

Embolization of pulmonary sequestration with Onyx: an unusual application

Pulmoner sekestrasyonun Onyx ile embolizasyonu: Farklı bir uygulama

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Summary– We report a baby with intralobar pulmonary sequestration who was successfully treated with a new embolization agent, Onyx. A 1.5-month-old female infant was admitted to our hospital with sweating and fatigue. Telecardiography showed cardiomegaly, dextrocardia, and increased pulmonary vascular markings. In thoracic computerized tomography, pulmonary sequestration, right pulmonary hypoplasia, and large collateral arteries were seen. The collateral arteries were originating from the celiac trunk and aorta. Echocardiography revealed enlargement of the left atrium and ventricle and left ventricle systolic dysfunction. Angiography revealed a large feeding artery and three branches originating from the aorta and another feeding artery originating from the celiac trunk. We performed embolization of the feeding arteries and their branches, with coils and Onyx. The procedure was performed without complications, and all feeding arteries were completely occluded. The infant started to gain weight. One year later, the infant's body weight had increased and she had no respiratory problems or signs of congestive heart failure. In this case report, we suggest that embolization with Onyx is a reliable alternative method to surgery for infants with pulmonary sequestration. With future studies, pulmonary sequestration embolization with Onyx may become an acceptable and easy treatment option in pediatric patients.

Pulmonary sequestration (PS) treatment was previously based on the surgical removal of the tissue.

We report an infant with intralobar PS who was successfully treated with a new embolic agent, Onyx.

Özet– Bu yazıda, yeni embolizan ajan olan Onyx ile başarılı bir şekilde tedavi edilmiş intralober pulmoner sekestrasyonu olan bir bebek hasta sunuldu. Bir ay 15 günlük kız hasta terleme ve yorulma şikayetleri ile hastanemize getirildi. Telekardiyografide kardiyomegali ve dekstrocardi bulunduğ, pulmoner vasküler gölgelerin arttığı saptandı. Toraksın bilgisayarlı tomografisinde pulmoner sekestrasyon, sağ akciğer hipoplazisi, geniş kollateral arterler görüldü. Kollateral arterler trunkus çölyakus ve aortadan köken alıyordu. Ekokardiyografi ile sol atriyum ve sol ventrikülde genişleme, sol ventrikülde sistolik fonksiyon bozukluğu belirlendi. Anjiyografi ile, aortadan kaynaklanan geniş besleyici arter ve 3 dalı ile, trunkus çölyakustan kaynaklanan başka bir besleyici arter saptandı. Besleyici arterlere ve dallarına önce coil sonra Onyx ile embolizasyon uyguladık. İşlem komplikasyonsuz olarak uygulandı ve besleyici arterler tam olarak kapatıldı. Olgu, işlem sonrasında kilo almaya başladı. Bir yıl sonra yapılan kontrolde vücut ağırlığının arttığı, hiçbir solunum sorununun olmadığı ve konjestif kalp yetersizliği bulgularının kaybolduğu görüldü. Bu olgu sunumu ile, pulmoner sekestrasyon bulunan çocuklarda Onyx ile embolizasyonun cerrahiye alternatif, etkili bir yöntem olarak kullanılabileceğini göstermek istedik. Gelecekteki çalışmalarla, çocuklarda, Onyx ile pulmoner sekestrasyon embolizasyonu, kabul edilebilir ve kolay bir tedavi seçeneği olarak yerini alacaktır.

CASE REPORT

A 1.5-month-old female infant was admitted to our hospital with excessive sweating and fatigue during feeding. She had a history of recurrent respiratory in-

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Abbreviations:

AVMs Arteriovenous malformations
PS Pulmonary sequestration

fections. On physical examination, the patient's weight was 3600 g and height was 55 cm, both below the fifth percentile for age. Her saturation was 98%. A grade I/VI systolic ejection murmur was audible at the third intercostal space, to the right of the sternum. Telecardiography showed cardiomegaly, dextrocardia, and increased pulmonary vascular markings. On the electrocardiographic examination, the QRS axis was 45 degrees, PR 0.12 seconds (s), corrected QT 0.38 s, and heart rate 150 beats/minute. Echocardiography revealed enlargement of the left atrium and ventricle and left ventricle systolic dysfunction (ejection fraction: 50%). The thoracic computerized tomography revealed PS, right pulmonary hypoplasia, shift of cardiac and mediastinal structures to the right, and large venous and arterial structures in the mid and lower zones of the right lung. The arterial collaterals were originating from the celiac trunk and aorta. The diameter of the artery originating from the aorta was 6 and of the artery originating from the celiac trunk was 3 mm. After sedation with ketamine, midazolam and local anesthesia, the patient was catheterized via arterial and venous routes with 4 and 5 French sheaths, respectively. Aortic pressure was 98/50, mean 70 mmHg, and there was right pulmonary artery hypoplasia. Emboliza-

