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JCIMCR OPEN ACCESS Journal of Clinical Images and Medical Case Reports

ISSN 2766-7820

Clinical Image

Open Access, Volume 3

A rare cause of chronic respiratory failure in adult

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Received: Sep 25, 2022

Accepted: Oct 24, 2022

Published: Oct 31, 2022

Archived: www.jcimcr.org

DOI: www.doi.org/10.52768/2766-7820/2129

Keywords: Congenital heart disease; Digital clubbing; Respiratory failure.

Background

Tetralogy of Fallot is the most common form of cyanotic congenital heart disease, characterized by ventricular septal defect, overriding aorta, pulmonary stenosis and right ventricular hypertrophy.

A 64-year-old man, ex-smoker, with previous history of multiple myeloma and tetralogy of Fallot non surgically corrected. Admitted to the Pulmonology Department for worsening dyspnea in the last week. During the physical examination, the patient presented peripheral oxygen saturation of 58% and exuberant digital clubbing. Arterial blood gas analysis revealed respiratory acidemia (pH 7.31, pO_2 28, pCO_2 49, HCO_3- 24.7) and analytical study on peripheral blood showed polyglobulia and thrombocytopenia. No changes on chest radiograph. During hospitalization, he was eupneic, despite maintaining respiratory acidemia with bilevel noninvasive ventilation and never presenting pO_2 greater than 40 mmHg with different FiO_2 values. Chest CT confirmed the presence of a complex vascular malformation, with a small pulmonary artery diameter and exuberant collateral circulation in the subpleural region of the RUL and left lung fissure. There was also a right paravertebral mass and some left paravertebral nodules, suggesting extramedullary hematopoiesis. Transthoracic echocardiogram showed alterations compatible with tetralogy of Fallot with a possible right to left shunt due to interventricular communication and dilated and hypertrophied right ventricle. He was discharged home having a global respiratory failure with FiO_2 26% (pH 7.33, pO_2 33, pCO_2 59, HCO_3-31.1) and refused home ventilation.

This case report illustrates a rare cause of chronic respiratory failure in adult, with no relevant clinical improvement with oxygen or noninvasive ventilation therapy. It is also noteworthy for being a patient who refused surgical treatment and has long term survival comparing with other works described in literature.

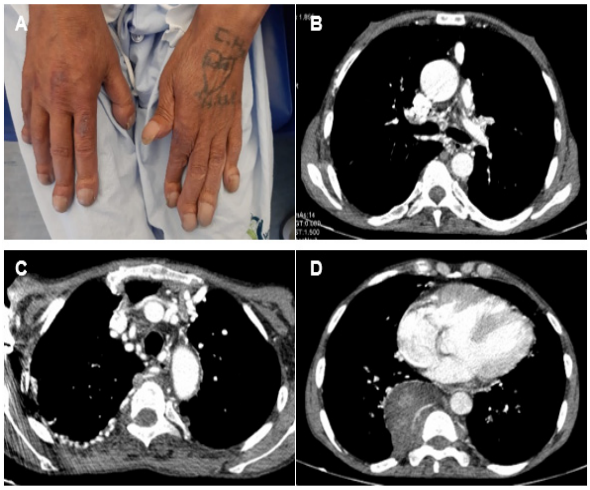

Figure 1: (A): Digital clubbing. (B,C): Chest CT confirmed the presence of a complex vascular malformation with a small pulmonary artery diameter and exuberant collateral circulation in the subpleural region of the RUL and left lung fissure. (D) Right paravertebral mass and some left paravertebral nodules, suggesting extramedullary hematopoiesis.