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RESEARCH ARTICLE

PLATYPNEA-ORTHODEOXIA SYNDROME : AN ATYPICAL CLINICAL CASE

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ABSTRACT

Platypnea-orthodeoxia is un uncommon syndrome associating dyspnea and refractory hypoxemia induced by upright posture. Multiple etiologies can be brought upfront: cardiac shunt (patent atrial septal defect), pulmonary resection and hepatopulmonary syndrome amongst others. Its diagnosis is made by ultrasonography, often misled by its nonspecific clinical presentation. We present an atypical case of patent atrial septal defect. In our clinical case, the latent defect had been made patent by an architectural cardiac modification following a bulky pericardial cyst located near the right lung base. The therapy is that of the shunt, surgical or percutaneous.

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INTRODUCTION

We report a clinical case about a 72 y.o. women presenting dyspnea with the least effort, free of chest pain. The chest x-ray (expiration and inspiration views) show a reduction of right diaphragmatic mobility (Fig.1). The spirometric breathing patterns are normals. The CT-scan highlights a basi-thoracic mass of 12,5 x 12,5 x 5,7cm associated with an elevated diaphragm dome (Fig.2). This fluid content lesion has no glycolytic activity on the TEP/CT. Overnight oximetry shows a pulse oximetry under 90% for 1 hour 52 minutes. A mild left ventricular hypertrophy, normal left ventricular systolic function, no signs of pulmonary arterial hypertension or of inferior vena cava dilatation. The diagnosis of right diaphragmatic palsy due to the voluminous pericardial cyst is made, explaining the dyspnea. A complete cyst resection is made via a right lateral thoracotomy (7th intercostal space) procedure. The pericardial cyst's collar is identified then ligated. The immediate post-operative period is complicated by a severe refractory hypoxemia and associated dyspnea, requiring an ICU transfert.

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The ICUE medical staff soon notice the relief on respiratory symptoms of the left lateral decubitus. Pulmonary embolism, lung contusion, atelectasis and bronchial surinfection are excluded by further investigations. Episodes of normal oxygen level (100% SO2) on left lateral decubitus opposed to hypoxemia (80% SO2) on upright and seated position led to the diagnosis of platypnea-orthodeoxia syndrome. Transoesophageal echocardiography associated with high heart catheterism (Fig.3) explains the symptoms by the presence of a patent foramen ovale without pulmonary hypertension signs. The pericardial cystectomy might have resulted in an architectural modification of the mediastinum, leading to an alignement of the inferior vena cava to the foramen ovale (Fig 4.), creating a right-left positional shunt. Another explanation might be the transformation of a latent to a patent foramen ovale after the anatomical intra-thoracic modification induced by the surgery. The patient has been transferred to a university hospital for endovascular patent foramen ovale closure. The respiratory symptoms quickly amended after treatment.

DISCUSSION

Platypnea-orthodeoxia syndrome is the association of dyspnea and refractory hypoxemia induced by upright or seated position.Its etiology is one of an intracardiac right-left shunt

without pulmonary hypertension. Orthodeoxia is defined by an oxygen blood level diminution of 4% or more. (Zerhouni, 2013). This rare and misdiagnosed entity was first described by Burchell in 1949 although the term of orthodeoxia wasn't used for 20 more years (Zerhouni, 2015). This shunt is most often due to an interatrial communication such as a patent foramen oval. Many other pathological situations can lead to the development of the platypnea-orthodeoxia syndrome. Patent foramen ovale is a frequent anomaly leading to a left to right shunt due to higher left atrium blood pressure. As much as 20 to 30% of the population is carrier of this anomaly, often asymptomatic and requiring no further investigations or treatment.

