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Complex Case Of Moya Moya Syndrome In A Patient With Antiphospholipid Syndrome And Hyperthyroidism: A Case Report Highlighting Treatment Challenges.

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Abstract:
Moya Moya disease (MMD) is a chronic condition which involves occlusive cerebrovascular disorder characterised by the gradual narrowing or blockage of the internal carotid arteries and their main branches, leading to the formation of collateral vessels that appear as a "puff of smoke" on angiographic images.(1,2). Autoimmune disorders such as APLS(3) and autoimmune thyroiditis(4) can lead to chronic inflammation of blood vessels, damaging the endothelial cells lining the cerebral arteries and contributing to the progressive narrowing and occlusion characteristic of Moya Moya disease(5) which is defined as moya moya syndrome. A noteworthy case showcases the intricate management of multiple comorbidities associated with Moyamoya syndrome in a 27-year-old female patient. The presentation included recurrent stroke episodes, further complicated by autoimmune disorders such as APLA and Graves' disease. This particular case underscores the critical need for comprehensive, interdisciplinary care to enhance patient outcomes.

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I. Introduction

Moyamoya disease is a cerebrovascular condition linked to an increased susceptibility to stroke and/or haemorrhage among those affected. This disorder is characterised by a chronic and progressive constriction of the end segment of the bilateral internal carotid arteries or the initial sections of the anterior and/or middle cerebral arteries. Consequently, an anomalous vascular system is established through collateral pathways at the base of the brain. The term "Moyamoya," originating from the Japanese language, denotes the visual resemblance of these collateral blood vessels as observed in cerebral angiograms. Antiphospholipid syndrome (APLS) is a chronic autoimmune condition characterised by the detection of antiphospholipid antibodies in the bloodstream. The presence of these antibodies substantially elevates the likelihood of thrombosis, resulting in the development of blood clots within both arterial and venous systems. The syndrome can also contribute to repetitive pregnancy losses, stroke, and additional thromboembolic occurrences, thereby adding complexity to the clinical presentation of affected individuals. Autoimmune hyperthyroidism, notably Graves' disease, represents an additional pathological state capable of influencing the comprehensive vascular load. The hallmark of Graves' disease involves the excessive synthesis of thyroid hormones and the autoimmune nature of Graves' disease leads to the production of antibodies that target the thyroid gland, causing inflammation and has the capacity to induce angiogenesis(6). The presence of Moya Moya disease, APLS, and Graves' disease in combination poses a distinctive and demanding clinical situation. Prolonged inflammation linked to autoimmune conditions such as APLS and thyroiditis may harm the endothelial cells that line the cerebral arteries, contributing to the progressive vascular alterations observed in Moya Moya disease(6). This case study explores the instance of a 27-year-old woman who exhibited an intricate interplay of these ailments, emphasising the challenges in diagnosis and treatment that were encountered. It is hypothesised that autoimmune mechanisms are involved in the development of MMD, as research suggests that individuals with MMD might have a heightened occurrence of other autoimmune conditions. This case highlights the intricacy of caring for patients with numerous comorbidities and emphasises the significance of employing a thorough, interdisciplinary strategy to enhance patient outcomes. The objective of this report is to offer insights into the physiological mechanisms that connect these conditions, describe the clinical manifestation and diagnostic discoveries, and disclose the treatment strategy and extended outcomes of this multifaceted scenario.

