Case Report

A Rare Case of Subarachnoid Hemorrhage in a Normotensive Young Hemodialysis Patient Followed up With Familial Mediterranean Fever

Mehmet BİRİCİK[1,a]

[1]*Savur Prof. Dr. Aziz Sancar Devlet Hastanesi, İç Hastalıkları Kliniği, Mardin, Türkiye*

ABSTRACT

Nontraumatic Subarachnoid hemorrhage (SAH) is a rare but life-threatening complication in patients undergoing hemodialysis (HD). In this instance, we disclose the occurrence of a nontraumatic SAH in a 28-year-old male patient diagnosed with Familial Mediterranean Fever (FMF) while undergoing HD treatment. The SAH was diagnosed on the basis of clinical and radiological findings. The patient had a history of recurrent attacks of FMF and was undergoing HD for end-stage renal disease (ESRD). The patient was treated with supportive care and close monitoring. Despite all efforts, the patient passed away after 20 days. The cause of SAH in our patient remains unclear, but we think that it is multifactorial in nature and that AA-type amyloidosis may cause this condition with an autoinflammatory reason.

Keywords: *Subarachnoid Hemorrhage, Familial Mediterranean Fever, Hemodialysis.*

ÖZ

Ailevi Akdeniz Ateşi ile Takip Edilen Normotansif Genç Hemodiyaliz Hastasında Nadir Bir Subaraknoid Kanama Olgusu

Travmatik olmayan Subaraknoid kanama (SAK), hemodiyalize (HD) giren hastalarda nadir görülen ancak yaşamı tehdit eden bir komplikasyondur. Bu örnekte, HD tedavisi görürken Ailesel Akdeniz Ateşi (FMF) teşhisi konan 28 yaşında bir erkek hastada travmatik olmayan bir SAH oluşumunu sunuyoruz. SAK tanısı klinik ve radyolojik bulgulara dayanılarak konuldu. Hastanın tekrarlayan FMF atakları öyküsü vardı ve son dönem böbrek hastalığı (ESRD) için HD görüyordu. Hasta destekleyici bakım ve yakın takip ile tedavi edildi. Tüm çabalara rağmen hasta 20 gün sonra hayatını kaybetti. Hastamızdaki SAK'ın nedeni belirsizliğini koruyor, ancak doğası gereği multifaktoriyel olduğunu ve AA tipi amiloidozun otoinflamatuar bir nedenle bu duruma neden olabileceğini düşünüyoruz.

Anahtar Sözcükler: *Subaraknoid Kanama, Ailesel Akdeniz Ateşi, Hemodiyaliz.*

Bu makale atıfta nasıl kullanılır: Biricik M. Ailevi Akdeniz Ateşi ile Takip Edilen Normotansif Genç Hemodiyaliz Hastasında Nadir Bir Subaraknoid Kanama Olgusu. Fırat Tıp Dergisi 2024; 29(1): 60-62.
How to cite this article: Biricik M. A Rare Case of Subarachnoid Hemorrhage in a Normotensive Young Hemodialysis Patient Followed up With Familial Mediterranean Fever. Firat Med J 2024; 29(1): 60-62.

ORCID IDs: M.B. 0000-0002-5701-4978.

Subarachnoid hemorrhage (SAH) is a rare but life-threatening complication in patient undergoing hemodialysis (HD) and it is associated with a high mortality rate if not promptly diagnosed and treated. SAH is characterized by bleeding into the subarachnoid space between the arachnoid membrane and the pia mater (1). SAH has been reported to occur more frequently in patients undergoing HD than in the general population (2).Familial Mediterranean Fever (FMF) is an autosomal recessive inheritance pattern, characterized by recurring episodes of fever and serositis, which predominantly impact individuals with Mediterranean heritage. FMF has been associated with several systemic manifestations, including vasculitis, arthritis, and amyloidosis. In addition, FMF, an autoinflammatory disease, has been implicated as a potential risk factor for the development of SAH. Neurological complications are rare in FMF, but acute stroke has been reported in some cases. However, the exact mechanisms underlying the increased risk of SAH in HD patients with FMF remain unclear. Several mechanisms have been proposed to explain the association between FMF and SAH, including cerebral vasculitis, arterial wall inflammation, and endothelial dysfunction. This case report presents a rare case of spontaneous, nonaneurysmal, nontraumatic, SAH in a normotensive young HD patient with FMF and discusses the potential contributing factors and management strategies.

