OLGU SUNUMU / CASE REPORT

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Perkütan Atriyal Septal Defekt Kapama Sonrası Aorto-Atriyal Fistüle Bağlı İntravasküler Hemoliz

Intravascular Hemolysis Due to Aorto-Atrial Fistula After Percutaneous Atrial Septal Defect Closure

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ÖZ

Perkütan atriyal septal defekt (ASD) kapama, seçilmiş hastalarda cerrahiye kıyasla daha kısa iyileşme süresi ve daha düşük komplikasyon oranları gibi avantajlar olan ve giderek daha fazla tercih edilen bir yöntemdir. Her ne kadar güvenli bir tedavi yöntemi olarak kabul edilse de, nadiren kardiyak erozyon gibi yaşamı tehdit edebilecek komplikasyonlara yol açabilir. Bu vaka raporunda perkütan ASD kapamadan iki ay sonra intravasküler hemoliz bulguları ile başvuran 38 yaşındaki erkek bir hastaya ait nadir bir olgu sunulmaktadır. Başlangıçta hematüri nedeniyle üriner sistem kaynaklı bir kanama düşünülen hastada, ileri tetkikler sonucunda, cihaz kaynaklı aortik erozyona bağlı olarak non-koroner sinüs ile sol atriyum arasında gelişmiş bir fistül saptanmıştır. Kapama cihazının cerrahi olarak çıkarılması ve fistülün otolog perikardiyal yama ile onarılması sonrasında hastanın semptomları tamamen gerilemiştir. Bu vaka raporu, perkütan ASD kapama sonrası açıklanamayan hemoliz gelişen hastalarda aorto-atriyal fistül gibi yapısal komplikasyonların da göz önünde bulundurulması gerektiğini vurgulamaktadır.

Anahtar Kelimeler: amplatzer septal occluder, aorto-atriyal fistül, atriyal septal defekt, hemolitik anemi

ABSTRACT

Percutaneous closure of atrial septal defects (ASD) is an increasingly utilized method for selected patients, offering advantages such as shorter recovery times and lower complication rates compared to surgery. Despite being considered as a rather safe treatment method, it may rarely lead to life-threatening complications such as cardiac erosion. We report a case of a 38-year-old male who presented with symptoms of intravascular hemolysis two months after percutaneous ASD closure. Despite initial suspicion of urinary bleeding due to hematuria, further evaluation revealed a fistula between the non-coronary sinus and the left atrium, caused by device-related aortic erosion. Surgical removal of the closure device and successful repair of the fistula with autologous pericardial patches resolved the symptoms. This case underscores the importance of considering structural complications, including aorto-atrial fistula, in patients with unexplained hemolysis following percutaneous ASD closure.

Keywords: amplatzer septal occluder, aorto-atrial fistula, atrial septal defect, hemolytic anemia

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INTRODUCTION

Percutaneous atrial septal defect (ASD) closure has emerged as a safe and effective treatment method in selected patients with secundum type ASD [1]. Although this minimally invasive procedure has advantages over surgery such as shorter recovery times and lower complication rates, it may rarely be associated with serious adverse events [2]. During the early post-interventional period, potential complications may include residual shunts, device embolization, or thromboembolism [3]. On the other hand, late complications, such as aortic or atrial erosion, perforation, endocarditis, cardiac rupture, or development of aorto-atrial fistulas, may emerge months or even years after the procedure [4].

Intravascular hemolysis due to cardiac erosion and aorto-atrial fistula is an extremely rare complication following ASD closure. There are only two cases in the literature where cardiac erosion was diagnosed following findings consistent with hemolysis after ASD closure [5,6]. We present a case of an aorto-atrial fistula due cardiac erosion following ASD closure, which was incidentally diagnosed in the work-up of intravascular hemolysis, to highlight the importance of recognition and management of such anomalies in these patients.

CASE REPORT

A 38-year-old male patient was referred to our cardiology outpatient clinic for work-up of intravascular hemolysis. He was diagnosed with secundum type ASD (defect size: 32x17mm; rims: anteroinferior-23mm, posterosuperior-8mm, aortic-6mm, posteroinferior-12mm, superior vena cava-14mm, inferior vena cava-15mm) in another hospital two months ago, which was closed percutaneously by a 36mm The Amplatzer Septal Occluder (ASO) in that center. No complication was noted during the periprocedural and early post-procedural period and he was discharged with dual antiplatelet therapy (daily 100mg aspirin and 75mg clopidogrel). The patient's past medical history revealed no other significant chronic condition or prior utilization of long-term medications.

