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CASE REPORT

N-Butyl cyanoacrylate embolization of an extremely rare variant of sequestration complex — a high-flow left-to-left shunt between systemic artery and pulmonary vein

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Abstract
A 19-year-old female presented with hemoptysis. Computed tomography (CT) pulmonary angiography revealed aberrant vessels from descending thoracic aorta, draining into pulmonary veins (left-to-left shunt). She was managed by transcatheter embolization of the aberrant vessels using N-butyl cyanoacrylate (NBCA) with balloon occlusion. A systemic artery to pulmonary vein fistula is one of the least common congenital anomalies. Most of the reported cases have been managed by surgery. Only a few patients have been treated by transcatheter embolization, using coils or vascular plugs as the embolizing agents. To our knowledge, this is the first case of its kind that was managed by glue embolization. Favorable post-procedure results have led us to believe that glue embolization can be considered a suitable alternative to thoracotomy in such patients.

Keywords Butyl 2-cyanacrylate · Therapeutic embolization · Bronchopulmonary sequestration

Introduction

A systemic artery to pulmonary vein fistula is one of the least common congenital anomalies and literature on this is scarce. Most of the reported cases have been managed by surgery and rarely by endovascular treatment (vascular plugs/coils). There has been no case reported in literature to be treated by a liquid embolic agent. In this rare and unique case, we chose endovascular management as the preferred treatment strategy and used glue as the embolizing agent, as coil embolization could not result in complete embolization.

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Case report

A 19-year-old female presented with complaints of recurrent hemoptysis (30 cc/day) for 1 year. The patient had no history of trauma, infection, or surgery. Routine investigations were within normal limits except for moderate anemia (hemoglobin 9.5gm%).

The patient underwent a computed tomography (CT) pulmonary angiography, which revealed three systemic artery to pulmonary venous fistulas arising from the descending thoracic aorta (D5, D6, and D8 vertebral levels) entering the right lower lobe with venous drainage into the right superior and inferior pulmonary veins (left-to-left shunt). These arteries were neither bronchial arteries nor were associated with any pulmonary sequestration. The main right and left pulmonary arteries were normal.

The treatment options including surgery and endovascular embolization were discussed with the patient and she decided to go for transcatheter embolization. A transfemoral approach was used with a 5-Fr sheath. Selective cannulation of the aberrant vessel at D6 vertebral level was performed using 5F Cobra catheter. The angiogram revealed a high-flow systemic to pulmonary venous fistula with parenchymal blush and drainage into the left atrium (Fig. 1A and B). A plan for distal coil embolization was made as there was a risk of recurrence with proximal coil embolization.

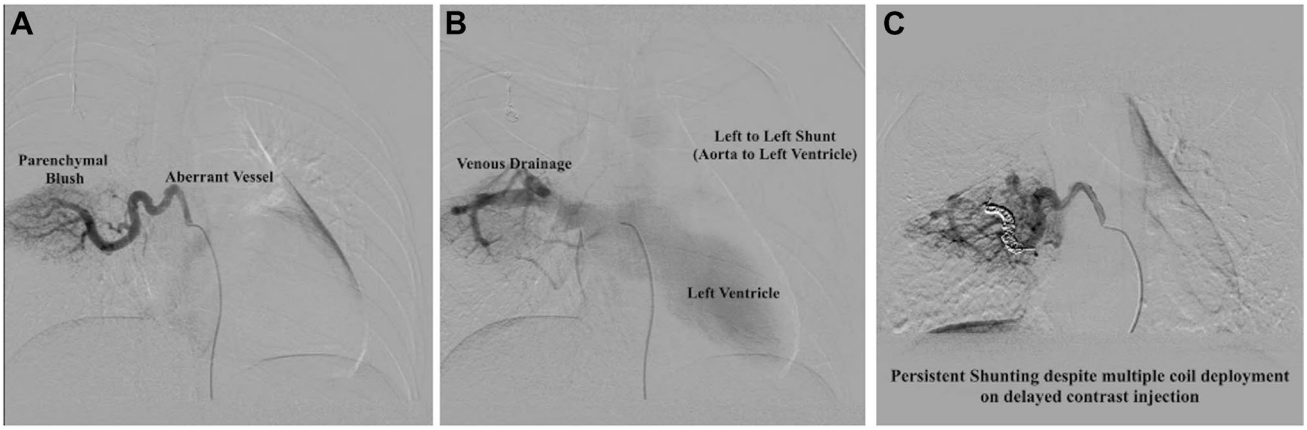

Fig. 1 Left-to-left shunt with failed coil embolization. **A** Angiogram (late arterial phase) after selective cannulation of D6 aberrant artery showing lung parenchymal blush. **B** Angiogram (venous phase) showing venous drainage into pulmonary vein and left atrium. **C** Persistent shunting despite multiple coil deployment on delayed contrast injection

A microcatheter (Direxion™ Boston Scientific) was used to super-selectively cannulate the aberrant vessel and embolization was performed by deploying four 18–14-10 and three 18–14-6 coils (Cook Inc). An angiogram just proximal to the coils demonstrated good embolization. However, on a more proximal angiogram (near the site of origin of the aberrant vessel), contrast opacification of the aberrant vessel with parenchymal blush similar to the initial angiogram was seen (Fig. 1C).

