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Case Report

Extreme recurrent massive hemoptysis in a cystic fibrosis patient requiring 22 separate embolization procedures prior to lung transplantation: A case report

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ABSTRACT

While bronchial artery embolization is an established, safe, and effective treatment for massive hemoptysis from a variety of causes including cystic fibrosis, patients rarely require more than 2 angiography and embolization treatments during their lifetime. We present a rare case of massive, recurrent hemoptysis requiring a total of 22 angiography and embolization procedures over a period of 8 years, prior to the patient receiving a double lung transplant.

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Introduction

Bronchial artery embolization (BAE) is an established, effective treatment for massive hemoptysis in cystic fibrosis (CF) patients [1,2]. In the western hemisphere, CF is a leading cause of massive hemoptysis, defined as acute bleeding with a volume greater than 240 mL within 24 hours [3]. It has been estimated that as many as 4.1% of CF patients will experience acute massive hemoptysis in their lifetime with an average annual incidence of 0.87% [4]. In CF, chronic inflammation in the lungs promotes hypertrophy of the bronchial arteries and

angiogenesis. Having undergone these deleterious changes, the systemic arterial supply to the lungs is the source of major hemoptysis in CF patients and can be life-threatening, carrying a substantial risk of asphyxiation or exsanguination [1].

BAE has been shown to be highly effective in achieving initial control of hemoptysis, with reported success rates ranging from 73% to 100% after one session [5]. However, recurrence of hemoptysis can be observed in CF patients having undergone an initial successful BAE, with rates of recurrence reported as high as 55% [5]. Sources of recurrent bleeding are highly variable, due not only to the high variability of bronchial artery

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anatomy but also to the formation of collateral vessels [2]. One study reported a 75% incidence of bleeding from nonbronchial systemic collateral vessels in patients with recurrent hemoptysis after a previous BAE [1]. Transarterial embolization of the bronchial arteries, as well as other systemic arterial collateral vessels, remains a safe and effective means of managing recurrent hemoptysis. BAE complications are few and studies suggest that it is unlikely to produce adverse effects on pulmonary function [6].

Despite the high recurrence of hemoptysis in CF patients, only a small percentage of patients require more than two embolization procedures [6,7]. To the best of our knowledge, the highest reported number of embolization procedures in an individual patient was 7, over the course of approximately 70 months [1,6]. We describe a patient who underwent a total of 22 angiography and transarterial embolization procedures over the course of 96 months prior to definitive surgical therapy in the form of lung transplantation.

Case presentation

At the authors' institution, IRB approval is not required for case reports. The patient was first diagnosed with CF in 2000, at age of 23. She had her first angiogram with BAE for massive hemoptysis in 2007. Over the next 8 years, she needed 21 additional angiography and embolization procedures for recurrent hemoptysis, with more than half of these procedures occurring over the 18 months preceding her transplant. Each of these procedures was performed for massive hemoptysis of at least 200mL over a 24-hour period. She also had pulmonary angiography to evaluate for any vascular abnormality in the pulmonary artery branches that could cause hemoptysis. Her pulmonary angiography was normal, thus no pulmonary artery branch vessel embolization was performed. These procedures were performed by 5 different interventional radiologists at 3 separate institutions, but the majority of her procedures were performed at the authors' institution. Her last embolization was performed in June 2015 and then she received a double lung transplant in August 2015. At the time of this submission, the patient is now 3.5 years postlung transplant with no evidence of rejection and excellent lung function.

Over the course of her 22 embolization procedures, she had a variety of vessels treated, including the bronchial arteries, internal mammary arteries, intercostal arteries, costocervical trunk branches, thyrocervical trunk branches, multiple unnamed parasitized vessels off of the subclavian arteries, and a parasitized vessel arising from an esophageal branch of the left gastric artery (Fig. 1). All procedures were performed via a right groin approach under moderate or deep intravenous sedation, and embolization was performed with particles of varying size, but most typically in the range of 100-300 microns, depending on the type of particle chosen by the interventional radiologist. Each of these procedures was technically successful, with embolization of one or more vessels supplying abnormal lung parenchyma. Due to profound collateralization and parasitization of arteries feeding this patient's lungs, she had more than 100 arterial branch

vessels embolized during this time period. The patient's massive hemoptysis resolved after each procedure, although the durability of the response became shorter over the 8 years during which the procedures were performed. Twelve of the 22 embolizations were performed over the last 2 years before transplant.

