Pulmonary Emphysema As A Cause Of Rib-notching
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Citation

Abstract
A rare case of unilateral rib notching in a patient with pulmonary emphysema is reported. Vascular alterations of the great vessels were ruled out and the right upper lobe (RUL) showed perfusion deficit, which was coincident with the rib notching site. A systemic arteriography proved the occurrence of dilated and tortuous intercostal arteries which caused notching. A review of the literature revealed no well documented case of pulmonary emphysema associated with rib notching.

INTRODUCTION
Rib notching consists of the erosion of the superior or inferior rib margin, showing a characteristic appearance in the chest roentgenogram. The erosion in lower regions is due to persistent pulsation of the dilated intercostal arteries. Coarctation of the aorta remains, by far, the most common cause of notching in the inferior aspects of the ribs, which usually causes notching in the inferior aspects of the third to ninth ribs1. Tetraplegia as a spinal cord lesion is the most common cause of upper marginal rib notching2.

Although the pulmonary emphysema has been described as a likely cause of inferior rib notching3, we found no well-documented case in the literature review.

CASE REPORT
A 42-year old male patient, who smoked 26 packs-year and suffered from progressive dyspnea for several years. On physical examination, asthenia, low respiratory murmur and clubbing in his hands were found. A chest roentgenogram showed air-trapping in the upper lobes with notching in the lower zone of the posterior regions of the fourth and fifth right ribs. (Figure 1).

The CT scan revealed clear signs of bullous emphysema mainly in the upper lobes. Alpha-1-antitripsin levels were normal. The functional respiratory tests showed a slight
obstructive alteration (FEV1 75%), increased RV (155%) and decreased CO diffusion (DLCO 54%, KCO 51%). ECG and arterial blood gas determinations were normal. Magnetic resonance of heart and great vessels revealed no cardiovascular alterations. An arteriography of the aorta and its branches showed remarkable dilatation of the right intercostal arteries. Bronchial arteries were dilated and tortuous. Fistulous communication between the intercostal, subclavian and internal mammary with the pulmonary circulation was observed (Figure 2-a). Lung scintigraphy showed no perfusion in the right upper lobe and heterogeneous distribution in both lungs (Figure 2b).

**DISCUSSION**

The location of the rib notching is of importance to determine the etiological diagnosis. Notching is less common in the upper margin than in the lower one, although they could sometimes be overlooked since they are often slightly visible in the chest x-ray. Rib notching may result from the absence of a stimulus secondary to the repeated contraction of the intercostal muscles, as occurs in tetraplegia.4

The most common cause of lower notching is the coarctation of the aorta, being unilateral or bilateral according to the level of the coarctation. From a hemodynamic standpoint, it is possible for rib notching to develop into other cardiovascular conditions due to increased intercostal arteries blood supply in the opposite direction to the aorta, unlike to the coarctation of the aorta. Among these conditions are the thrombosis of the abdominal aorta, the obstruction of the subclavian artery (Blalock-Taussig’s operation and Takayasu’s arteritis7) and those which result in decreased pulmonary artery supply (Tetralogy of Fallot, pulmonary atresia, pulmonary valve stenosis, Ebstein’s malformation and pulmonary emphysema). The pulmonary emphysema has been reported as an unusual cause of rib-notching.3

There are other disorders of venous aetiology (superior vena cava obstruction), due to vascular shunt (pulmonary or intercostal arteriovenous fistula), neurogenic (intercostal neurinoma) and osseous (hyperparathyroidism or idiopathic). The idiopathic forms are more common than expected. The location of the rib-notching may be helpful to differentiate the pathological forms from the normal ones. The pathological forms are usually in the mid-third of the posterior arcs of the ribs. In this patient the notching was unilateral, inferior and were located in the posterior mid-third.

The pulmonary emphysema may often be associated with a vascular deficit located in one or more pulmonary regions, the vascularization being normal or increased in the remaining areas. In this patient the region of absence of perfusion was consistent with the less ventilated emphysematous area. This progressive diminishment of perfusion in this area leads to the occurrence of intercostal collateral circulation, in a compensatory form. At that level, dilated intercostal arteries develop, as it is shown in the arteriography, which contact with the lower margin of the upper ribs, resulting in rib-notching.
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In a post-mortem study including 18 cases of pulmonary emphysema, diminished bronchial circulation was observed which compensated with increased pulmonary systemic circulation, by precapillary anastomoses leading to widening and tortuosity of the intercostal arteries which result in rib-notching.

In our patient the deficit located on the pulmonary vascular bed with the presence of bullae, led to the hypertrophy of the intercostal vascularization at that level and the occurrence of rib erosion in this area.

References

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