Chronic thromboembolic pulmonary hypertension associated with indwelling Port-A-Cath® central venous access systems

Xavier Jais, Delphine Natali, Marjorie Abraham, Laurent Savale, Dermot O’Callaghan, David Montani, Azzedine Yaïci, Florence Parent, Marc Humbert, Olivier Sitbon, Gérald Simonneau

French referral centre for Pulmonary Hypertension, Université Paris-Sud, Hôpital Antoine Béclère, Clamart, France

Background:

Chronic thromboembolic pulmonary hypertension (CTEPH) is a frequent cause of pulmonary hypertension, the treatment of choice of which is pulmonary endarterectomy (PEA) in cases of surgically accessible disease. Ventriculoatrial shunts and infected cardiac pacemakers are known to predispose to the development of CTEPH that is generally inaccessible to PEA. However, there are no cases reported in the literature of CTEPH in patients with indwelling Port-A-Cath® (PAC) central venous access systems.

Methods:

We reviewed consecutive patients with PACs admitted to our department between January 2006 and March 2010 for assessment of CTEPH. We described these patients’ characteristics, clinical and surgical management and outcome.

Results:

Seventeen patients (7 F, 10 M; age 55±16 y) were identified. The interval between insertion of PAC and diagnosis of CTEPH was 5.6±4.0 years ago. In 15 patients, the PAC had been inserted for administration of chemotherapy in a context of malignant disease, which was in complete remission in 93% of cases at the time of diagnosis of CTEPH. At first assessment, all patients were in NYHA functional class III or IV, 6-minute walk distance was 242±154 m. Baseline hemodynamics were as follows: mPAP 46±11 mmHg, RAP 9±7 mmHg, CI 2.0±0.6 L/min/m², TPR 1127±549 dyn.s.cm⁻⁵. Eleven patients (67%) were considered operable candidates, only 8 patients underwent surgery. PEA was considered successful in 75% of cases. One patient died during the immediate postoperative period; one other patient developed refractory right heart failure and subsequently underwent lung transplantation. All PACs were removed and 30% were culture-positive for staphylococcus epidermidis. Six months after surgery, 50% of patients had significant clinical and hemodynamic improvement.

Conclusion:

These data suggest that PAC may predispose to the development of CTEPH. For some patients, staphylococcal infection may play a role in the development of the disorder. Patients with CTEPH associated with PACs may be more likely to have disease that is amenable to PEA than patients with CTEPH that develops in association with ventriculoatrial shunts and infected pacemakers.