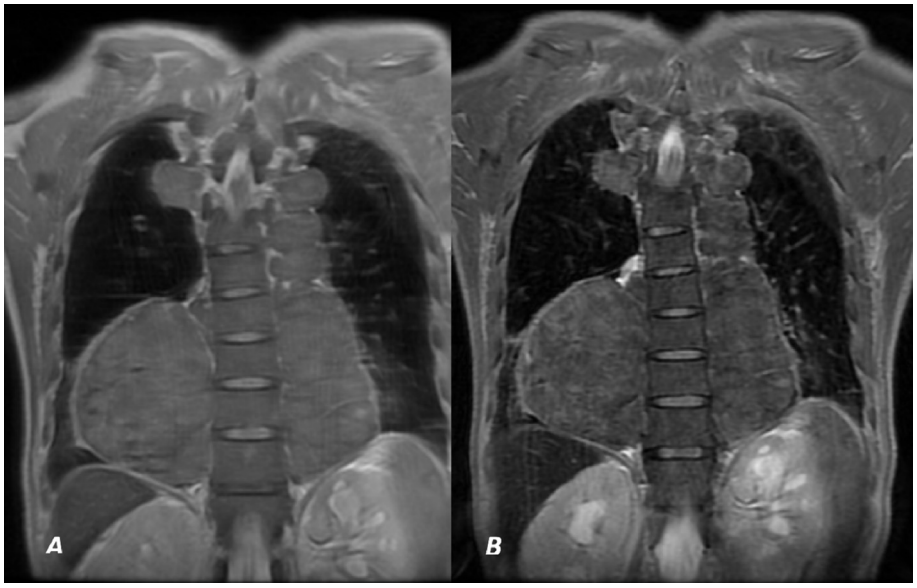

Fig. 2. Chest MRI. ***A*** (T2 weighted image) and ***B*** (STIR) shows heterogeneous, but predominantly hypointense paraspinal tumors.

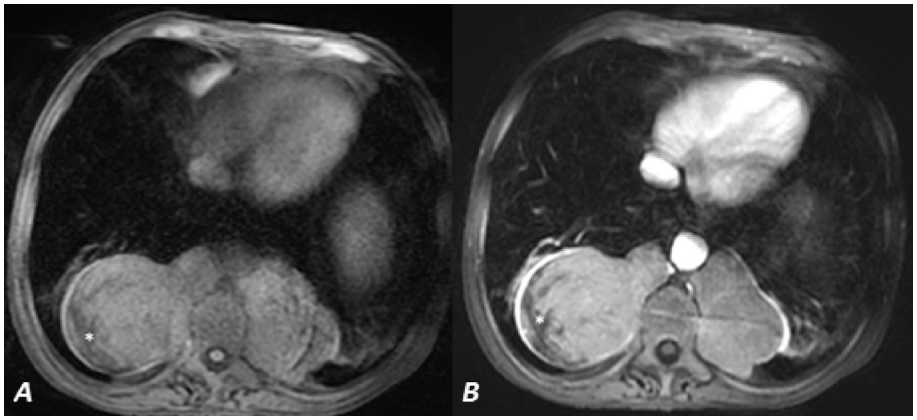

Fig. 3. Chest MRI. Pre (***A***) and post-contrast (***B***) fat suppressed T1-weighted gradient-echo sequences shows mild contrast enhancement and fat foci (*).

the abnormality is more common in men, in a ratio of 5:1 [1,4]. Since the first report in the literature, made by Gatto et al. in 1954, a large number of cases have been described.

Chest radiographs may show lobulated paravertebral tumors and costal deformities. The CT of the chest shows the internal characteristics of the masses, as fat attenuation foci, calcifications

and contrast enhancement [5,6]. The magnetic resonance arose as an important method for the evaluation of this condition. Areas of fat replacement will be bright on T1 and T2-weighted sequences and the areas of iron deposit will show low signal in both T1 and T2-weighted sequences. Contrast enhancement is usually mild to moderate, a finding related to the pattern of vascularization of the lesions [5]. The diffusion weighted sequence is a useful tool, allowing assume a non-neoplastic nature of the masses. The evaluation with nuclear medicine may be important to rule out extramedullary hematopoiesis occurring in the pulmonary parenchyma, when it can be presented as nodules, ground glass opacities, interstitial thickening and signs of pulmonary hypertension [7]. SPECT/CT with technetium-99m sulfur colloid may show increased uptake in the lungs [8].