CASE REPORT

Following one day after his HD session, a 28-year-old male patient with a medical history of FMF, arrived at the emergency department experiencing a sudden onset of severe headache and vomiting. The patient had been undergoing HD three time in a week for the past 3 years due secondary end-stage renal disease (ESRD) to FMF. He had a history of recurrent attacks of FMF for the past 10 years, which were not well-controlled with colchicine therapy. The patient had no history of hyper-

[a]Yazışma Adresi: Mehmet BİRİCİK, Savur Prof. Dr. Aziz Sancar Devlet Hastanesi, İç Hastalıkları Kliniği, Mardin, Türkiye
Tel: 0544 643 1879 e-mail: dr.mehmetbiricik@gmail.com
Geliş Tarihi/Received: 29.03.2023 Kabul Tarihi/Accepted: 06.11.2023

tension or other comorbidities. On examination, the patient was alert and oriented with no focal neurological deficits. His blood pressure was 140/80 mmHg. In the examinations of the patient, PT, aPTT, INR, thrombocyte and bleeding time were within the normal range. The non-contrast computed tomography (NCCT) showed no evidence of aneurysms or vascular malformations (Figure 1, 2). CT brain showed SAH (Figure 2).

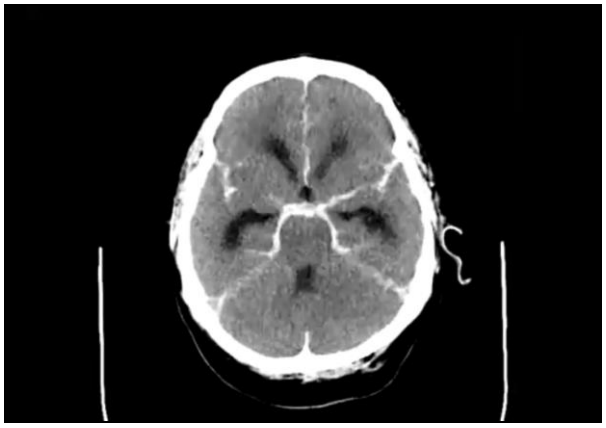

Figure 1. *Non-contrast computerized tomography brain showing subarachnoid hemorrhage in a young Hemodialysis patient (black arrow).*

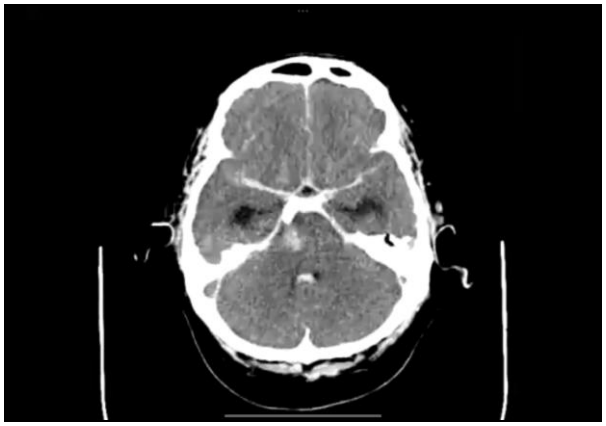

Figure 2. *Repeat non-contrast computerized tomography brain showed Subarachnoid hemorrhage (black arrow).*

The patient did not report any incidents of head trauma or falling, nor had a bleeding diathesis or intracerebral bleed in his medical history. Additionally, following consultation with a neurosurgeon, nimodipine was initiated, while Mannitol, Dexamethasone, and other "anti-edema" treatments were not administered.

The patient was admitted to the intensive care unit for close monitoring and supportive care. He was continued on nimodipine and started antiepileptic therapy. The patient's HD was continued without heparin and he remained hemodynamically non-stable throughout his hospitalization. The patient passed away after 20 days of hospitalization in intensive care unit.

DISCUSSION

SAH is a rare but serious complication that can occur in patients undergoing HD. SAH is defined as bleeding into the subarachnoid space, which is the area between the arachnoid mater and the pia mater that covers the brain. In HD patients, SAH is associated with higher morbidity and mortality rates compared to the general population (3). The exact etiology of SAH in HD patients is not fully understood, but it is believed to be multifactorial in nature. Research conducted on the general public has demonstrated that the likelihood of developing SAH is higher in females, individuals with hypertension, smokers, those who abuse alcohol, and people of non-white racial backgrounds. (4-6).

Approximately 85% of all cases of SAH arise from the rupture of saccular aneurysms located at the base of the brain. Non-aneurysmal perimesencephalic hemorrhages constitute 10% of SAH cases, with the remaining 5% stemming from other uncommon causes. Risk factors for SAH include genetic predisposition and modifiable factors such as smoking, hypertension, and excessive alcohol consumption. A significant risk factor is having a family history of SAH, as first-degree relatives have a 3 to 7 times higher risk of experiencing it. Certain inherited disorders, such as Autosomal Dominant Polycystic Kidney Disease (ADPKD), are linked to a higher susceptibility of SAH. Roughly 2% of SAH patients have been diagnosed with ADPKD. While other inherited conditions, such as Ehlers-Danlos Disease IV, neurofibromatosis type 1, and Marfan's Syndrome, have also been linked to SAH, they are less commonly found in affected patients. The rupture of aneurysms may be triggered by a sudden increase in intramural arterial pressure. Activities that can cause bleeding, such as physical exercise, sexual intercourse, and straining, have been reported in up to 20% of cases. Individuals receiving maintenance dialysis have an elevated likelihood of experiencing bleeding compared to the overall populace. Earlier studies have indicated that this group is especially susceptible to gastrointestinal bleeding and subdural hematomas. (7, 8). The heightened risk of bleeding observed in patients receiving maintenance dialysis is believed to result from defects in the coagulation pathway and the administration of anticoagulation treatment during dialysis. Studies have reported an increased incidence of SAH in HD patients, with a prevalence of up to 6.8% in some populations (9). A retrospective study analyzing the incidence of SAH in HD patients found that the risk of SAH was higher in HD patients compared to non-HD patients, with a standardized incidence ratio of 4.4 (9,10). Another study found that HD patients with SAH had a higher mortality rate and worse outcomes compared to non-HD patients with SAH (9,11). Cerebrovascular accidents such as acute ischemic stroke, subarachnoid hemorrhage and hematoma are not common in FMF patients. In a study of 23 FMF patients, a total of 35 acute stroke events were observed. Acute ischemic stroke attack was observed in 19 of these patients,

