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Case Report Open Access

Klippel-Trenaunay-Weber Syndrome and Heart Failure, an Uncommon Presentation

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Abbreviations: KTWS: Klippel Trenaunay Weber Syndrome; LA: Left Atrium; LVHY: Left Ventricular Hypokinesia; EF: Ejection Fraction

Introduction

Klippel-Trenaunay-Weber syndrome is a rare congenital disorder characterized by the triad of capillary malformations, bony or soft tissue hypertrophy usually of lower limbs, and venous varicosities or malformations [1], both genders are equally affected. The prevalence is estimated 1 in 20 000 to 1 in 100 000 live births [2].

Case Report

A 25-year-old male with Klippel - Trenaunay - Weber syndrome (KTWS) presented with a new onset atrial fibrillation in May 2013 without reversion to sinus rhythm. His medical history included varicose disease in lower limbs (Figure 1), mental retardation and a history of progressive dyspnea. Initial assessment was done with an echo showing a dilatation of left atrium (LA), generalized left ventricular hypokinesia (LVHY), ejection fraction (EF): 40% with preserved right ventricular function, moderate mitral insufficiency, pulmonary and tricuspid mild insufficiency without evidence of pulmonary hypertension (Figure 2). A multidetector computed angiotomography showed normal coronary arteries (Figure 3), global LVHY, EF: 40% and dilated LA (Figure 4), then a cardiac magnetic resonance revealed the same global LVHY, and atrial dilatation, with no late gadolinium enhancement or patognomonic findings of other disease. A lower limb magnetic resonance revealed varicose dilatation of the superficial venous system of both lower limbs. He has no history of myocarditis and an enzyme-linked immunosorbent assay and indirect fluorescent antibody tests were negative for Chagas disease. The patient receive therapy with metoprolol 50mg twice a day, furosemide 20mg twice a day, spironolactone 25mg a day and acenocumarol. After a follow, up of 5 years the patients maintain the same ejection fraction (40%) without progression of mitral insufficiency and with a New York Heart Association Class I status.

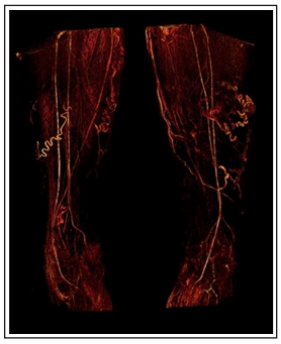

Figure 1: Tomographic reconstruction of varicose disease in lower limbs.

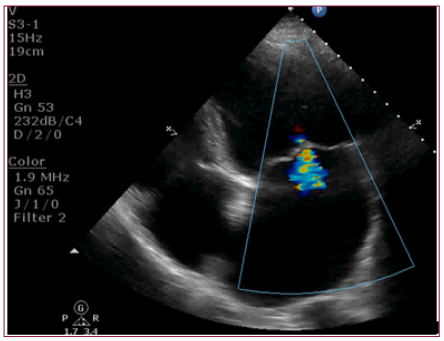

Figure 2: Echocardiogram, left atrial enlargement and mitral insufficiency.

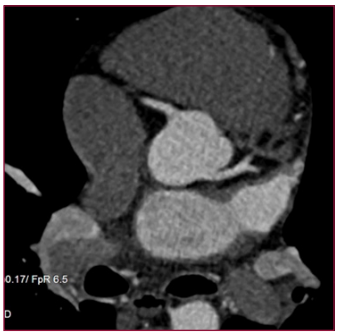

Figure 3: Coronary CT angiography without lesions.

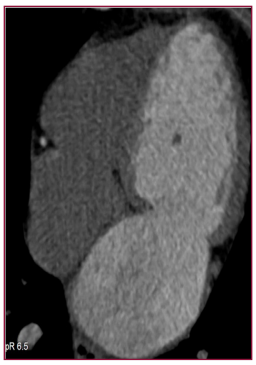

Figure 4: CT showing left ventricular and atrial enlargement.

Discussion

Some theories suggest a mesodermal abnormality during fetal growth, abnormal regulation, or production of growth factors affecting angiogenesis in veins [3]. Major characteristic is hemihypertrophy, subcutaneous masses of various types, such as lipomas, hamartomas, hemangiomas, lymphohemangiomas, and epidermal nevus, are major findings in more than one-half of the patients. The case presents a diagnosis of KTW syndrome showing varicose disease in lower limbs, mental retardation and a history of progressive dyspnea with a new onset atrial fibrillation with an enlarged left atrium and low ejection fraction without an identifiable cause, there are some reports of chronic thromboembolic pulmonary hypertension [4] and heart failure [3,5,6]. Heart failure could be caused by chronically increased preload associated to extensive varicosities of superficial and deep veins especially after the exclusion of the vast majority of causes of heart failure.

Conclusion

The presence of heart failure in an adult patient with Klippel-Trenaunay-Weber syndrome has to be studied trying to find a conventional cause but considering that the syndrome itself can promote the development of chronically increased preload or an enlargement of pulmonary trunk that causes chronic increase in the cardiac index and severe pulmonary hypertension.

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