IJC Heart & Vasculature 8 (2015) 103–107

Contents lists available at ScienceDirect

IJC Heart & Vasculature

journal homepage: http://www.journals.elsevier.com/ijc-heart-and-vasculature

Catheter-based intervention for pulmonary vein stenosis due to fibrosing mediastinitis: The Mayo Clinic experience

Shiva P. Ponamgi [a], Christopher V. DeSimone [b], Charles J. Lenz [c], Megan Coylewright [d], Samuel J. Asirvatham [b,e,*], David R. Holmes [b], Douglas L. Packer [b]

[a] Division of Hospital Internal Medicine, Mayo Clinic Health System, Austin, MN, United States
[b] Division of Cardiovascular Diseases, Department of Internal Medicine, Mayo Clinic, Rochester, MN, United States
[c] Department of Internal Medicine, Mayo Clinic, Rochester, MN, United States
[d] Section of Cardiology, Department of Internal Medicine, Dartmouth-Hitchcock Medical Center, Lebanon, NH, United States
[e] Department of Pediatrics and Adolescent Medicine, Mayo Clinic, Rochester, MN, United States

ARTICLE INFO

Article history:
Received 30 April 2015
Received in revised form 15 June 2015
Accepted 16 June 2015
Available online 18 June 2015

Keywords:
Fibrosing mediastinitis
Pulmonary vein
Stenosis
Stent
Angioplasty

ABSTRACT

Introduction: Fibrosing mediastinitis (FM) is a rare but fatal disease characterized by an excessive fibrotic reaction in the mediastinum, which can lead to life-threatening stenosis of the pulmonary veins (PV). Catheter-based intervention is currently the only viable option for therapy. However, the current literature on how best to manage these difficult cases, especially in regards to sequential interventions and their potential complications is very limited.

Methods: We searched through a database of all patients who have undergone PV interventions at the Earl H. Wood Cardiac Catheterization Laboratory in Mayo Clinic, Rochester. From this collection, we selected patients that underwent PV intervention to relieve stenosis secondary to FM.

Results: Eight patients were identified, with a mean age of 41 years (24–59 years). Five were men, and three were women. Three patients underwent balloon angioplasty alone, and five patients had stents placed. The majority of patients had acute hemodynamic and symptomatic improvement. More than one intervention was required in five patients, four patients had at least one episode of restenosis, and four patients died within four weeks of their first PV intervention.

Conclusions: We describe the largest reported case series of catheter-based intervention for PV stenosis in FM. Although catheter-based therapy improved hemodynamics, short-term vascular patency, and patient symptoms, the rate of life-threatening complications, restenosis, and mortality associated with these interventions was found to be high. Despite these associated risks, catheter-based intervention is the only palliative option available to improve quality of life in severely symptomatic patients with PV stenosis and FM. Patients with PV stenosis and FM (especially those with bilateral disease) have an overall poor prognosis in spite of undergoing these interventions due to the progressive and recalcitrant nature of the disease. This underscores the need for further innovative approaches to manage this disease.

1. Introduction

Fibrosing mediastinitis (FM), also known as collagenosis or sclerosing mediastinitis [1,2] is a rare but fatal disease with relatively few therapeutic options. It is thought to occur due to exposure to the *Histoplasma capsulatum* antigen within the mediastinum, triggering an intense inflammatory host response resulting in proliferation and invasion of fibrous tissue into vital structures [3,4]. One complication of the fibrotic reaction is pulmonary vein (PV) stenosis, previously described in case reports and series [1,3–12]. Diagnosis of PV stenosis is challenging due to its gradual onset of nonspecific symptoms including fatigue and dyspnea, and thus presentation is typically delayed. In the late stages of this disease, recurrent episodes of pulmonary edema and hemoptysis can occur, eventually becoming fatal [6,7,13].

Medical and surgical therapies for FM remain largely ineffective due to its extensive fibrotic invasion of mediastinal structures [2,14–18]. For those patients suffering from PV stenosis, angioplasty and stent deployment are potential treatment options. Relieving obstruction with timely angioplasty in PV stenosis caused by other etiologies (i.e. pulmonary vein isolation for atrial fibrillation) normalizes venous flow into the left atrium [19,20]; however, there is limited data on how best to manage PV stenosis caused by FM [1,5–7,21].

* Corresponding author at: Division of Cardiovascular Diseases, 200 First Street SW, Rochester, MN 55905, United States.
E-mail address: asirvatham.samuel@mayo.edu (S.J. Asirvatham).

http://dx.doi.org/10.1016/j.ijcha.2015.06.005

We report the largest case series of catheter-based interventions for patients with PV stenosis due to FM.

2. Methods

We searched amongst all patients who had undergone pulmonary vein intervention at the Earl H. Wood Cardiac Catheterization Laboratory in Mayo Clinic Rochester. From this cohort, we identified eight patients who had at least one pulmonary vein procedure (angioplasty +/− stent placement) for PV stenosis due to FM. We then performed a detailed chart review to evaluate patient presentation, symptoms, interventions, complications, and outcomes.

3. Results

We identified eight cases of PV stenosis due to FM that underwent catheter-based intervention. There were five men and three women, with a mean age at presentation of 41 years (range 24–59 years).

