Marfan syndrome

Marfan syndrome can demonstrate tortuous and elongated intracranial artery. However, these arteries rarely cause neurovascular compression resulting in hemifacial spasm or trigeminal neuralgia [1].

Complications

The underlying cause of spontaneous intracranial hypotension is usually a spontaneous cerebrospinal fluid fistula, however, some cases may be due to low cerebrospinal fluid volume. Evidence supports an underlying weakness of the meninges as a contributing factor; for instance, connective tissue disorders like Marfan syndrome, and Ehlers-Danlos syndrome.

Case reports

Marques et al. report a five-generation kindred with two brothers with pituitary gigantism due to AIP mutation-positive GH secreting pituitary neuroendocrine tumors and their first-cousin coincidently also having gigantism due to Marfan syndrome [2].

A 33-year-old woman who was diagnosed as Marfan syndrome, suffered from trigeminal neuralgia. Magnetic resonance (MR) angiography showed tortuous and elongated left vertebral artery (VA). The coronal section of three dimensional (3D) MR cisternography with contrast enhancement showed that the left trigeminal nerve was compressed from underneath by the tortuous and elongated left VA. After successful surgery of microvascular decompression, the patient's symptom resolved and no recurrence was encountered.

Neurosurgeons should not only be aware of hemifacial spasm but also of trigeminal neuralgia caused by elongated vessels in a patient with Marfan syndrome, although it is an extremely rare condition. In addition, offending vessel is not atherosclerotic in younger patients unlike usual cases of trigeminal neuralgia. Thus, microvascular decompression can be easier than usual cases [3].

[1], [3]
Sakakura K, Akutsu H, Yamamoto T, Masuda Y, Ishikawa E, Matsumura A. Trigeminal Neuralgia in a Patient with Marfan Syndrome: Case Report. Neurol Med Chir (Tokyo). 2013 Dec 27. [Epub ahead of print] PubMed PMID: 24390183.

[2]
Marques P, Collier D, Barkan A, Korbonits M. Coexisting pituitary and non-pituitary gigantism in the same family. Clin Endocrinol (Oxf). 2018 Sep 17. doi: 10.1111/cen.13852. [Epub ahead of print] PubMed PMID: 30223298.