Endovascular Treatment of Bilateral Pulmonary Artery Stenoses and Superior Vena Cava Syndrome

in a Patient with Advanced Mediastinal Fibrosis

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Vascular stenosis is a relatively uncommon and often fatal sequela of mediastinal fibrosis. There are very few reports in the medical literature of endovascular treatment for concomitant bilateral pulmonary artery stenoses and superior vena cava syndrome. We report the endovascular treatment of these conditions in a 54-year-old man, and the long-term outcome. (Tex Heart Inst J 2016;43(3):249-51)

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Mediastinal fibrosis (MF) is a rare condition that results in fibrous tissue proliferation of variable severity and is typically attributed to an exaggerated immune response to infection by *Histoplasma capsulatum*.¹ In severe cases, it can progress to obstruction of the superior vena cava (SVC), pulmonary vessels, esophagus, or airways. Whereas SVC obstruction is relatively common, pulmonary artery (PA) involvement is less frequently encountered.² Very few reports of long-term outcomes exist in the medical literature of either SVC or PA stents in this patient population. We discuss a case in which MF resulted in PA and SVC stenoses that were treated endovascularly, with early symptom improvement and 5-year survival with primary patency.

Case Report

A 54-year-old man with histoplasmosis presented at our institution with worsening right-sided chest tightness and difficulty sleeping on his right side because of shortness of breath. Three years before this episode, he had developed SVC syndrome from progressive MF that we had successfully treated endovascularly with an SVC stent. Therefore, differential diagnosis for the current episode of dyspnea included restenosis of the SVC stent, pulmonary embolism, pneumonia, and pulmonary hypertension. A computed tomographic angiogram (CTA) revealed high-grade right-PA stenosis caused by continued progression of MF. In addition, imaging revealed near-complete occlusion of the truncus anterior to the right PA. Results of CTA also revealed that the SVC stent remained widely patent.

Subsequently, we performed pulmonary arteriography to evaluate right-PA stenosis and PA hypertension, and to judge the possibility of intervention. Using a femoral approach, we advanced a Mott pulmonary catheter over the wire and into the right ventricle. The catheter was next advanced into the main PA, where the pressure was 35/12 mmHg, and then into the left PA—where digital subtraction arteriography revealed no abnormalities. The left main PA pressure was 36/10 mmHg. The catheter was then modified to make it more amenable to entering the stenosed right main PA. We used a 260-cm, modified stiff GLIDEWIRE® (Terumo Medical Corporation; Somerset, NJ), advancing the wire and catheter into the right main PA. Digital subtraction arteriography revealed approximately 75% stenosis at the origin of the right main PA, extending distally 3.6 cm (Fig. 1). A pressure recording obtained distal to the obstruction was 11/5 mmHg (pressure gradient, 22 mmHg). Given the patient's symptoms, imaging findings, and pressure gradient, intervention was judged necessary.

A 10 × 37-mm Express™ LD balloon-expandable stent (Boston Scientific Corporation; Natick, Mass) was advanced through the sheath, positioned, and deployed across

the stenosis. Thereafter, a digital subtraction arteriogram of the right PA showed substantial improvement of flow in the right main PA (Fig. 2). An H1 catheter was placed over the wire through the long sheath distal to the stent. The right-PA pressure after stenting now was 24/16 mmHg (improved from 11/5 mmHg). After the catheter, wire, and sheath were withdrawn, pressures in the main PA were 33/10 mmHg. The gradient of 9 mmHg had improved from 22 mmHg and was within normal limits. Attention was then turned to the evaluation of the SVC stent. A digital subtraction arteriogram of the SVC showed widely patent flow through the SVC stent.

Fig. 1 Pulmonary arteriogram reveals proximal right pulmonary stenosis. Note the previously placed endovascular superior vena cava stent, which has only a mild contour deformity.

Fig. 2 After placement of a balloon-expandable endovascular stent, this pulmonary arteriogram shows improved flow and a patent proximal right pulmonary artery.

riogram of the SVC showed widely patent flow through the SVC stent.

After the procedure, the patient reported relief of both his chest discomfort and his shortness of breath. A follow-up CTA at the 1-year interval showed both stents to be patent (Fig. 3). Our patient experienced immediate normalization of physiologic values and symptomatic relief after right-PA stenting.

Over the 2 years that followed, however, his MF continued to progress to the extent that a digital subtraction arteriogram at the end of that time showed approximately 75% stenosis at the origin of the left PA. An 8 × 27-mm Express™ LD balloon-expandable stent (Boston Scientific) was placed at another institution. Approximately 1 month later, the patient developed hemoptysis and underwent emergency diagnostic angiography at our institution (Fig. 4). Over the next 2 months, the patient had recurrent episodes of hemoptysis and friable bronchial mucosa. After a protracted medical course, he died of respiratory complications—5 years from his initial endovascular intervention.