In our series of eight patients, a total of eighteen catheterizations were performed, including one aborted procedure and seventeen angioplasties either with balloon, stent, or both (three patients had balloon angioplasty only). Seven patients had severe stenosis in two or more PVs (Table 1). Five of our patients required more than one intervention. Acute hemodynamic improvement was seen in all eight patients with balloon angioplasty +/− stent placement (Table 2).

3.1. Case 1

A 27-year-old woman with six months of progressively worsening dyspnea developed respiratory failure during diagnostic bronchoscopy. CT angiogram (CTA) of the chest showed bilateral patchy ground glass opacities suggestive of pulmonary edema with occlusion of both the right lower pulmonary vein (RLPV) and left upper pulmonary vein (LUPV) and a marked focal narrowing of the right upper pulmonary vein (RUPV). Successful balloon angioplasty of RUPV was done but the patient continued to clinically deteriorate and died in two days. Autopsy revealed an 8 cm fibrosing mediastinal mass with fungal stains consistent with Histoplasma, as well as partial encasement of the right and left pulmonary hilar regions with constriction or obliteration of the vasculature.

3.2. Case 2

A 52-year-old man with a history of FM and a prior pericardial patch enlargement of the RLPV 12 years ago presented with gradual worsening of fatigue and dyspnea on exertion. CTA demonstrated stenosis of the RLPV and occlusion of all other pulmonary veins. No intervention was performed initially due to lack of significant gradient across this lesion. The patient's symptoms worsened over the next five months and required two balloon angioplasties in the RLPV. The patient was asymptomatic afterwards but passed away the following month from complications of a newly diagnosed metastatic cancer.

Table 1
Patient characteristics.

Case	Age*	Sex	Pulmonary veins involved				
			RUPV	RMPV	RLPV	LUPV	LLPV
1	27	F	Stenosed	Absent	Occluded	Occluded	Patent
2	52	M	Occluded	Absent	Stenosed	Occluded	Occluded
3	47	F	Occluded	Absent	Stenosed	Occluded	Stenosed
4	38	M	Occluded	Absent	Occluded	Stenosed	Stenosed
5	59	M	Stenosed	Absent	Stenosed	Occluded	Patent
6	42	M	Stenosed	Occluded	Patent	Absent	Absent
7	24	F	Stenosed	Absent	Occluded	Patent	Patent
8	43	M	Occluded	Absent	Stenosed	Stenosed	Stenosed

* Age at first catheterization; left upper pulmonary vein; LLPV, left lower pulmonary vein; PV, pulmonary vein; RLPV, right lower pulmonary vein; RUPV, right upper pulmonary vein.

3.3. Case 3

A 47-year-old man with FM was admitted for worsening shortness of breath over the past three months with pulmonary edema. CTA showed severe stenosis of the left lower pulmonary vein (LLPV) and the right middle pulmonary vein (RMPV) along with occlusion of LUPV and RUPV. The patient underwent balloon angioplasty followed by placement of two stents in the LLPV and RMPV respectively. This resulted in a significant improvement in hemodynamics. Repeat CTA was performed at 1 month, 1 year, 1.5 years, and 4 years post procedure and showed no significant restenosis; the patient has remained asymptomatic.

3.4. Case 4

A 38-year-old woman with FM presented with six months of worsening shortness of breath and hypoxia requiring 4 L of oxygen at rest. CTA (Fig. 1) demonstrated that the right pulmonary veins were occluded, and the left pulmonary veins were stenotic but patent. Balloon angioplasty was performed in the LUPV resulting in improved symptoms. Nine months later, symptoms recurred requiring a repeat balloon angioplasty of the LLPV. At three-month follow-up, the patient was still requiring 4 L of oxygen at rest and remained dyspneic with exertion, but symptoms had improved while at rest.

3.5. Case 5

A 59-year-old male with FM presented with gradually worsening hemoptysis and exercise intolerance for the last two years. Angiography revealed stenosis of the RLPV and RUPV and occlusion of the LUPV (Fig. 2). A stent was successfully placed in the RLPV but the patient developed severe chest pain during the procedure. An occlusive thrombus was found in the left anterior descending artery, which was removed, and the patient was started on eptifibatide and bivalirudin infusions. A drug eluting stent was then placed in the RUPV, but the patient developed severe intra-procedural hemoptysis. At this time, all anticoagulation was stopped and an endobronchial blocker was placed in the left main stem bronchus. Overnight, the patient suffered a large embolic stroke with temporal lobe herniation and died.

3.6. Case 6

A 42-year-old male with FM and history of a left pneumonectomy presented with worsening hemoptysis and exercise intolerance over the last six months. CTA demonstrated high-grade stenosis of RUPV and chronic RMPV occlusion. Two stents were initially placed in the RUPV and in the next ten months, two more balloon angioplasties were performed for restenosis; despite this, the RUPV re-occluded (Fig. 3). Rotational atherectomy was then performed within the stent followed by cutting balloon angioplasty, but it resulted an acute in-stent thrombosis the next day. In the next eight months, the patient required three more angioplasties with cutting balloons as well as another stent in the RUPV. The patient is now being considered for a heart and lung transplant.

3.7. Case 7

A 24-year-old woman with FM and von Willebrand disease presented with worsening dyspnea on exertion over the course of the last month. CTA revealed complete occlusion of the RLPV and near complete occlusion of the RUPV. A stent was successfully placed in the RUPV but it re-stenosed in six months with resultant intermittent hemoptysis requiring another stent. The second stent lasted for thirteen months