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Case Report

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Role and Safety of Plasmapheresis in the Treatment of Moyamoya Syndrome

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Abstract

We are presenting a case of Graves' disease with acute deterioration in mental status secondary to moyamoya syndrome that developed because of untreated severe thyrotoxicosis. The patient did not respond well to medications used to treat thyrotoxicosis and underwent plasmapheresis aiming to improve symptoms and neurologic outcome. After one session of plasmapheresis, she developed cerebral herniation. This article will focus in brief on the role of plasmapheresis in the treatment of complicated cases and the safety of this procedure, especially in critically ill patients.

Keywords: Moyamoya syndrome; Thyrotoxicosis; Graves' disease; Plasmapheresis

Introduction

Moyamoya syndrome (MMS) is a rare cause of cerebrovascular insult that can lead to brain ischemia and bleeding. It is an angiogenic disorder of progressive narrowing of the cerebral arteries at the base of the brain, typically involving the intracerebral portion of the internal carotid arteries. MMS can develop in association with head irradiation, chemotherapy, and many other disorders, such as neurofibromatosis type 1 (NF1), Down syndrome and autoimmune disorders. It can be idiopathic as well. The diagnosis can be easily missed given the rarity of this vascular syndrome. Graves' disease (GD) has been rarely reported to cause MMS, and the underlying coexisting mechanism remains unclear. MMS is incurable, and the goal of treatment is directed toward improving cerebral blood flow. We are reporting a case of a young woman admitted for thyroid storm with acute onset of focal neurologic symptoms and altered mentation with tragic outcome. She was treated with plasmapheresis and developed cerebral edema after one procedure.

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Case Report

Investigations

A 28-year-old Hispanic female with a history of GD presented to the emergency department with worsening symptoms of palpitations, sweating, weight loss, and leg edema for about 1 month prior to admission. She had not taken antithyroid medications or beta blockers for months, as she was asymptomatic and had not been following up with a provider. The patient was tachycardic on admission, normothermic, alert and oriented with no focal neurologic deficit.

Diagnosis

Thyroid-stimulating hormone (TSH) level on admission was < 0.007 µIU/mL (0.36 - 3.7 µIU/mL), free thyroxine (FT4) > 8.0 ng/dL (0.76 - 1.46 ng/dL), and total triiodothyronine (TT3) was 2.75 ng/mL (0.6 - 1.8 ng/mL).

Treatment

Her methimazole was resumed, and she was placed on beta blocker as well.

Outcomes

A day later, symptoms were improving, and she was planned for discharge, but before discharge, she developed sudden weakness of the right upper and lower extremities. A few minutes later, she became unresponsive and developed seizures. Magnetic resonance angiogram of the brain showed ischemic infarcts with occlusion/near occlusion of the right and left internal carotid arteries, and a diagnosis of MMS was made. After intubation, thyroid storm treatment with higher dose of methimazole, steroids, and beta blocker were initiated. The patient's mental status did not improve, and her Glascow Coma Scale score remained at 5. However, her vital signs were sta-

ble and controlled until she was extubated, at which point she started to have tachypnea, tachycardia and hyperthermia. So, the decision was made to intubate her again, and to start with plasmapheresis, trying to improve symptoms and neurologic outcome. The patient was hemodynamically stable during plasmapheresis. Few hours after the first session of plasmapheresis, the patient's neurologic outcome deteriorated and brain computed tomography (CT) scan showed cerebral edema and herniation, which ultimately led to brain death.

Discussion

MMS is a vascular endothelial disorder that can lead to cerebrovascular accidents and variable neurologic symptoms. It involves a progressive narrowing of cerebral arteries at the base of the brain, more specifically the circle of Willis, where it leads to the development of fragile collateral arteries, leading to the characteristic features found on brain imaging that is diagnostic of MMS [1]. This vascular syndrome is linked to several distinct clinical conditions, including sickle cell disease, systemic lupus erythematosus, Down syndrome, fibromuscular dysplasia NF1, and rarely can be associated with GD [2]. The pathophysiology of MMS, especially in GD, is incompletely understood. Thyroid toxicity is thought to play a role in the pathology of developing cerebral arterial stenosis in patients with GD [3]. This was confirmed by Kim et al, who reported that elevated thyroid autoantibodies and thyrotoxicosis were frequently observed in patients with MMS [4]. While the three most common treatments for hyperthyroidism are antithyroid drugs, radioactive iodine, and thyroidectomy, plasmapheresis is an alternative treatment that has been proposed for many years for hyperthyroidism, especially in severe thyrotoxicosis and thyroid storm [5]. It entails the separation of plasma from blood to eliminate proteins such as antibodies and cytokines; and it is commonly used to improve neurologic outcomes in many autoimmune-associated neurologic disorders, such as myasthenia gravis crisis and Guillain-Barre syndrome, where it is considered as a safe and effective first-line treatment [6, 7]. Plasmapheresis may also have the potential to stabilize the neurovascular progression in patients with MMS associated with GD and possibly prevent further progression of the cerebral arterial occlusive process [8]. Plasmapheresis is considered a safe procedure in literature reviews, with a reported incidence of severe complications of up to 4.75% [9]. In a retrospective study using medical records of 73 patients who underwent 1,283 plasmapheresis procedures for different neurologic diseases, no fatalities were reported [10]. The study concluded the safety of plasmapheresis in patients with neurologic symptoms when the procedure is performed by skilled and well-trained persons [10].