More dense packing with coils would increase the overall cost of the procedure. In addition, there was no guarantee that the goal of embolization could be achieved using coils. Amplatzer vascular plugs were not available at that time. Polyvinyl alcohol (PVA) particles would have been a great option if the fistula had been low-flow. Given the high-flow nature of the fistula with drainage into the left atrium, the use of PVA particles would have been similar to a daredevil stunt due to risk of non-target embolization.

Finally, we thought of a plan to convert this high-flow fistula to a low-flow fistula so that embolization by glue can be done safely. The catheters and sheath were exchanged for a 7Fr guiding renal double curve (RDC) catheter. After selectively cannulating the aberrant vessel (D6 level), we used an assembly of microcatheter (Direxion™ Boston Scientific) in distal position and 5-mm × 20-mm monorail balloon catheter (Abbott Viatrac 14-Plus) in proximal position. The balloon was inflated to occlude the inflow to the vessel. A contrast injection was administered to look for any change in the flow of the systemic artery to the pulmonary venous fistula, and to our expectation, there was a significant decrease in the flow to the aberrant vessel, which was appreciated as a significant delay in the opacification of the pulmonary vein and left atrium (Fig. 2A). With this new insight, the decision to use glue in combination with lipiodol (1:1) as the embolizing agent was made (Fig. 2B). An angiogram performed after the glue embolization revealed complete occlusion of the aberrant vessel (Fig. 2C). Since there was complete embolization, the use of additional coils for occluding the remaining stump of the artery was not required. Using the same technique (glue embolization after balloon inflation), the other two aberrant vessels (D5 and D8) were also embolized successfully. No coils were used for embolization of the other two aberrant vessels (Supplementary video showing the main steps of the procedure is attached).

Post-procedure, the patient was stable and maintained normal oxygen saturation on room air. Over the next 3 days, the patient complained of mild right-sided chest pain, which was managed using analgesics. She was discharged after 3 days in a stable condition. Follow-up at 1, 6, and 12 months revealed no episodes of hemoptysis.

Discussion

The systemic arterial supply to the lung can be congenital or acquired. The congenital causes provide an aberrant systemic artery supply to the lung. These include bronchopulmonary sequestration and congenital pulmonary veno-lobar syndrome [1]. In the acquired causes (bronchiectasis, pulmonary infections, and pulmonary thromboembolism), there is hypertrophy of the normal systemic arteries.

The sequestration complex can be categorized based on the anatomy of the tracheobronchial tree, pulmonary artery, and lung parenchyma. True pulmonary sequestration is defined as non-functioning lung tissue that is not in normal continuity with the tracheobronchial tree and pulmonary arteries and derives its blood supply from the systemic arteries [2]. Pseudo-sequestration is the lung tissue that has a normal tracheobronchial tree but an absent pulmonary arterial supply, with blood supply from the systemic vessels [3].