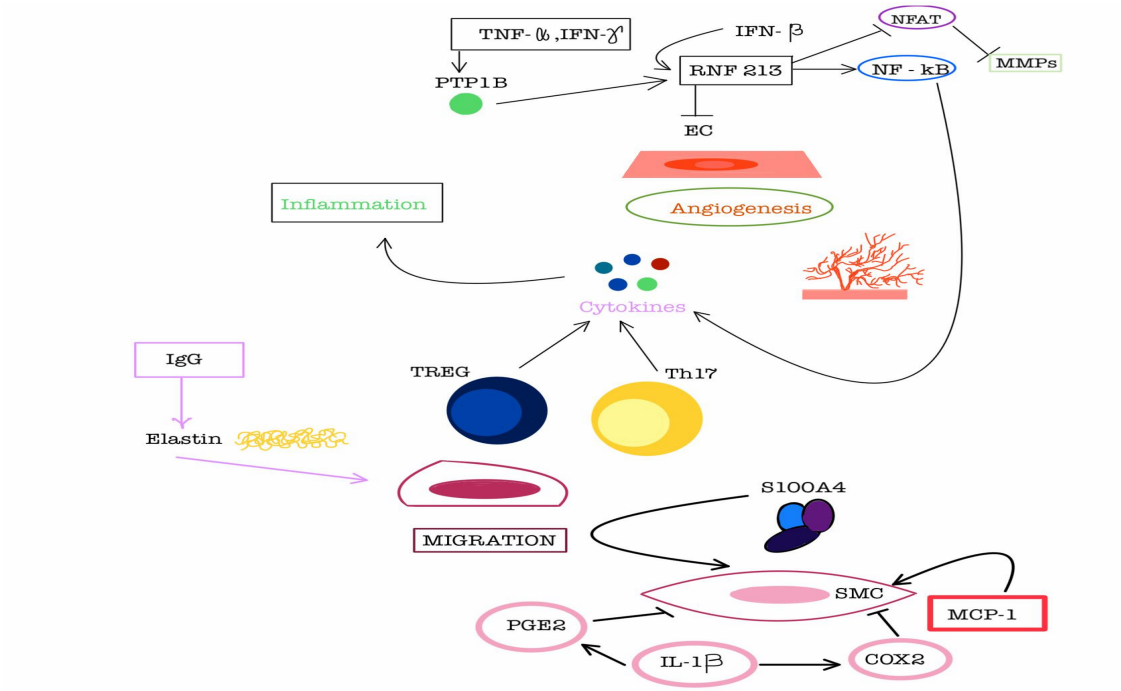

Figure 1: A mechanical review of inflammatory markers and their role in Moya Moya disease. COX2, cyclooxygenase 2; EC, endothelial cell; MMPs, matrix metalloproteinases; PGE2, prostaglandin E2; SMC, smooth muscle cell.

II. Case Presentation:

A 27-year-old homemaker was admitted to the medical department of Pt. BD Sharma University of Health Sciences, Rohtak exhibiting a two-day history of aphasia, dysphagia, and facial asymmetry. She complained of mild to moderate cephalalgia, vertigo, visual disturbances, and palpitations. Previously, she encountered a similar episode with hemiparesis on the left side, resulting in persistent gait and grip difficulties. Her medical record includes a third-trimester intrauterine demise (IUD) and hypertension with edema in both lower extremities during previous pregnancies, resolving postpartum. Upon evaluation, she was conscious but aphasic, displaying left facial nerve paralysis, tachycardia, hypertension (BP 160/90 mmHg), and a tender midline neck mass indicative of thyroid involvement. Neurological examination revealed significant motor impairment (left-sided paresis). Most cranial nerves appeared intact, except for left facial asymmetry. Laboratory findings showed elevated thyroid hormones (T3: 16.48 ng/dL, T4: 4.70 ng/dL, TSH: <0.01 µIU/mL) and increased anti-TPO antibodies (187.77 IU/mL), consistent with Graves' disease. Positive ANA with a nuclear speckled pattern and elevated APLA levels, including cardiolipin IgG (1.14 IU/mL and 13.36 IU/mL) and IgM (11.58 IU/mL and 32.82 IU/mL), along with anti-beta-2 glycoprotein I IgM (41.23 IU/mL and 4.33 IU/mL) and IgG (<1.80 IU/mL and 28.69 IU/mL) were noted. Lupus anticoagulant was not detected. Imaging studies revealed intimal thickening and severe stenosis in the left ICA and MCA, with collateral vessels suggestive of Moya Moya disease. The diagnosis and treatment of MMD in the presence of concurrent APLS and Graves' disease pose substantial challenges. The patient's manifestation of stroke-like symptoms and imaging findings of severe stenosis and collateral circulation indicated advanced MMD. Confirmation of coexisting APLS and Graves' disease through positive APLA profile and thyroid evaluations complicated therapeutic decision-making. Managing this patient necessitated a multidisciplinary approach, incorporating dual antiplatelet therapy for thrombotic risk reduction, beta blockers for thyroid-related cardiovascular strain, and thionamide for hyperthyroidism control. This comprehensive therapeutic strategy aimed to tackle the multifaceted aspects of the patient's condition. Long-term Outcome: A personalised treatment plan involving dual antiplatelet therapy, beta blockers, and thionamide demonstrated efficacy in maintaining the patient's stability, with no recurrent strokes observed during a two-year follow-up period. Regular monitoring and adjustments to the treatment regimen played a pivotal role in sustaining the patient's well-being and preventing further cerebrovascular incidents.

