

An Endocrine Perspective of Juvenile Moyamoya Syndrome/ Disease: A Literature Review

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Abstract

Moyamoya disease is a progressive rare type of cerebrovascular disease marked by stenosis and occlusion of the distal internal carotid arteries and circle of Willis branches, leading to the development of a collateral network of blood vessels at the base of the brain. For Moyamoya angiopathy, endocrinopathies may be risk factors, sequelae, or just coincidental findings. So, it is worth checking and regularly monitoring for these endocrinopathies in patients with Moyamoya angiopathy, aiming for early diagnosis and appropriate treatment.

Keywords: Adrenal, diabetes, endocrinopathy, growth hormone, hypogonadism, moyamoya, parathyroid, thyroid

INTRODUCTION

In spite of being Moyamoya (MM) cerebral angiopathy a rare condition, it is the most common pediatric cerebrovascular disease in some countries.^[1] It is characterized by progressive stenosis or occlusion of the intracranial internal carotid artery and/or the proximal portion of the anterior cerebral artery and middle cerebral artery. This steno-occlusive pattern is associated with a compensatory development of a collateral network of vessels at the base of the brain, appearing as a “puff of smoke” on conventional angiography (“Moyamoya” in Japanese). This angiopathy was first described in Japan in 1957^[2] and was called “MM disease” for the first time by Suzuki and Takaku in 1969.^[3]

MM syndrome is an MM angiopathy associated with an underlying condition (acquired or inherited), whereas patients with no underlying risk factors (idiopathic) are said to have MM disease.^[1,4] The MM disease etiology is unknown to date. There is an obvious familial tendency, in which there is a 6–12% risk of developing the disease if a person has a first-degree relative with MM disease.^[5,6] There is also an ethnic predilection for Asian populations, especially for people with Korean and Japanese ancestry.

The MM disease incidence in East Asia is about 10 times that of Western countries.^[7] Genetic linkage studies revealed putative chromosomal locations linked to MM disease.^[8–11] Apart from the genetic hypotheses, various environmental factors have been proposed as etiological factors of MM disease, including infectious agents, immunological responses with cellular components and autoantibodies, and hemodynamic stress to specific vascular loci.^[12–14] Whatever the cause, MM angiopathy increases the risk of ischemic and hemorrhagic brain damage.^[4]

MM angiopathy has two peaks: between 5 and 10 years of age and during the fourth decade,^[15–19] with a more rapid progressive course during childhood.^[20] Clinical presentation in adults may be due to ischemia or hemorrhage, whereas in children it is mainly due to ischemic events,^[18,21,22] which result in focal neurological signs, seizures, and progressive cognitive impairment.^[23]

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Other common presenting symptom is headache (migraine).^[4]

The anterior cerebral arteries and internal carotid arteries supply the hypothalamic-pituitary areas with blood; these arterial branches are close to the stenosed carotid fork in MM.^[24] Then through links to posterior cerebral and external cerebral artery branches, MM arteries provide collateral circulation to ischemic regions. The hypothalamus is perfused by reversed flow through these collaterals. Therefore, this defect can result in hypothalamic vascular insufficiency, leading to hypothalamic and pituitary dysfunction.^[24] In contrast, endocrinopathies may be a risk factor for MM angiopathy.^[25] This review will highlight the endocrinopathies that had been reported in conjunction with MM disease/syndrome, aiming to achieve optimal health through early diagnosis and treatment.

MATERIALS AND METHODS

We searched the PubMed database using the following keywords: “MM growth failure,” “MM growth hormone deficiency,” “MM hypopituitarism,” “MM precocious puberty,” “MM hypogonadism,” “MM adrenal,” “MM diabetes insipidus,” “MM diabetes mellitus children,” “MM hypoparathyroidism,” “MM hyperparathyroidism,” “MM thyroid children,” “MM hypothyroidism children,” and “MM Gravis disease children” (last search 17 September 2022). We took into account the articles analyzing patients ≤ 18 years.

The main reported MM-associated endocrinopathies are as follows.

