To the Editor,

We report the case of 72-year-old man, former smoker, with moderate chronic obstructive pulmonary disease (post-bronchodilator FEV1, 69%), and previous left pneumonectomy for squamous cell lung cancer with invasion of the pulmonary artery.

He presented in the emergency department of our hospital 2 months post-surgery with progressively worsening dyspnea, even on minimal exertion, aggravated in a sitting or upright posture.

Clinical signs included tachypnea, oxygen saturation (SatO2) 88%, d-dimer 1226 ng/ml, and arterial blood gases pH 7.42, pO2 65.1 mmHg, pCO2 31.2 mmHg and HCO3 20.1 mmol/l. Post-surgical changes were seen on chest X-ray, and no signs of pulmonary embolism were found on lung scintigraphy/SPECT-CT.

In view of the patient’s history of lung surgery and oxygen desaturation in an upright posture, platypnea–orthodeoxia syndrome (POS) was suspected.

SatO2 was tested in supine and sitting positions, showing a significant fall in hypoxemia in an upright posture (from 93% to 88% with nasal cannulas at 4 l/min). Transthoracic echocardiography (TTE) with bubble study revealed a right-to-left shunt due to a patent foramen ovale (PFO). These findings were confirmed by transesophageal echocardiography (TEE).

The patient underwent heart surgery for percutaneous closure of the PFO, using the Amplatzer® Cribriform occluder (Fig. 1). Successful placement of the device and absence of residual shunt was confirmed with TEE. In subsequent follow-ups, the patient’s dyspnea was fully resolved and standing SatO2 was normal.

POS is an unusual phenomenon, consisting of dyspnea in an upright posture, which diminishes in recumbency (platypnea), accompanied by increased blood oxygen levels in that posture (orthodeoxia).

Causes of POS can be divided into 3 groups, depending on the pathogenesis: cardiac, pulmonary or other.1,4

Two components are required in cardiac POS, one being anatomical (common in all patients) and the other functional. The anatomical component must involve an intracardiac communication causing shunt. The most common defect in the general population and in POS patients is PFO. PFO does not generally cause disease because it causes left-to-right shunt, but this reverses in the presence of certain functional changes.

The most common functional components include ascending aorta aneurysm, pneumonectomies (most often right-sided) and lobectomies, pericardial and myocardial disease, and tricuspid valve disease. The reduction in blood flow experienced in recumbency causes the shunt to diminish, with the consequent rise in SatO2.

A high level of suspicion is needed in the diagnosis of POS, and a careful anamnesis is essential if it is to be identified.

The most simple, but no less important, examination is the determination of SatO2 in different postures. Definitive diagnosis of cardiac POS is reached with bubble-contrast TEE, which produces fewer false-positives than TTE. Both tests may be normal when the patient is supine, or if microbubble contrast medium is administered via an upper limb. This is because in PFO, the flow is received primarily by the inferior vena cava.

Cardiac POS treatment consists of closing the PFO with Amplatzer® devices, resulting in resolution of symptoms5 and normal SatO2 in an upright posture.

Fig. 1. Spectroscopy image in hemodynamics unit. Unfolded Amplatzer® Cribriform 25 mm occluded before release in the interatrial septum (A). Transesophageal echocardiogram 3-dimensional image showing device after percutaneous closure of the PFO (B).

Hematic Pleural Effusion in a 17-Year-Old Woman

Derrame pleural hemático en mujer de 17 años de edad

To the Editor,

We report the case of a 17-year-old woman with a history of well-controlled allergic asthma, diagnosed in November 2010 with a right frontal space-occupying lesion, consistent with WHO grade III anaplastic ependymoma. She was treated the same month with resection of the mass and, one month later, was operated again for local recurrence. She subsequently received a 3-month course of radiation therapy.

Five months after the initial diagnosis, the patient was referred to our respiratory medicine department due to a 1-week history of dyspnea. On physical examination, she showed signs of pleural effusion in the lower two-thirds of the left hemithorax, so a diagnostic thoracocentesis was performed. The pleural fluid sample was consistent with uncomplicated exudate with mildly predominant mononuclear cells and a very low glucose level. Pathology study results were negative. In the 6 days following her first visit to the clinic, she required 2 evacuating thoracocentesis procedures for increased dyspnea, so she was hospitalized for further study. On day 7 after admission, in view of her progress, a medical diagnostic and therapeutic thoracoscopy was considered, but this was abandoned due to deterioration of her general condition, and poorly controlled dyspnea. Her clinical progress over the following 24 h worsened and she became moribund. Sedation was administered and the patient died on day 8 of admission, from respiratory failure.

In view of the lack of a confirmed diagnosis of the left pleural effusion in a patient with the above-mentioned history, an autopsy was requested, which revealed multiple pleural implants (Fig. 1). The definitive diagnosis was anaplastic ependymoma (WHO grade III), with tumor recurrence in the frontal lobe of the brain and extended metastatic disease, primarily in the pleura and also in the peritoneal serous membrane, subcutaneous tissue of the cranial vault, and the meninges.

Ependymoma is one of the rarer glial tumors that typically occur within or adjacent to the ependymal cells lining the ventricles of the brain. It accounts for 3%–9% of all neuroepithelial tumors. Four histological subtypes have been described: classic, anaplastic, myxopapillary, and subependymoma.1,2 It occurs equally in both sexes and mean age at diagnosis is 5 years. The most widely accepted

therapeutic option to date is complete surgical resection followed by adjuvant radiation therapy. This generally yields a 5-year survival rate of 60%–89%, in contrast to incomplete resection, for which 5-year survival is less than 50% (21%–49%). Chemotherapy has also been applied; initial response is good in most cases but not all.3 Despite advances in diagnostic and therapeutic techniques, 50% of cases present recurrence or disease progression.4,5

Extracranial metastasis is uncommon in these tumors (0.5%–0.98% of cases).4 The main sites are the lymphoid organs and viscera (particularly the lung). Time between the appearance of the primary tumor and metastatic involvement is usually about 8 years, although it can be shorter.

In conclusion, our patient’s presentation and progress is similar to the pattern observed in the literature to date, although the time between appearance of the primary tumor and respiratory symptoms was shorter, so her death, 6 months after diagnosis, was earlier. Her disease also appeared to be more aggressive than cases reported by other authors, with metastasis in the surgical site and in distant sites, developing within a very short period of time. Thus, in the case of anaplastic ependymoma WHO grade III, pleuropulmonary involvement must be taken into consideration as a possible complication, even shortly after the appearance of the intracranial lesion, as was the case in our patient.

References


Oriol Plans Galván, a,* Ignasi García-Olived, b Maria Sol Prats, b Elena Ferrer Sistach, c Eduard Fernández-Ofierias, c Juan Ruiz Manzano b

a Servicio de Medicina Intensiva y Crítica, Hospital Universitari Germans Trias i Pujol, Badalona, Barcelona, Spain
b Servicio de Neumología, Hospital Universitari Germans Trias i Pujol, Badalona, Barcelona, Spain
c Servicio de Cardiología, Hospital Universitari Germans Trias i Pujol, Badalona, Barcelona, Spain

* Corresponding author.
E-mail address: oriolplans@hotmail.com (O. Plans Galván).

Fig. 1. Image of the thoracic cavity during autopsy: multiple nodular neoplastic implants are seen in the visceral pleura, along with a moderate amount of serous, bloody pleural fluid.