One week after the procedure, the patient had dark-colored urine. Standard abdominal and urogenital examinations and abdominal computed tomography scanning revealed no abnormality. Due to persistent dark-colored urine and a decrease in hemoglobin levels (pre-procedure hemoglobin: 16.5g/dL and current hemoglobin: 10.1g/dL) suggestive of potential urinary bleeding, the dual antiplatelet therapy was discontinued. However, red urine persisted and jaundice was developed. Biochemical analysis revealed elevated lactate dehydrogenase, aspartate aminotransferase and bilirubin levels, indicating a potential hemolytic process. Peripheral blood smear analysis revealed normochromic normocytic erythrocyte morphology with acanthocytes and polychromasia. The negative results of both direct and indirect Coombs tests collectively indicated a non-immune intravascular hemolysis. For work-up of intravascular hemolysis, the patient was referred to our outpatient clinics for an echocardiographic examination, especially to look for a complication of ASD closure process since the symptoms of the patients started one week after the procedure.

The transthoracic echocardiography showed a strong echo of the occluder in the interatrial septum with no apparent shunt (Figure 1). However, during transesophageal echocardiography, a fistula connecting the left atrium to the aortic root, which originated from the non-coronary sinus due to aortic erosion, was observed (Figure 2). An aortogram provided passage of the contrast agent from aorta to the left atrium. The heart team decided a surgical repair. The patient underwent aortotomy and right atriotomy, which allowed identification of the fistula between the left atrium and the non-coronary sinus (Figure 3), and visualization of the closure device with multiple small thrombi adhered to its surface (Figure 4).The ASD closure device was carefully removed from the interatrial septum. Subsequently, the fistula was surgically closed using autologous pericardial patches. A patch measuring 10x10mm was applied on the aortic aspect, accompanied by a corresponding patch measuring 15x15mm on the atrial side. Control transesophageal echocardiography confirmed the successful closure of the fistula. The patient was discharged without any complications and was asymptomatic during the follow-up visits.

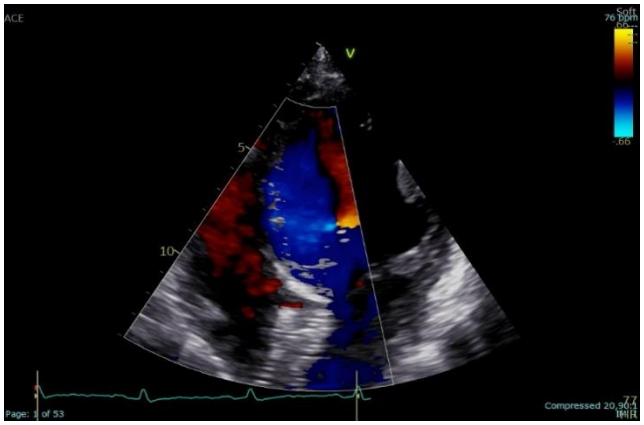

Figure 1. Transthoracic echocardiography – apical 4 chamber view: the strong echo of the occluder in the atrial septum is seen with no apparent Color Doppler flow at the atrial level

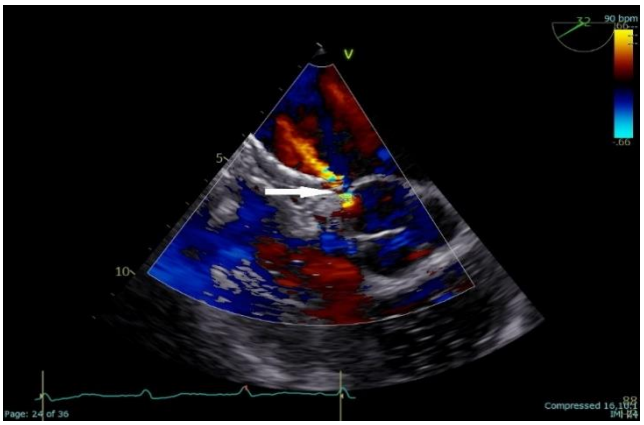

Figure 2. Transesophageal echocardiography – mid esophageal short axis view: a fistula is seen connecting the left atrium to the aortic root, which originated from the non-coronary sinus due to aortic erosion.