1. Growth hormone (GH) deficiency

MM disease can be associated with GH deficiency, which could be caused by chronic cerebrovascular insufficiency.^[26]

In 1990, the first case report of GH deficiency with MM disease was published, in which a 7-year-old boy with hypopituitarism presented with short stature. Then he had his first generalized convulsion 6 months after starting GH replacement therapy, which warranted further neuroradiological investigations to reveal the diagnosis of MM disease.^[27]

In 1999, Mootha *et al.* reported two cases of MM disease in association with GH deficiency. One of them was diagnosed with MM disease at the age of 17 months. After that, her growth was deteriorated and central hypothyroidism with GH deficiency was the diagnosis at the age of 9 years. An appropriate increase in growth velocity was achieved with hormonal replacement therapy (thyroxine and GH). The second one was a 6-year-old male, presented with short stature [height of -3 standard deviation score (SDS)], with a bone age of 4.5 years. He failed two GH provocation tests; therefore, it was decided to start him on GH therapy. However, he developed

transient left hemiparesis before starting GH therapy for which a brain magnetic resonance imaging (MRI) study was conducted revealing the ischemic changes in white matter of both hemispheres, with the right one showing more alterations. The magnetic resonance angiogram showed bilateral anterior and middle cerebral arteries narrowing, MM disease was recognized, and the child underwent a right-side revascularization operation. GH replacement was started a year later. A recurrence of neurological symptoms was reported for which he underwent a revascularization surgery on the left side, and GH continued. The patient remained stable after those surgeries, and no side effects were reported from the GH therapy. He had exhibited an appropriate rise in his growth velocity.^[24]

Another case was reported by Kalina *et al.*^[26] in 2004. A 16-year-old boy was referred because of headaches and short stature. The height was -4.3 SDS; and the bone age was 11.5 years. No focal neurological deficits were observed. Isolated GH deficiency was diagnosed on the basis of baseline and stimulated GH measurement. On brain MRI, a malformation of the cerebral vessels was suspected, and angio-computed tomography and pan-angiography revealed an image suggestive of MM disease. Daily doses of GH were provided at a dose of 0.025 mg/kg/day. The first-year growth velocity was 12 cm per year and no side effects have been reported as a result of the treatment.

Parker *et al.*,^[28] in 2009, reported a 15-year-old male known to have thalassemia since 15 months; he was assessed for short stature and found to have GH deficiency for which he was started on GH therapy. MRI was performed in view of GH deficiency and stenotic arteries were found suggestive of MM syndrome.

Abdullah and Alabedi,^[29] in 2022, reported two brothers with MM angiopathy and GH deficiency. The diagnosis of MM angiopathy for those brothers was made at the ages of 11 and 8.5 years when they presented with neurologic manifestations, and revascularization surgery was done for both of them. The diagnosis GH deficiency was made later (at ages of 13 and 10 years, respectively) as their growth was slowed. The brothers showed a good response to GH therapy (about 4 cm per 6 months).

Growth failure was also reported in a number of families with BRCC3-related MM and majority of the affected members had partial GH deficiency.^[30-32]

According to this review, GH deficiency is common in MM angiopathy, and their presentations may occur at the same time, or one precedes the other.

2. Precocious puberty

Precocious puberty is uncommon in MM disease. Kazumata *et al.*^[33] reported the first case of precocious puberty associated with MM disease in 1996, in which an 8-year-old girl presented with menarche, advanced breast

development (Tanner's stage III), and presence of pubic hair, on a medical background history of pustular psoriasis. Her pubertal precocity started at the age of 7, which led to advancing in her bone age (11 years) at presentation. A normal neurological examination was reported, but a brain MRI revealed numerous flow voids in the area of basal ganglia on both sides, indicating the existence of MM disease. The proximal segment of the anterior and middle cerebral arteries and the distal segment of the internal carotid arteries were all stenosed on cerebral angiograms. In the base of the brain, a network system of collateral vessels (MM vessels) was also found, confirming the presence of MM disease. The precocious puberty was treated medically. Kazumata *et al.* suggested that ischemic changes or a little destructive damage of the hypothalamus caused by dilated perforators was the cause of this patient's early puberty.

3. Hypogonadism

BRCC3-related MM angiopathy had been reported in the literature with a syndromic presentation including hypergonadotropic hypogonadism and partial GH deficiency.^[30-32]

In 2022, another unique association was described: a 14-year-old male who was surgically treated for cryptorchidism at a younger age suffered from recurrent seizures and decline cognitive skills, presented with transient bilateral hemiparesis, found to have MM angiopathy. Further testing revealed Klinefelter syndrome.^[34]

No case of hypogonadotropic hypogonadism had been reported in association with MM angiopathy in patients 18 years old or younger, whereas one was mentioned for a 34-year-old man with empty sella syndrome.^[35]

4. Hypothyroidism

Mootha *et al.*^[24] in 1999 reported a Caucasian female who had MM, central hypothyroidism, and GH deficiency.