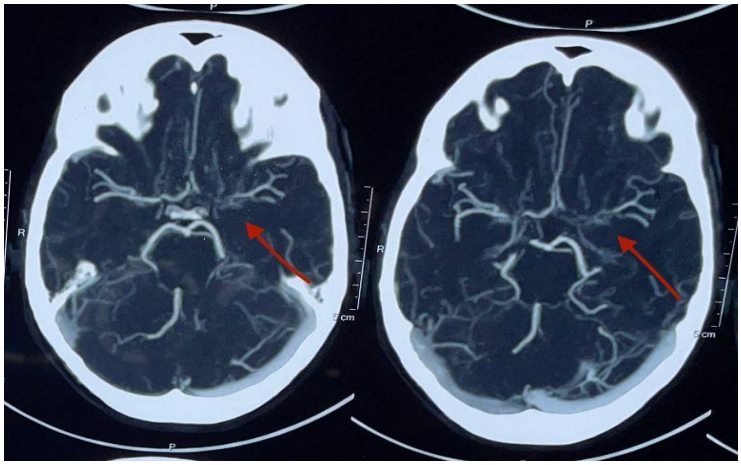

Figure 2 shows the Non visualisation of A1 segment of left ACA with severe luminal narrowing of the proximal M1 segment of left MCA with attenuated calibre of A2 segments of b/l ACA. Multiple lenticulostriate and leptomeningeal collateral vascular channels can also be seen.

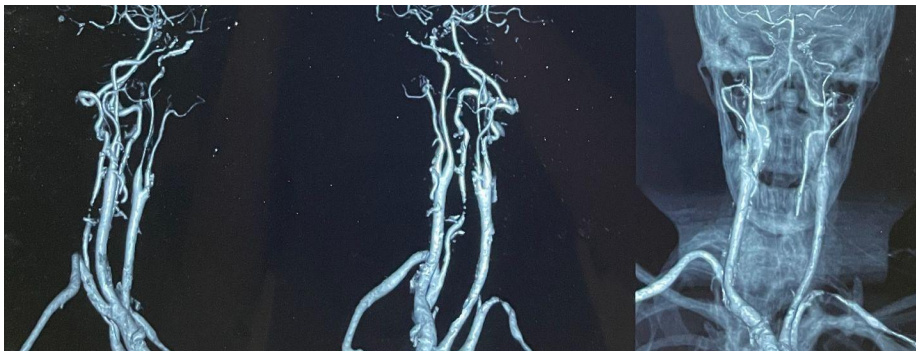

Figure 3: Intimal thickening with diffuse tubular narrowing of cervical and intracranial segments of left ICA with complete occlusion of clinoid and supraclinoid segment.

III. Conclusion:

Although APS often prompts the initiation of anticoagulant therapy, opting for antiplatelet medications instead of anticoagulation is advisable due to the potential risk of bleeding complications associated with the delicate vessels in moyamoya pathology(7). This example highlights the critical need for an integrated approach in addressing the various facets of MMD and its coexisting comorbidities to improve patient outcomes.

IV. Research And Future Directions:

Further research is warranted in order to enhance comprehension of the pathophysiological connections between MMD and autoimmune disorders. Such endeavours have the potential to result in the development of more precise treatment modalities and enhanced prognoses for individuals impacted by said conditions. The discourse underlines the intricate relationship between Moya Moya disease and autoimmune ailments, underscoring the necessity for an interdisciplinary strategy to the identification and treatment of these conditions.

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