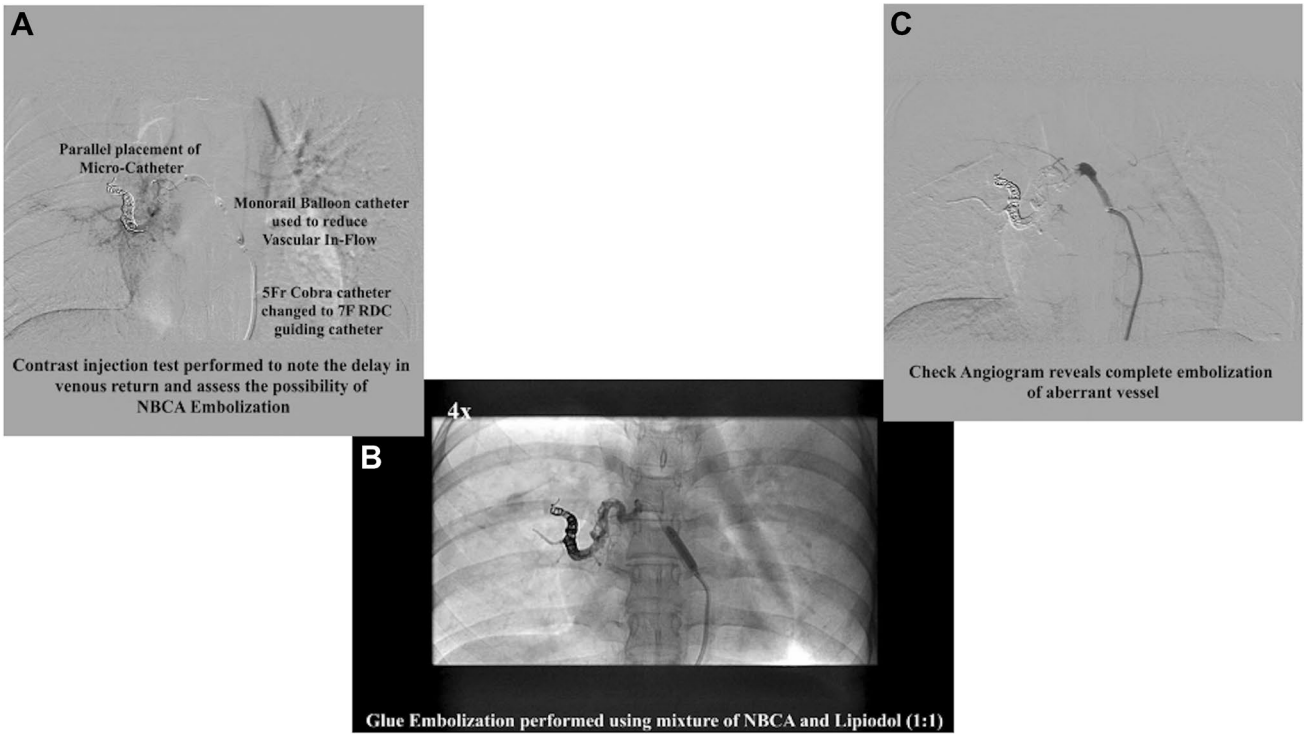

Fig. 2 Glue embolization of the aberrant artery. **A** Assembly of microcatheter in distal position and monorail balloon catheter in proximal position with inflated balloon and contrast injection showing significant decrease in the blood flow. **B** Glue embolization performed using mixture of NBCA and lipiodol (1:1). **C** Post-embolization angiogram revealing complete embolization of the aberrant vessel

The third type is a systemic to pulmonary vein fistula (non-sequestration), which is supplied by an aberrant systemic vessel with no parenchymal or bronchial abnormalities and presence of a normal pulmonary arterial supply [4].

A systemic artery to pulmonary vein fistula is one of the least common congenital anomalies. It was first described as a type I sequestration complex [5]. Some authors have also described this condition as systemic arterialization of the lung without sequestration [2, 4]. It is a rare condition that can be seen in patients of all age groups. Most of the patients are from the pediatric population. The most common presentations are a continuous murmur, tachypnoea, cyanosis, and failure to thrive [2–4]. However, the patients in the adult age group are mostly asymptomatic or they may present with exertional dyspnoea and hemoptysis [2–4]. On imaging, there is an aberrant systemic artery shunting to the pulmonary vein, finally draining into the left heart, creating left-to-left shunt. The lower lung lobes are invariably involved, more frequently on the left side [2, 6–8].

In literature, most of these cases have been managed by surgical ligation and lobectomy and only a few have been treated by endovascular embolization [2, 4–10]. In addition, it is important to realize that embolization in these cases has its own inherent problems as most of these cases are from the pediatric population. These include limit on the use of contrast, use of smaller hardware, higher risk of bleeding, and other factors related to pediatric anesthesia and post-procedure care.

In all of these described cases in the literature, which have been managed by endovascular embolization, embolization was performed using an Amplatzer vascular plug or metallic coils and not using glue. The use of Amplatzer® vascular plug for the occlusion of vascular malformations has a very high technical success rate. Its only disadvantage is the requirement of relatively large delivery systems and catheters (at least 5 Fr), which can be problematic in small children. Controlled-release coils or detachable coils also have an equally good technical success rate but have an added advantage over the vascular plugs as they can be implanted via 4-Fr catheters. So, in most of the pediatric cases, the authors have used coils for closure of smaller fistulas and vascular plugs for larger fistulas. However, in our case, this limitation of sheath size is not applicable as the patient was 19 years old and there was no problem with the use of larger delivery system.

The use of liquid embolic agents like glue does carry a risk of distal embolization to the right atrium and systemic circulation. However, if the arterial flow is reduced like in our case, and the injection of glue is performed in a controlled manner by an experienced operator, the risk is significantly

reduced. In our case, we had to use glue as coil embolization (7 coils in 1 aberrant vessel as described above) was unsuccessful and we did not want to use more number of coils due to financial constraints. Amplatzer vascular plug was not available. So, the only option left was using glue. In addition, we have been using glue in other types of embolization routinely and thus felt that controlled injection of glue could also be done in this case without significant risk of complications. To our knowledge, this is the first case of its kind to be managed by N-butyl cyanoacrylate as the embolizing agent (PubMed search, using MeSH keywords — Arteriovenous Malformations, Butyl 2-Cyanacrylate, Therapeutic Embolization, Bronchopulmonary Sequestration). Favorable post-procedure results indicate that transcatheter glue embolization can be considered a suitable alternative to coil/plug embolization in these cases. However, further studies are required to support this technique of embolization.

Abbreviations CT: Computed tomography; NBCA: N-Butyl cyanoacrylate; PVA: Polyvinyl alcohol; RDC: Renal double curve

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Data Availability The data that support the findings of this study are available from the corresponding author, [AG], upon reasonable request.

Declarations

Ethics committee approval Not required.

Human and Animal Rights Statement All procedures performed in studies involving human participants were in accordance with the ethical standards of the institutional and/or national research committee and with the 1964 Helsinki declaration and its later amendments or comparable ethical standards.

Informed consent for publication Consent for publication was obtained for every individual person's data included in the study.

Conflict of interest The authors declare that they have no conflict of interest.

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