



Diffuse pulmonary ossification: A case report



Diogo Roriz*, Inês Abreu, Cristina Marques, Luisa Teixeira, Pedro Belo Soares, Filipe Caseiro Alves

Medical Imaging Department and Faculty of Medicine, University Hospital of Coimbra, Portugal, Praça Mota Pinto/Av. Bissaya Barreto, 3000-075 Coimbra, Portugal

ARTICLE INFO

Article history:

Received 18 January 2016

Received in revised form 20 January 2016

Accepted 20 January 2016

Available online 18 February 2016

Keywords:

Diffuse pulmonary ossification

Mitral stenosis

High-resolution

CT

ABSTRACT

Diffuse pulmonary ossification (DPO) is a rarely diagnosed entity that may present with characteristic imaging features. It is listed in the differential diagnosis of lung parenchymal calcifications and should be considered by the radiologist if the appropriate findings are identified. We report a case of DPO secondary to mitral stenosis in a patient whose severe cardiac pathology lead to death few weeks after a chest CT was done. To date, there are no specific treatments with proved benefit in this pathology.

© 2016 The Authors. Published by Elsevier Ltd. This is an open access article under the CC BY-NC-ND license (<http://creativecommons.org/licenses/by-nc-nd/4.0/>).

1. Introduction

Lung parenchymal calcifications are uncommon, but may occur in numerous conditions so their main differential diagnosis should be known by the radiologist. We demonstrate a rare etiology for lung calcifications in a 87 year-old male that came to our emergency department complaining of dyspnea and was diagnosed with diffuse pulmonary ossification secondary to a long-standing mitral stenosis. Radiographic and CT findings of the patient are described and the most important imaging features of this pathology are here highlighted.

2. Case report

A 87 year-old male was admitted to the emergency department of our hospital referring progressively increased dyspnea over the last two days, together with anorexia in the last months. He had a history of long known moderate to severe mitral stenosis, but due to his age was not considered for valvuloplasty. He was also a smoker (30 pack-year).

The chest radiography done at arrival (Fig. 1) revealed the presence of bilateral micronodular opacities that were more prominent on the right lung, where they were associated with coarse calcifications. A chest CT was suggested to further characterize these findings.



Fig. 1. Chest radiography with postero-anterior incidence demonstrating exuberant calcified opacities, more prominent on the right lung. Calcification of the aortic arch and a tortuous descending aorta can also be seen.

The chest CT (Fig. 2) demonstrated a dilated left atrium associated with dilated pulmonary arteries. An organized pleural effusion on the right. In the lung parenchyma, there were exuberant confluent microcalcifications, more numerous in the lung bases. Despite being bilateral, they were rather asymmetric, being more

* Corresponding author. Fax: +351 239482840.

E-mail address: djr.radiologia@gmail.com (D. Roriz).

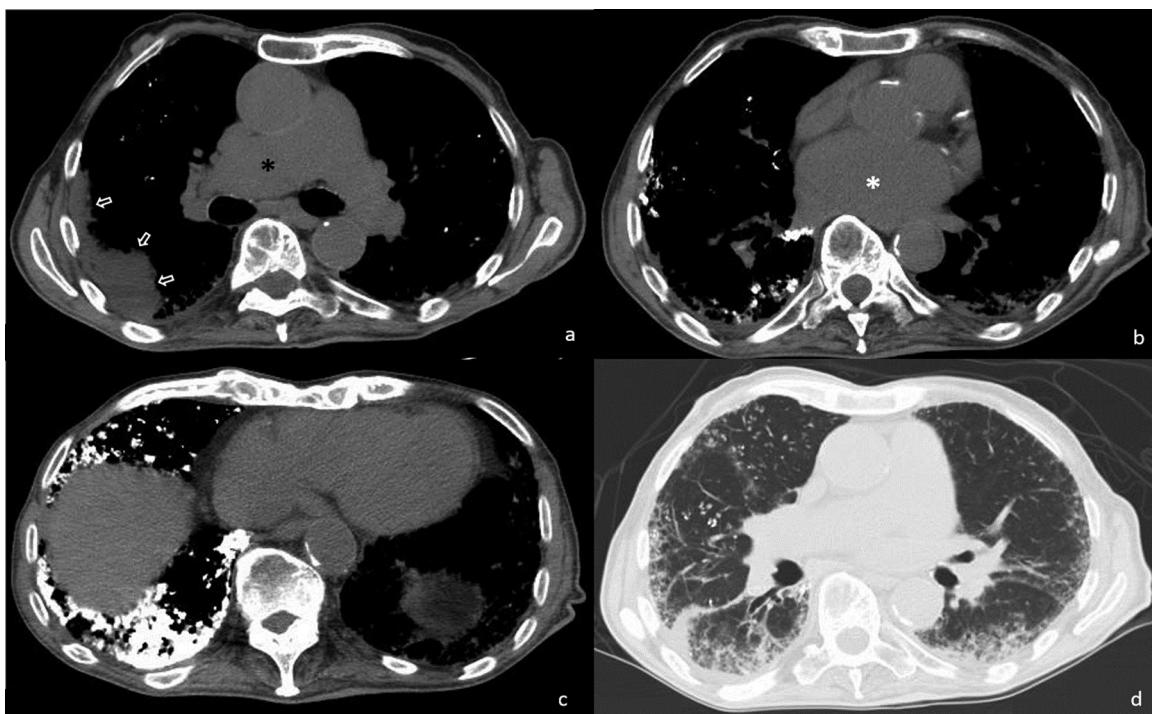


Fig. 2. The non-contrast-enhanced chest CT revealed: a) increased sized right pulmonary artery (black asterisk) and organized right pleural effusion (open arrows); b) dilated left atrium (white asterisk); c) small nodular bilateral calcifications, more prominent on the right lung (also seen in b); d) subpleural bronchiectasis and reticulation.

conspicuous on the right side and rare on the left side. Bilateral subpleural bronchiectasis and reticulation were also seen on the lung inferior lobes.