tion of PS was decided. Angiography revealed a large feeding artery and three branches originating from the aorta and another feeding artery originating from the celiac trunk (Figure 1a). We performed embolization of the feeding arteries and their branches with two 4/30, three 4/41, four 3/23, and two 5/58 microcoils (MA, USA) (Figure 1b). However, angiography after embolization with microcoils indicated incomplete occlusion. Therefore, we used Onyx, a very expensive agent comparatively, for embolization of all feeding arteries (Figure 1c). Ultraflow microcatheter (Covidien, USA) was used during Onyx embolization, 0.25 ml dimethyl sulfoxide (DMSO) was injected in the ultraflow microcatheter to fill the dead space, and then 2.5 ml Onyx (ev3, USA) was injected slowly under fluoroscopy to fill the feeding arteries. The procedure was performed without complications, and the feeding arteries were completely occluded. One year later, the infant's body weight had increased and she had no respiratory problems or signs of congestive heart failure. On the follow-up, left ventricle systolic function had improved (ejection fraction: 65%).

DISCUSSION

Pulmonary sequestration (PS) is a rare congenital anomaly of nonfunctional pulmonary tissue that does

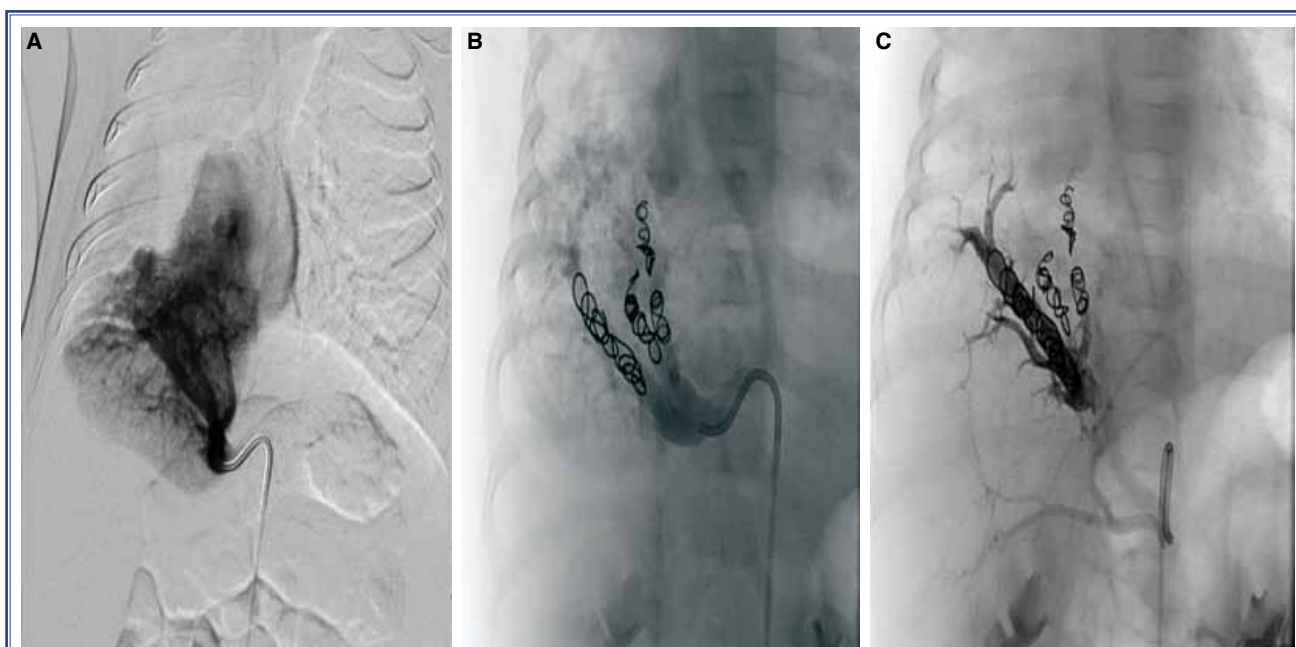


Figure 1. (A) Angiogram showing the arterial collaterals originating from the aorta. (B) Three branches from the feeding artery after embolization with microcoils. (C) Large vessel embolization with Onyx.