5. Hyperthyroidism

In several studies assessing the risk factors for MM disease progression, Graves' disease is a well-known medical disorder that has been associated with a rapid progression of MM disease.^[36,37] Previous research had demonstrated that vasculopathy can be prevented by suppressing thyrotoxicosis,^[38] as the clinical progression of MM disease patients with concurrent Graves' disease is quicker than that of patients without Graves' disease.^[39] Appropriate antithyroid medication can reduce the risk of ischemic episodes or infarctions by reducing the hemodynamic load on the brain.^[37] The association between MM and hyperthyroidism was thought to be attributed to increased metabolism and oxygen consumption in the brain, which are harmful to arterial walls, sympathetic nervous activation that may cause cerebral arteries stenosis, and cellular proliferation and vascular dysregulation due to T-cell dysfunction in autoimmune thyroid disease.^[40-42] It

was postulated that in MM disease with hyperthyroidism, the need for revascularization surgery in addition to antithyroid therapy depends on the mechanisms of the vascular changes. So, antithyroid therapy is sufficient if hyperthyroidism is the cause of vascular changes and symptoms. However, if immune-mediated mechanisms are considered to be the cause of vascular changes and symptoms, surgical revascularization might be needed.^[43]

In 2005, Golomb *et al.*^[44] reported a 10-year prepubertal girl with bilateral MM angiopathy and Graves' disease diagnosed after the sudden onset of left face, arm, and leg weakness. There was a history of easy fatigability, warm feeling, loose stools, and throbbing headache in the previous 2 years. Thyroid gland examination showed 4 and 3 cm diameter right and left lobes, respectively. Thyroid radioablation was the modality of therapy used in this patient. Hypothyroidism developed 6 weeks later and levothyroxine was started. MM was treated by cranial revascularization followed by daily aspirin and verapamil.

Another case was reported in 2014 when a 12-year-old girl with Graves' disease started to develop neurological symptoms (deterioration in physical and school performances, seizure, and speech difficulty). MM angiopathy was found in brain MRI. Graves' disease was treated by subtotal thyroidectomy as it was not controlled with antithyroid medication. MM angiopathy was treated by shunt operation between the left superficial temporal artery and middle cerebral artery 2 months after thyroidectomy.^[45]

In 2011, a case-control study that involved 114 patients with MM disease aged <16 years demonstrated a significant association of MM disease with hyperthyroidism (overt and subclinical) and increased thyroid antibodies (antithyroperoxidase or antithyroglobulin), whereas there was no such association with hypothyroidism (overt and subclinical).^[46]

A retrospective study published in 2017 that included 37 patients (1–67 years old) with MM disease found that elevated thyroid antibody (not thyroid function) was the only variable that was significantly associated with an aggressive presentation of MM disease (hemorrhage, major stroke, or frequent transient ischemic attacks), which supports the hypothesis that MM pathophysiology is related to immune-mediated processes.^[47]

6. Hypoparathyroidism and hyperparathyroidism

While there was no reported case of hypoparathyroidism in association with MM angiopathy, there was one adult case for hyperparathyroidism (60-year-old male) with neurofibromatosis type 1.^[48]

7. Adrenal disease

There were only three reported adult cases. Two of them had congenital adrenal hyperplasia and the third one

had pheochromocytoma as a part of multiple endocrine neoplasia type 2A complicated by MM syndrome.^[49-51]

8. Diabetes insipidus

In 2010, Shibata *et al.*^[52] reported a 15-year-old girl with central diabetes insipidus and her radiologic examination revealed neuroepiphyseal germinoma and MM disease that represent a rare incidental combination.

Another case of diabetes insipidus was described in 2017 for an 18-year-old female with MM disease who underwent an external carotid to internal carotid bypass. Diabetes insipidus was related to ketamine infusion that was used during the procedure, not to MM disease itself.^[53]

In conclusion, when MM patient develops diabetes insipidus another cause must be searched.

9. Diabetes mellitus

The association between MM angiopathy and diabetes mellitus in adults was documented in many studies.^[54-60] For children, in 2005, Krebs *et al.*^[61] reported MM disease in a 12-year-old boy who had poorly controlled type 1 diabetes mellitus for 10 years. The authors considered their concurrence a coincidental, and the MM angiopathy may be reinforced by the underlying accelerated atherosclerosis. Concurrence of MM angiopathy with diabetes mellitus/insulin resistance had also been described in microcephalic osteodysplastic primordial dwarfism type II, an autosomal recessive inherited disease.^[62]

CONCLUSION

For MM angiopathy, endocrinopathies may be risk factors, sequelae, or just coincidental findings. So, it is worth checking and regularly monitoring for these endocrinopathies in patients with MM angiopathy aimed for early diagnosis and appropriate treatment.

Ethical consideration

Not applicable.

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Conflicts of interest

There are no conflicts of interest.

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