Discussion

BAE is an effective short-term intervention in the management of massive hemoptysis and also can improve long-term outcomes in CF patients [7]. Despite its utility in the context of recurrent hemoptysis, fewer than 15%-20% of patients require more than 2 sessions [6,7]. We report a patient who required 22 angiography and transarterial embolization procedures over a period of 8 years to treat recurrent massive hemoptysis related to underlying CF. Despite the large number of hemoptysis episodes, she was not deemed a candidate for lung transplant until less than 1 year before she received her surgery, due to relatively preserved lung function on her pulmonary function tests. However, she suffered from relatively severe, recurrent acute on chronic respiratory infections, which in addition to the chronic inflammatory changes seen in CF, were thought to be the underlying reason for the patient needing so many embolizations. This led to a particularly large number of embolizations needing to be performed over an 8 year period, making this an extreme case from both the perspective of the sheer number of procedures needed as well as the number and variety of vessels that required treatment.

Prior studies have reported the utility of multiple embolizations for some CF patients, including treatment of vessels in the thorax that can be parasitized and feed abnormal lung parenchyma after prior BAE had been performed [1,2,6–8]. The two most common reasons for repeat BAE for hemoptysis were recanalization (72%) and neoangiogenesis (19.1%) based on analysis of angiography [8]. Woo found a higher hemoptysis-free survival when using N-butyl cyanoacrylate instead of polyvinyl alcohol particles [9]. However, to our knowledge, the mainstay of transarterial embolization for CF patients with hemoptysis is particle embolization.

This case demonstrates the importance of thorough angiography, superselective microcatheter positioning for safe embolization, and recognition that some CF patients require a uniquely high number of embolizations to prevent asphyxiation or exsanguination prior to receiving a transplant. While it is atypical for CF patients to require such a large number of embolization procedures in their lifetime, interventional radiologists and pulmonologists must be aware that recurrent hemoptysis can occur. This is primarily due to the marked parasitization and collateralization that may develop because of the underlying lung parenchymal inflammatory disease which is often complicated by chronic lung infections.

While computed tomography angiography (CTA) or bronchoscopy was performed before every procedure, CTA was obtained prior to 3 procedures and bronchoscopy was performed prior to 2 procedures. A consensus has not been reached