In patients with severe comorbidities, especially those requiring mechanical ventilation and on pressors, like our case, the prevalence of adverse events during plasmapheresis is generally higher. Arrhythmias, life-threatening hypotension, and water and electrolyte imbalance are more common to occur in those patients. Such complications, among others, were observed in a study entitled 370 plasmapheresis procedures in 54 patients in an intensive care unit level [11]. In this study, 88.9% of the procedures were carried out without complications [11].

In this case, no water imbalance was reported during plasmapheresis, and it is unknown whether the plasmapheresis played a role in cerebral edema and herniation in the presence of extensive ischemic cerebrovascular insult.

Conclusions and learning points

It is well known that a significant proportion of thyroid hormones bind to serum proteins. This makes plasmapheresis an effective therapeutic option to achieve euthyroidism and improve clinical outcomes in patients with thyrotoxicosis complications. As an invasive procedure, plasmapheresis is not without complications. However, it can be considered a relatively safe therapeutic method for patients hospitalized in intensive care units in literature reviews. On the other hand, to the best of our knowledge, cerebral edema and herniation were not reported as a complication of plasmapheresis. However, continuous observation and monitoring of patients provided by well-trained personnel are essential for better safety outcomes.

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None to declare.

Financial Disclosure

None to declare.

Conflict of Interest

No conflict of interest to disclose.

Informed Consent

Informed consent was obtained by the patient's next of kin as the patient is deceased.

Author Contributions

Mohammed Al Tameemi: writing the manuscript and conducting the literature search. Basma Ataallah: conducting the literature search.

Data Availability

All data in our report were obtained from the patient's hospitalization. Any inquiries regarding supporting data availability of this study should be directed to the corresponding author.

References

1. Khurram MSA, Rao SP, Khan A. Moyamoya disease with review of literature. Journal of Evolution of Medical and Dental Sciences. 2013;2(48):9419-9422,
2. Miller R, Unda SR, Holland R, Altschul DJ. Western moyamoya phenotype: a scoping review. Cureus. 2021;13(11):e19812. doi pubmed
3. Ohba S, Nakagawa T, Murakami H. Concurrent Graves' disease and intracranial arterial stenosis/occlusion: special considerations regarding the state of thyroid function, etiology, and treatment. Neurosurg Rev. 2011;34(3):297-304; discussion 304. doi pubmed
4. Kim SJ, Heo KG, Shin HY, Bang OY, Kim GM, Chung CS, Kim KH, et al. Association of thyroid autoantibodies with moyamoya-type cerebrovascular disease: a prospective study. Stroke. 2010;41(1):173-176. doi pubmed
5. Ashkar FS, Katims RB, Smoak WM, 3rd, Gilson AJ. Thyroid storm treatment with blood exchange and plasmapheresis. JAMA. 1970;214(7):1275-1279. pubmed
6. Vikrant S, Thakur S, Sharma A, Gupta D, Sharma S. Safety and efficacy of therapeutic membrane plasmapheresis in the treatment of Guillain-Barre syndrome: A study from a tertiary care hospital from India. Neurol India. 2017;65(3):527-531. doi pubmed
7. Strasser E. Principles of therapeutic apheresis in neurological disease. Transfus Med Hemother. 2023;50(2):88-97. doi pubmed
8. Malik S, Russman AN, Katramados AM, Silver B, Mitsias PD. Moyamoya syndrome associated with Graves' disease: a case report and review of the literature. J Stroke Cerebrovasc Dis. 2011;20(6):528-536. doi pubmed
9. Vinan-Vega M, Mantilla B, Jahan N, Peminda C, Nugent K, Lado-Abeal J, Rivas A. Usefulness of plasmapheresis in patients with severe complicated thyrotoxicosis. Proc (Bayl Univ Med Cent). 2020;34(2):279-282. doi pubmed
10. Vucic S, Davies L. Safety of plasmapheresis in the treatment of neurological disease. Aust N Z J Med. 1998;28(3):301-305. doi pubmed
11. Szczeklik W, Wawrzycka K, Wludarczyk A, Sega A, Nowak I, Seczynska B, Fajfer I, et al. Complications in patients treated with plasmapheresis in the intensive care unit. Anaesthesiol Intensive Ther. 2013;45(1):7-13. doi pubmed