Sclerosteosis

Sclerosteosis is a rare bone disorder characterized by a progressive craniotubular hyperostosis. The diagnosis of sclerosteosis is based on characteristic clinical and radiographic features and a family history consistent with autosomal recessive inheritance. The skull overgrowth may lead to lethal elevation of intracranial pressure, distortion of the face, and entrapment of cranial nerves, resulting in recurrent facial palsy or secondary trigeminal neuralgia.

Treatment

The treatment of the trigeminal neuralgia secondary to hyperostosis and resistant to medications presents a dilemma. The narrowing of the foramen oval and difficulty in the identifying and approaching of the foramen makes the percutaneous technique a challenge for the neurosurgeon in patients harboring sclerosteosis. Microvascular decompression should not be considered since the primary cause of the trigeminal neuralgia is the nerve entrapment by the narrowing of neurovascular foramina and not the neurovascular conflict related to essential trigeminal neuralgia. Stereotactic radiosurgery may be a good treatment option, but there is a lack of published data supporting the use of this method in cranial hyperostosis ¹⁾.

1)

de Andrade EM, Beer-Furlan A, Duarte KP, Fonoff ET, Teixeira MJ. Management of trigeminal neuralgia in sclerosteosis. Surg Neurol Int. 2013 Nov 20;4(Suppl 6):S455-9. doi: 10.4103/2152-7806.121644. PubMed PMID: 24349870; PubMed Central PMCID: PMC3858807.

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