transient ischemic attack in 3 and parietal hematoma in 1 patient. Subarachnoid hemorrhage was not seen (12).
The management of SAH in HD patients can be challenging, with surgical intervention being the mainstay of treatment. However, these patients may be at increased risk of complications such as cerebral vasospasm and rebleeding. Therefore, early diagnosis and appropriate management are crucial in improving patient outcomes.
The exact etiology of SAH in HD patients remains unclear, but it is thought to be multifactorial in nature, and further studies are needed to better understand the underlying mechanisms of this complication. In this case report, we present the case of a 28-year-old male patient with FMF who developed SAH during his HD session. Despite prompt diagnosisand supportive care, the patient passed away after 20 days of hospitalization. In this publication, we aimed to present a case of SAH in a patient who undergoing HD due to ESRD secondary to secondary FMF. We did not come across any publications with a history of FMF in previous studies or case reports. In this aspect, the case we presented has the feature of being the first. We thought that the underlying reason for this may be that FMF is an autoinflammatory condition and may have triggered SAH in our patient. We wanted to emphasize that researchers should keep in mind that HD patients with an autoinflammatory disease such as vasculitis or FMF are at risk of SAH.

Conclusion

The etiology of SAH in our patient remains unclear. Further studies are needed to elucidate the underlying mechanisms of SAH in HD patients with FMF. Prompt diagnosis and appropriate management are crucial in improving the outcome of such patients.

REFERENCES

1. Onoyama K, Kumagai H, Miishima T etal. Incidence of strokes and its prognosis in patients on maintenance hemodialysis. Jpn Heart J 1986; 27: 685-91.
2. Seliger SL, Gillen DL, Longstreth WT Jr, Kestenbaum B, Stehman-Breen CO. Elevated risk of stroke among patients with end-stage renal disease. Kidney Int 2003; 64: 603-9.
3. Iseki K, Fukiyama K, Okawa Dialysis Study (OKIDS) Group. Clinical demographics and long-term prognosis after stroke in patients on chronic haemodialysis. The Okinawa Dialysis Study (OKIDS) Group. Nephrol Dial Transplant 2000; 15: 1808-13.
4. van Gijn J, Rinkel GJ. Subarachnoid haemorrhage: Diagnosis, causes and management. Brain 2001; 124: 249-78.
5. Feigin VL, Rinkel GJ, Lawes CM et al. Risk factors for subarachnoid hemorrhage: an updated systematic review of epidemiological studies. Stroke 2005; 36: 2773-80.
6. Sandvei MS, Romundstad PR, Müller TB, Vatten L, Vik A. Risk factors for aneurysmal subarachnoid hemorrhage in a prospective population study: the HUNT study in Norway. Stroke 2009; 40: 1958-62.
7. Sood P, Kumar G, Nanchal R et al. Chronic kidney disease and end-stage renal disease predict higher risk of mortality in patients with primary upper gastrointestinal bleeding. Am J Nephrol 2012; 35: 216-24.
8. Sood P, Sinson GP, Cohen EP et al. Subdural hematomas in chronic dialysis patients: significant and increasing. Clin J Am Soc Nephrol 2007; 2: 956-9.
9. Sakhuja A, Schold JD, Kumar G, Katzan I, Navaneethan SD. Nontraumatic subarachnoid hemorrhage in maintenance dialysis hospitalizations: Trends and outcomes. Stroke 2014; 45: 71-6.
10. Murakami M, Hamasaki T, Kimura S, Maruyama D, Kakita K. Clinical features and management of intracranial hemorrhage in patients undergoing maintenance dialysis therapy. Neurol Med Chir (Tokyo) 2004; 44: 225-32.
11. Davenport A. Changing the hemodialysis prescription for hemodialysis patients with subdural and intracranial hemorrhage. Hemodial Int 2013; 17: S22-7.
12. Topcuoglu MA, Ayvacioglu-Cagan C, Arsava EM et al. Acute stroke in familial Mediterranean fever: An analysis of 23 new cases and systematic review of the literature. Int J Rheum Dis 2023; 26: 657-66.