The symptoms are associated to spinal cord compression, if infiltration of the spinal canal is present, or on paravertebral nerve structures. However, it is believed that over 80% of cases are asymptomatic and discovered incidentally in imaging studies [9].

The treatment of this condition involves transfusion and hydroxyurea (lowering the demand of the insufficient bone marrow), splenectomy in the case of hereditary spherocytosis and small doses of radiation, used mainly in cases of spinal cord compression [2].

The radiological evaluation was crucial in our case to manage the patient as having extramedullary hematopoiesis. There are several reports in the literature of spontaneous massive haemothorax [10–12]. Therefore, the confirmation with histological analysis was put aside due to the high vascularity of the lesion and it was decided to treat the patient to reduce the possibility of post-procedure bleeding.

References

[1] A. Taher, H. Ismaeel, M.D. Cappellini, Thalassaemia inter- media: revisited, Blood Cells Mol. Dis. 37 (2006) 12–20.
[2] R. Haidar, H. Mhaidli, A.T. Taher, Paraspinal extramedullary hematopoiesis in patients with thalassemia intermedia, Eur. Spine J. 19 (2010) 871–878.
[3] L. Pantongrag-Brown, N. Suwanwela, Case report: chronic spinal cord compression from extramedullary haematopoiesis in thalassemia BMRI findings, Clin. Radiol. 46 (1992) 281–283.
[4] S.A. Salehi, T. Koski, S.L. Ondra, Spinal cord compression in betathalassemia: case report and review of the literature, Spinal Cord. 42 (2004) 117–123.
[5] Y.M. Berkmen, et al., Case 126: extramedullary hematopoiesis, Radiology 245 (3) (2007) 905–908.
[6] Christos S. Georgiades, Edward G. Neyman, Isaac R. Francis, Michael B. Sneider, Elliot K. Fishman, Typical and atypical presentations of extramedullary hemopoiesis, Am. J. Roentgenol. 179 (5) (2002) 1239–1243.
[7] M.R. Bowling, et al., Pulmonary extramedullary hematopoiesis, J. Thorac. Imaging 23 (2) (2008) 138–141.
[8] S.Z. Ali, M.J. Clarke, A. Kannivelu, D. Chinchure, S. Srinivasan, Extramedullary pulmonary hematopoiesis causing pulmonary hypertension and severe tricuspid regurgitation detected by technetium-99m sulfur colloid bone marrow scan and single-photon emission computed tomography/CT, Korean J. Radiol. 15 (3) (2014 May-Jun) 376–380.
[9] K. Parsa, A. Oreizy, Nonsurgical approach to paraparesis due to extramedullary hematopoiesis, J. Neurosurg. 82 (1995) 657–660.
[10] P.R. Smith, D.L. Manjoney, J.B. Teitcher, K.N. Choi, A.S. Braverman, Massive hemothorax due to intrathoracic extramedullary hematopoiesis in a patient with thalassemia intermedia, Chest 94 (1988) 658–660.
[11] P. Pornsuriyasak, T. Suwatanapongched, N. Wangsuppasawad, M. Ngodngamthaweesuk, P. Angchaisuksiri, Massive hemothorax in a beta-thalassemic patient due to spontaneous rupture of extramedullary hematopoietic masses: diagnosis and successful treatment, Respir. Care vol. 51 (3) (2006) 272–276.
[12] N. Xiros, et al., Massive hemothorax due to intrathoracic extramedullary hematopoiesis in a patient with hereditary spherocytosis, Ann. Hematol. 80 (1) (2001) 38–40.