Bilateral pulmonary artery aneurysm in Behçet syndrome: Report of two operative cases

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Pulmonary artery aneurysm is uncommon and may rupture causing fatal hemoptysis. We report 2 cases of bilateral pulmonary artery aneurysm for which we attempted conservative surgical treatment.

Clinical Summary

PATIENT 1. A 16-year-old boy with previously diagnosed Behcet syndrome was hospitalized because of intermittent hemoptysis that had lasted for 2 weeks. Erythrocyte sedimentation rate was 40 mm in the first hour, and C-reactive protein level was greater than 3.5 mg/dL. Chest radiography (Figure 1, A) showed rounded right parahilar opacity. Computed tomographic scan (Figure 1, B) of the chest revealed aneurysmal dilatation of both main pulmonary arteries, with thrombus within. The patient received intravenous corticosteroid therapy and was operated on 7 days after admission. At the time of operation, he had an erythrocyte sedimentation rate of 13 mm/h and a C-reactive protein level of 1.5 mg/dL. Median sternotomy was performed. The proximal right pulmonary artery was clamped under the superior vena cava. The aneurysm was opened, the thrombus was evacuated, the arterial defect was located, and endoaneurysmorrhaphy was performed. There was no leakage through the suture line after removal of the aortic clamp. The left pulmonary artery was operated on in the same operative session. The left pulmonary artery was controlled and clamped in the left pulmonary hilum; subsequent surgical steps were the same as for the right aneurysm. In the next 24 postoperative hours, while still under mechanical ventilation, the patient died suddenly of massive hemoptysis.

PATIENT 2. A 28-year-old man was admitted as an emergency after 10 days of hemoptysis. His medical history revealed oral and genital ulceration 1 year before admission indicative of Behçet syndrome. Erythrocyte sedimentation rate was 87 mm/h, and C-reactive protein level was 4.5 mg/dL. Chest radiography showed a giant, rounded opacity on the right lung field and a rounded, external opacity on the left lung field. An air-fluid level in the upper third of the right opacity indicated fissured pulmonary aneurysm (Figure 2, A). Thoracic computed tomographic scan of the chest revealed aneurysmal dilatation of both main pulmonary arteries, with thrombus within (Figure 2, B). Intravenous corticosteroid therapy (2 mg/[kg · d]) allowed the erythrocyte sedimentation rate to decrease from 87 to 32 mm/h in 3 days. The patient was immediately operated on after this delay, with the same procedure as in the previous case. He was extubated within the first postoperative hour. He was discharged 10 days after admission with a course of oral corticotherapy and cyclophosphamide therapy. No postoperative complications had developed after 3.5 years.

Discussion

Our 2 patients both had confirmed Behçet syndrome according to the criteria of the International Society for Behçet’s Disease. Pulmonary aneurysms are generally multiple, and as in our case may lead to hemoptysis by fistulizing into the bronchi. Hemoptysis of varying degrees is the most common symptom, with more than half of patients dying of pulmonary hemorrhage within 3 years.2

Behçet syndrome is the only known acquired syndrome that leads to the formation of pulmonary artery aneurysm. Such aneurysms, although small angiographically, can develop rapidly. Sudden hilar enlargement or the appearance of polylobular and round opacities on the chest radiograph can represent pulmonary artery aneurysms. Our preoperative investigations were limited to chest radiography and thoracic computed tomographic scan. Despite benefits that pulmonary artery catheterization can offer, it carries significant risk.

Various treatment modalities in pulmonary aneurysm include immunosuppressive therapy, surgery, and endovascular surgery. In case of massive hemoptysis, surgery may be necessary. The main problem facing the vascular surgeon is the 25% incidence of recurrent anastomotic aneurysm.3 The previously reported surgical technique consisted of pneumonectomy or lobectomy to the pulmonary structure involved by the aneurysm.4 The technique we used is more elective and avoids pulmonary amputation. Aneurysmectomy with direct closure of the aneurysmal defect has been reported to carry better long-term patency.5 On the basis of these data, we prefer a surgical approach with simple aneurysmectomy and occlusion of the arterial defect by direct suture (endoaneurysmorrhaphy). This technique seems to be compatible with the morphologic character of false and saccular aneurysms in Behçet syndrome. Preoperative steroid coverage has been suggested to reduce the risk of complications.1 The massive hemoptysis was
certainly a result of suture line rupture. Many factors may have played role in this hemorrhage. We emphasize the deleterious action of prolonged ventilation, which places cyclic positive pressure on the suture line. This pressure can be amplified if the patient is not in phase with the ventilator.

In patients with recurrent or massive hemoptysis, urgent surgery is required. Endoaneurysmorhaphy should be preferred to pneumonectomy or lobectomy. In the postoperative period, extubation must occur as soon as possible.

References