A diagnosis of diffuse pulmonary ossification secondary to mitral stenosis was suggested based on both the imaging and clinical findings.

Despite the medical therapy instituted, dyspnea of the patient increased over the following days, leading to his death from cardiorespiratory arrest two weeks after this exam. No autopsy was performed.

3. Discussion

Diffuse Pulmonary Ossification (DPO) is a rare diagnosis defined by the deposition of mature heterotopic bone in the lung parenchyma. It either can be idiopathic or secondary, most commonly due to pulmonary or cardiac pathologies, particularly mitral stenosis. The association of pulmonary ossification and mitral stenosis was firstly described by Salinger in 1932 [1]. Estimates establish it to occur in about 3–13% of patients with long-standing mitral stenosis at autopsies [2], but proper medical or surgical treatment of mitral stenosis and eventual underdiagnosis of DPO make it a rarely reported disease in the literature.

A male predominance has been reported with patients being diagnosed more commonly between the 2nd to the 4th decades of life [2,3], although there have been reports in individuals over 60 years-old. A history of repeated insults to the lung, such as recurrent pneumonia, might be present. DPO is believed to be indolent and slowly progressive [4].

Several theories have been proposed for explaining the physiopathology of this disease, focusing on different factors: pulmonary congestion, connective tissue proliferation, hemosiderin deposition or recurrent small pulmonary hemorrhages. Serum calcium and phosphorus levels are usually normal [5].

Patients may be asymptomatic. When symptoms and physical signs are present, they are not different from the ones of mitral

stenosis [3]. Dyspnea and fatigue are common and lower limb edema might also be present due to congestive cardiac failure. Restrictive pulmonary physiology and low diffusion capacity might be present. Hemoptysis are rare [3].

Imaging features play a prominent role in the diagnosis of DPO. Chest radiography is the first-line imaging technique and might reveal signs of enlarged left atrium, such as: cardiomegaly, double right heart border, prominent left atrium and increase of subcarinal angle. It also displays characteristic features such as micronodular calcified opacities that are asymmetrical, usually more conspicuous on the right lung and distributed unhomogeneously, sparing the apices and being more numerous on lung bases. The chest CT, particularly with high-resolution algorithm, is more sensitive in the identification of these features [6]. It may reveal enlarged left atrium and dilated pulmonary arteries, signs of pulmonary hypertension. It can also depict the small calcifications, in the same distribution seen in the chest radiograph, often concentrated near the pleura. They show a tendency to converge and may even form trabeculae [7].

Pathologic findings include the presence of round or mulberry-shaped bone nodules that originally form in the alveoli but may then occupy interstitial air spaces [3]. Osteoblast and osteoid tissue might also be found at the periphery of these nodules.

There is no directed treatment proved to benefit these patients. Treatment must be adjusted to the overlying cause of DPO on secondary forms. Low-calcium diets and systemic steroids have not shown to be effective [5]. Warfarin and bisphosphonates benefit is still to be proven on ossification.

4. Conclusion

Diffuse pulmonary ossification is commonly associated with mitral stenosis, with high-resolution chest CT being the most accurate imaging technique for this diagnosis. Characteristic features include small calcified opacities more numerous on subpleural

locations, especially on the right lung base and eventual signs of pulmonar hypertension. No specific treatment is known.

Conflict of interest

The authors wish to confirm that there are no known conflicts of interest associated with this publication and there has been no financial support for this work that could have influenced its outcome.

References

- [1] H. Salinger, Die Knochenbildungen in der Lunge mit besonderer Berücksichtigung der tuberösen Form, *Fortschr. Roentgenstr.* 46 (1932) 269.
- [2] R. Galloway, E. Epstein, N. Coulshed, Pulmonary ossific nodules in mitral valve disease, *Br. Heart J.* 23 (3) (1961) 297–307.
- [3] W. Wilson, R. Sasaki, C. Johnson, Disseminated nodular pulmonary ossification in patients with mitral stenosis, Ahsassociation. *Circulation* 19 (1959) 323–331, <http://dx.doi.org/10.1161/01.CIR.0000084300.60000>.
- [4] A. Burkett, N. Coffey, N. Voduc, Diffuse pulmonary ossification as a rare cause of interstitial lung disease, *Can. Respir. J.* 21 (1) (2014) 23–24.
- [5] E. Chan, D. Morales, C. Welsh, et al., Calcium deposition with or without bone formation in the lung, *Am. J. Respir. Crit. Care Med.* 165 (12) (2002) 1654–1669.
- [6] M. Konoglou, P. Zaragoulidis, A. Balaika, et al., Lung ossification: an orphan disease, *J. Thoracic Dis.* 5 (1) (2013) 101–104.
- [7] K. Woolley, P. Stark, Pulmonary parenchymal manifestations of mitral valve disease, *Radiographics* 19 (4) (1999) 965–972.