not have a normal connection with the bronchial tree or the pulmonary arteries.^[1,2] Blood supply of the tissue comes from an anomalous systemic artery or from arteries that originate from the aorta, usually the thoracic or abdominal aorta. Venous drainage is commonly directed to the pulmonary veins, and rarely to the azygos, hemiazygos, or portal vein.

The aim of the treatment is to prevent right-to-left shunting of blood in arteriovenous fistulas, to prevent left ventricular volume overload, and to occlude unwanted shunts.^[3] Patients with PS were previously treated by surgical removal of the abnormal tissue. Endovascular treatment of PS is another method of management. Selective embolization of the feeding arteries is a less-invasive therapeutic option and carries fewer complications than conventional surgery.^[4]

The embolization materials used are coils, vascular plugs and N-butyl cyanoacrylate. Coil embolization can be an effective procedure for managing a wide variety of aorta-pulmonary collateral vessels and shunts in children with congenital heart disease. The vascular plug is a self-expandable cylindrical mesh device, particularly useful for the embolization of large vessels with high flow. N-butyl cyanoacrylate (glue) is a liquid embolization agent that is mixed with lipiodol in a certain ratio. Onyx is a new effective embolic agent, with several applications in peripheral embolizations. Onyx is an elastic polymer comprised of ethylene-vinyl alcohol copolymer in DMSO with micronized tantalum powder.^[5] It provides contrast for fluoroscopic visualization and is relatively simple to use. It is a liquid, nonadhesive, nonabsorbable, injectable, and permanent embolic agent that can be used off-label for small and large vessels.^[6] It has been used mainly for the treatment of peripheral and cerebral arteriovenous malformations (AVMs), cerebral aneurysms and abdominal aorta stent graft-related endoleaks.^[7] While Onyx is currently only approved for the treatment of intracranial AVM, this case report aims to describe a different and successful application, such as PS, for which this effective and safe embolic agent may be used. In our patient, Onyx was seen to be more effective in the embolization of PS than microcoils with open configuration. After Onyx embolization, the clinical findings of congestive heart failure disappeared.

Saxena and colleagues^[8] reported a 40-year-old man with PS in whom embolization with polyvinyl

alcohol particles was decided. Unfortunately, he required lobectomy three weeks after the embolization. However, in our case, it was sufficient to embolize PS with Onyx.

In contrast to previously used liquid embolic materials such as N-butyl cyanoacrylate, Onyx allows longer injection times and better control of injections because of its longer polymerization time. Due to the lack of adherence, it also has a better penetration advantage than coils, N-butyl cyanoacrylate and vascular plugs.^[9] As it is nonadhesive, it is easier and safer to deliver, with low risk of sticking to the microcatheter tip, and it is viscous enough to allow for controlled motion.^[10] These features are important in the treatment of peripheral AVMs with variable nidus size and number of pedicles. There is low risk of the microcatheter lodging in the target area.

The most important factor creating difficulty in Onyx use is depositing an adequate amount of a liquid embolic agent in AVMs. The incidence of complications is relatively low. Complications such as infection, coloration of the skin, embolization of an erroneous localization, insufficient embolization, or lodging of the microcatheter in the target vessel are very rare. We did not encounter any complication related with Onyx use in our patient, except for the very bad odor from the skin due to the Onyx itself.

In conclusion, this case report suggests that embolization with Onyx is a safe alternative method to surgery and to other embolization agents in infants with PS who present with congestive heart failure. This case provides the first data on the safety and efficacy of embolization of PS with Onyx, and its preference over coils, N-butyl cyanoacrylate and plugs. With future studies, PS embolization with Onyx may become a valuable treatment option in pediatric patients.

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Key words: Child; congenital abnormalities/therapy; embolization, therapeutic; chemoembolization, therapeutic; Onyx copolymer; vascular malformations.

Anahtar sözcükler: Çocuk; doğumsal anormallikler/terapi; embolizasyon, terapötik; kemoembolizasyon, terapötik; Onyx kopolimer; vasküler malformasyon.