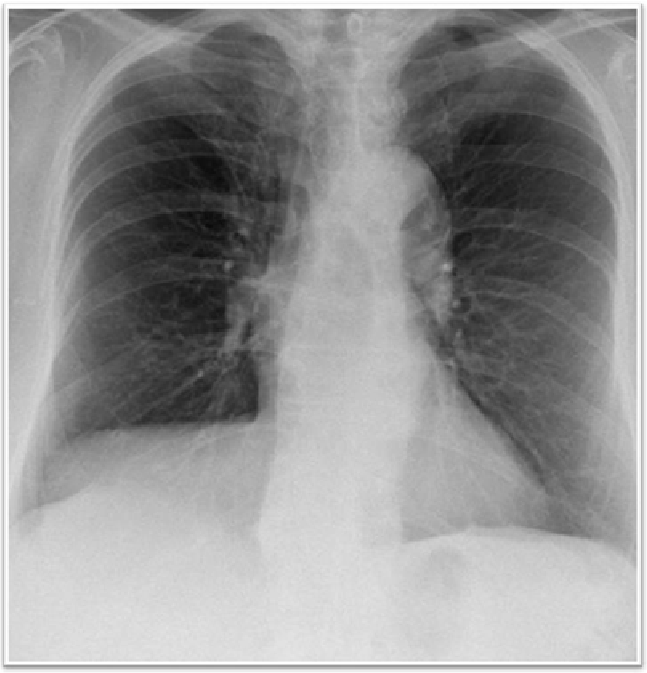

Fig. 1. Chest X-Ray

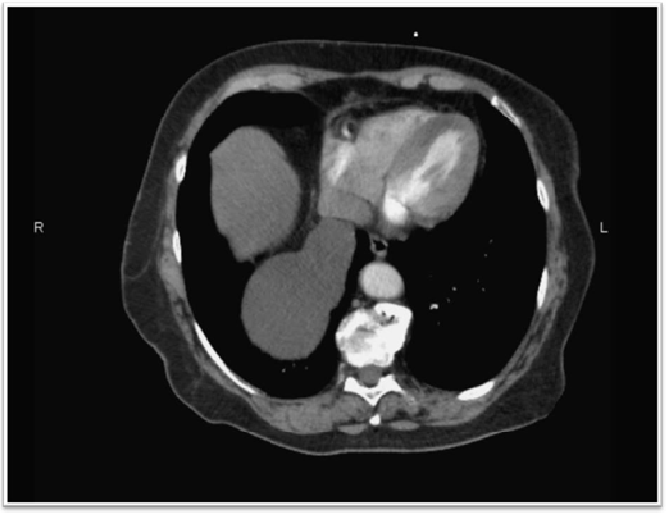

Fig. 2. Basithoracic CT-Scan

In rare cases, it can lead to this platypnea-orthodeoxia syndrome or be the cause of migraine with aura. (Calvert, 2011 and Pierce, 2010). The physiopathology is complex due to the postural variations of the inter-atrial septum, with a reorientation of the vena cava flow trough the patent foramen ovale. This flow occurs without a measurable dynamic pressure gradient. 6 Nowadays, physipathologic mechanisms leading to a right-left atrial shunt whiteout pulmonary hypertension are unknown and subject to debate. Anatomical modification of the atria or the mediastinum may be the cause of this flow modification (Zerhouni, 2013; Desouza, 2009 and Pierce, 2010).

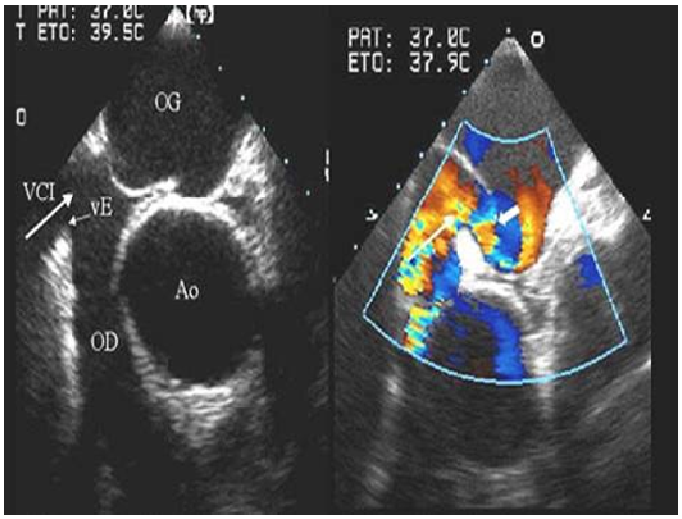

Fig. 3. Transoesophageal echocardiography

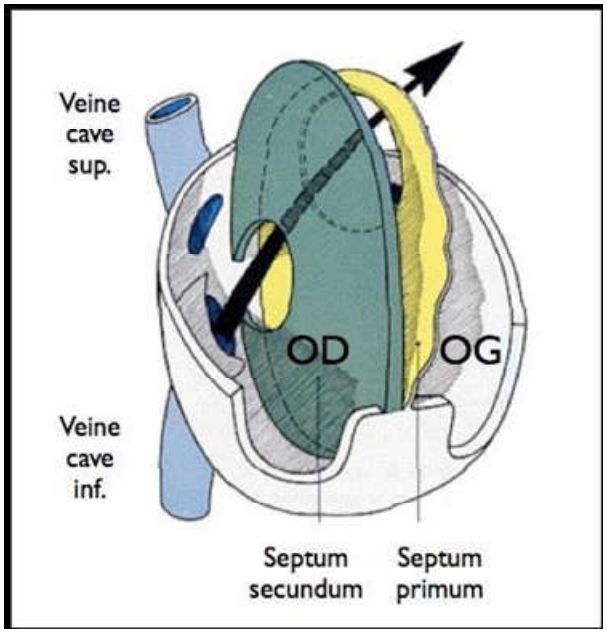

Fig. 4. Alignment of the inferior vena cava to the foramen ovale

However, two components must be present in order for the platypnea-orthodeoxia syndrome to occur : an anatomical interatrial communication such as a patent foramen ovale and a functional modification leading to an inter-atrial deformation and thus a flow reorientation of the vena cava. 10 The diagnosis is first made by pulse oxymetry. The lack of blood oxygen saturation after an hyper oxygenation test must lead to this diagnosis. A bubble contrast and a transoesophagial echocardiogram with tilt-test are the diagnosis gold standards. The transoesophagial echocardiogram is more specific and helpful than the transthoracic echocardiogram (Desouza, 2009; Roxas-Timonera, Miriam, 2001 AND Al Khouzaie, Thamer, 1997). 2,7,8 The therapeutical approach is one of the shunt : surgical or percutaneous with a foramen ovale occluder device (ombrella).

The latter being less invasive, with less complications and granting a durable right-left shunt occlusion (97% good results). The complications are scarce and durable (Schwerzmann, 2004).

Conclusions

Platypnea-orthodeoxia syndrome is a rare clinical entity that must be brought upfront in any case of positional dyspnea and refractory hypoxemia. The diagnosis is made by at least an echocardiogram, be it transthoracic or transoespophagial, to locate the right-left shunt. The treatment is one of the shunt, immediate closure by surgical or percutaneous means. The clinical amelioration after closure is most often spectaculary. The only contraindication is a chronic pulmonary artery hypertension (PAP >5 50-70% of inter-atrial pression).

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