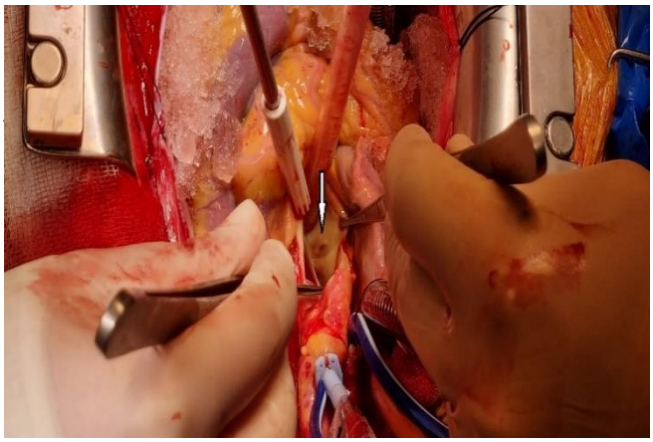

Figure 3. The fistula between the non-coronary sinus and the left atrium

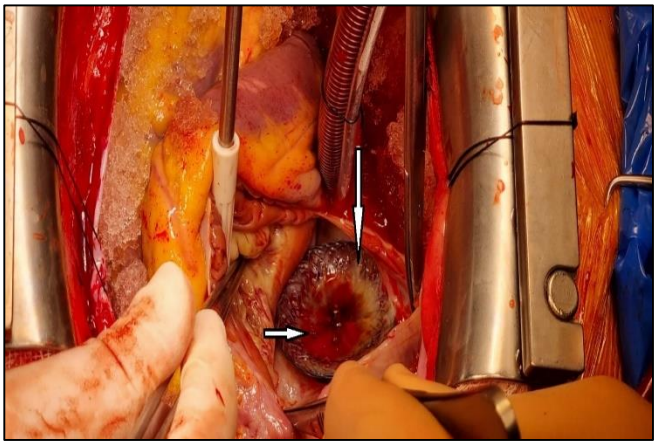

Figure 4. After aortotomy and right atriotomy, the closure device and small multiple thrombi are seen.

DISCUSSION

This case highlights intravascular hemolysis secondary to an aorto-left atrial fistula resulting from device-related erosion complication following percutaneous ASD closure. Although similar cases have been described in the literature, our report adds new insights by emphasizing the diagnostic challenge of hemolysis mimicking hematuria, the potential impact of early dual antiplatelet therapy discontinuation, and the surgical findings of thrombus formation on the device surface.

Cardiac erosions, with a reported incidence ranging from 0.1% to 0.3% are usually observed within the initial few days following the procedure while they may develop even years after the procedure [7,8]. Clinical presentation of erosion may vary from mild symptoms to severe symptoms such as acute cardiac tamponade requiring urgent pericardiocentesis [9]. Insufficient aortic rims and oversized devices are usually associated with the risk of erosion [10,11] while aortic aneurysm and hypertension may increase aortic wall tension, leading to erosion in a later period [12]. Although the rims seemed anatomically appropriate for closure in this case, the choice of a relatively large device (36 mm) may have increased mechanical stress or friction on nearby cardiac structures, potentially resulting in erosion and the formation of a fistula.

Aorto-atrial fistula may occur from device erosion or pressure necrosis on the aortic wall and are often diagnosed during routine follow-ups upon detection of a newly developed murmur unless they result in acute pericardial tamponade. Intravascular hemolysis due to cardiac erosion and aorto-atrial fistula is an extremely rare complication of percutaneous ASD closure with only two reported cases in the literature [5,6]. Intravascular hemolysis is usually caused by high-velocity flow, vortices, and rapid deceleration, exerting significant shear stress on red blood cells. The most likely mechanism for hemolysis in this case is related to the high-velocity blood flow from the aorta into the left atrium through the fistulous tract, which impacts the closure device directly. This high-shear flow likely leads to mechanical fragmentation of red blood cells due to collisions with the occluder surface. The development of an aorto-atrial fistula, accompanied by hemolysis, suggests that the high-velocity blood flow through the fistula was the primary cause of the symptoms. The resolution of these symptoms after surgically closing the fistula and removing the device further supports this hypothesis.

Urgent surgery with removal of the closure devices and repair of the defects is the primary treatment modality in patients experiencing cardiac erosion following percutaneous ASD closure [13]. In our patient, the Amplatzer device was removed and fistula was closed with autologous pericardial patches. During the surgical removal of the device, intense thrombus material was observed, probably due to premature discontinuation of dual antiplatelet therapy as he was supposed to have urinary bleeding. Current recommendations suggest the continuation of dual antiplatelet therapy for the first 6 months following percutaneous ASD closure [14].

CONCLUSION

This report points out a rare complication of percutaneous ASD closure procedure. Intravascular hemolysis may develop due to aorto-atrial erosion and fistula. Accurate diagnosis and surgical treatment of the fistula are essential in the management of these patients.

Conflict of Interest: All authors declare no conflicts of interest**.**

Informed Consent: The patient was thoroughly informed about the potential scientific contribution of the case report and provided both written and verbal consent for its publication.

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