Discussion

Mediastinal fibrosis is a non-neoplastic condition characterized by the proliferation of fibrous tissue in the mediastinum.¹ Most patients are young, and males and females are similarly affected.² The cause is varied but frequently relates to fungal infections such as *H. capsulatum*, or to tuberculosis. It presents with symptoms of cough, shortness of breath, and even hemoptysis, and it carries a high mortality rate (up to 30%).³⁻⁵ The literature has referred to the condition as fibrotic mediastinitis or sclerosing mediastinitis.⁵ Two forms of the disease exist—namely, the focal and the diffuse. On images, the diffuse form tends not to show calcifications. Because of differences in origin, presentation, and

Fig. 3 Follow-up computed tomographic angiogram, 1 year after right pulmonary artery stenting and 3 years after superior vena cava stenting, shows patency of both stents.

Fig. 4 Diagnostic pulmonary angiogram at our institution shows patency of all 3 stents.

sequelae, the 2 entities vary in prognosis and treatment. It is often thought that the focal form is triggered by *H. capsulatum*, whereas the diffuse form—which is less prevalent—is usually idiopathic or inflammatory. Treatment varies from medical therapy (the systemic administration of antifungal agents or corticosteroids) to surgical or endovascular treatment.^{6,7} Although stenting provides an important alternative to surgery, there are potential sequelae. These include stent migration, dissection, thrombosis, intimal hyperplasia, pseudoaneurysm formation, vascular rupture, and death. Long-term complication rates of 9.1% have been reported,⁸ but most of the available medical literature derives from experience with pulmonary stents in pediatric patients with congenital heart disease.

Some authors⁵ postulate that only genetically susceptible individuals will develop this pathologic fibroinflammatory response to the above (and various other) antigens. These patients will develop symptoms when the fibrosis compromises a major vessel or the airway.⁹ Because of its anatomic course through the mediastinum, the right PA is more often affected than the left PA. However, SVC involvement is still more common.^{10,11}

In this case of MF, we were able to improve the quality of life of our patient and prolong his survival after di-

agnosing and treating 2 high-grade stenoses—without the need for open surgery. Improvements in stent technology and in our understanding of the disease process are enabling us to lengthen survival and improve quality of life for patients with advanced MF.

References

1. Gustafson MR, Moulton MJ. Fibrosing mediastinitis with severe bilateral pulmonary artery narrowing: RV-RPA bypass with a homograft conduit. *Tex Heart Inst J* 2012;39(3):412-5.
2. Novella Sanchez L, Sanz Herrero F, Berraondo Fraile J, Fernandez Fabrellas E. Mediastinal fibrosis and superior vena cava syndrome [in Spanish]. *Arch Bronconeumol* 2013;49(8):340-2.
3. Murphy JC, Johnston N, Spence MS. A pressing matter—mediastinal fibrosis with near obliteration of the pulmonary arteries. *Catheter Cardiovasc Interv* 2013;81(6):1079-83.
4. Thiessen R, Matzinger FR, Seely J, Aina R, Macleod P. Fibrosing mediastinitis: successful stenting of the pulmonary artery. *Can Respir J* 2008;15(1):41-4.
5. Rossi SE, McAdams HP, Rosado-de-Christenson ML, Franks TJ, Galvin JR. Fibrosing mediastinitis. *Radiographics* 2001;21(3):737-57.
6. Smith JS, Kadiev S, Diaz P, Cheatham J. Pulmonary artery stenosis secondary to fibrosing mediastinitis: management with cutting balloon angioplasty and endovascular stenting. *Vasc Endovascular Surg* 2011;45(2):170-3.
7. Guerrero A, Hoffer EK, Hudson L, Schuler P, Karmy-Jones R. Treatment of pulmonary artery compression due to fibrous mediastinitis with endovascular stent placement. *Chest* 2001;119(3):966-8.
8. Law MA, Shamszad A, Nugent AW, Justino H, Breinholt JP, Mullins CE, Ing FF. Pulmonary artery stents: long-term follow-up. *Catheter Cardiovasc Interv* 2010;75(5):757-64.
9. Thomas BP, Bream PR Jr, Milstone AP, Meranze SG. Treatment of SVC syndrome and hemoptysis in a patient with mediastinal fibrosis. *Emerg Radiol* 2006;12(5):240-3.
10. Sherrick AD, Brown LR, Harms GF, Myers JL. The radiographic findings of fibrosing mediastinitis. *Chest* 1994;106(2):484-9.
11. Rizvi AZ, Kalra M, Bjarnason H, Bower TC, Schleck C, Gloviczki P. Benign superior vena cava syndrome: stenting is now the first line of treatment. *J Vasc Surg* 2008;47(2):372-80.