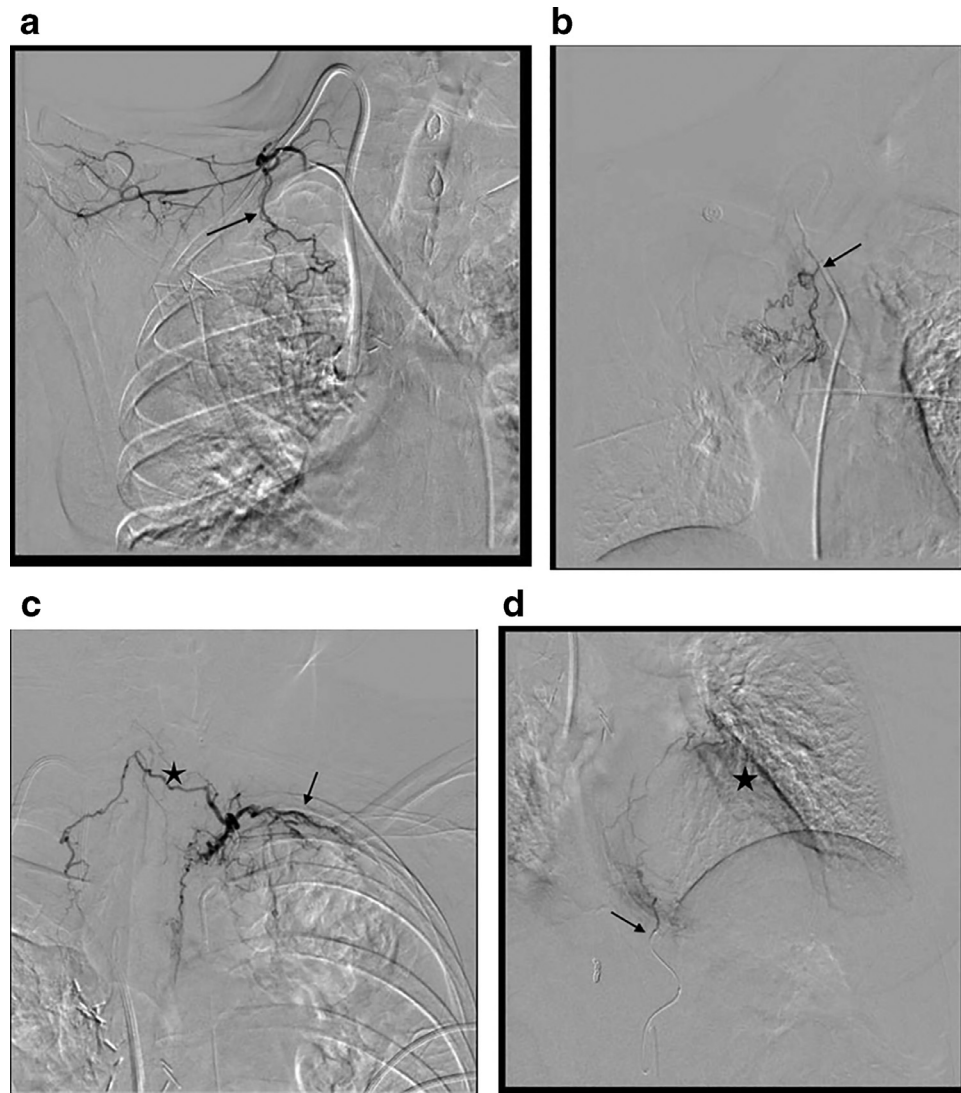


Fig. 1 – Angiographic images demonstrating superselective microcatheter embolization of arteries other than the bronchial arteries (which had previously been embolized) to treat a cystic fibrosis patient with recurrent massive hemoptysis until her disease progressed enough to be selected for lung transplantation. Microcatheter digital subtraction angiographic (DSA) image demonstrating parasitized arterial vasculature feeding cystic fibrotic right upper lobe abnormality which arose off of the right suprascapular artery (arrow), a branch of the thyrocervical trunk (A). Superselective DSA image via microcatheter in parasitized branch vessel arising off of the superior intercostal artery (arrow), a branch of the right costocervical trunk (B). DSA image via a microcatheter in the left costocervical trunk, demonstrating parasitized vasculature supplying abnormal lung in the left upper lobe (arrow) and another branch crossing midline (star) to supply abnormal right upper lobe disease (C). DSA image via microcatheter (arrow) in a parasitized branch off of an esophageal branch of the left gastric artery that traversed the diaphragm and had branches supplying abnormal cystic fibrotic lung (star) in the region of the lingula (D). Each vessel was superselectively catheterized with a microcatheter and embolized with particles, without complication.

regarding the recommendation to perform preprocedural studies for localization in patients with CF [3]. However, it has been suggested that CTA may be useful to study the anatomy of CF patients where unrecognized collateral vessels are associated with an increased need for repeated interventions [1,5].

In conclusion, we describe a case of a CF patient requiring a total of 22 angiography and embolization procedures over a period of 8 years to treat massive, recurrent hemoptysis until her overall disease progressed to the point of being appropri-

ate for and ultimately receiving a lung transplant. Interventional radiology plays a critical role in the management and survival of CF patients in this setting.

Supplementary material

Supplementary material associated with this article can be found, in the online version, at doi:[10.1016/j.radcr.2019.01.020](https://doi.org/10.1016/j.radcr.2019.01.020).

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