Klippel Trenaunay Syndrome

Klippel-Trenaunay-Weber syndrome (KTWS) involves a triad of conditions, including cutaneous hemangiomas, venous varicosities, and osseous and soft-tissue hypertrophy of the affected limb.

The English-language literature contains six reports of associations between KTWS and spinal cord cavernous angioma (CA), but the management of these patients has not been well described. [1)]

Case reports

2017

Yoshinaga et al. describe a rare case of multiple cavernomas in the central nervous system in a patient with KTWS. A-64-year-old man with KTWS and a previous brain hemorrhage presented with sudden onset of gait and vesicorectal disturbance. The magnetic resonance imaging scan showed a cavernoma associated with hemorrhage in his lumbosacral spinal cord. Moreover, numerous cavernomas were identified in his brain. He was treated conservatively with no adverse events. Gait disturbance improved, but vesicorectal disturbance did not improve [2)].

A extremely rare case of a giant P1-P2 partially thrombosed aneurysm associated with bilateral ICA occlusion in a Klippel-Trenaunay syndrome patient [3)].

A 23-year-old woman was admitted because of acute onset of leg weakness accompanied by upper back pain. Thoracic magnetic resonance imaging showed a heterogeneous mass with a slit component at the T1/2 level of the spinal cord. After the patient underwent left hemilaminectomy followed by removal of the tumor, her neurological symptoms improved. Pathological examination showed that the spinal lesion was characterized by hemosiderin deposition and thin-walled vascular channels surrounded by fibrous tissue. This is the first report to provide a detailed pathological description of the features of a spinal CA in a patient with KTWS. The discussion includes an assessment of the clinical features and management of CA associated with KTWS. Because this syndrome is a rare disease entity, further experience in the treatment of these patients is needed. However, considering that the pathological findings of spinal CA in patients with KTWS include the typical features of CA, the management of CA in patients with KTWS may be identical to that of isolated CA [4)].

The first and unique case of a rapid-growing skull hemangioma in a patient with Klippel-Trénaunay-Weber syndrome. This case report provides evidence that not all rapid-growing, osteolytic skull lesions need to have a malignant character but certainly need a histopathological verification. This material offers insight into the list of rare pathological diagnoses in an infrequent syndrome [5)].

[1)], [4)]
Oda K, Morimoto D, Kim K, Yui K, Kitamura T, Morita A. Spinal cavernous angioma associated with

Klippel-Trenaunay-Weber syndrome: a case report and literature review. World Neurosurg. 2017 Oct 17. pii: S1878-8750(17)31767-9. doi: 10.1016/j.wneu.2017.10.040. [Epub ahead of print] PubMed PMID: 29054771.

2)

Yoshinaga T, Yagi K, Morishita T, Abe H, Nonaka M, Inoue T. Cerebral and spinal cavernomas associated with Klippel-Trenaunay syndrome: case report and literature review. Acta Neurochir (Wien). 2017 Dec 1. doi: 10.1007/s00701-017-3408-5. [Epub ahead of print] PubMed PMID: 29197023.

3)

Fukaya R, Yanagisawa K, Fukuchi M, Fujii K. Posterior cerebral artery giant aneurysm associated with bilateral internal carotid artery occlusion in a Klippel-Trenaunay syndrome patient: a case report. Br J Neurosurg. 2017 Oct 26:1-3. doi: 10.1080/02688697.2017.1394446. [Epub ahead of print] PubMed PMID: 29069941.

5)

van der Loo LE, Beckervordersandforth J, Colon AJ, Schijns OE. Growing skull hemangioma: first and unique description in a patient with Klippel-Trénaunay-Weber syndrome. Acta Neurochir (Wien). 2017 Feb;159(2):397-400. doi: 10.1007/s00701-016-3012-0. Epub 2016 Nov 7. PubMed PMID: 27822901; PubMed Central PMCID: PMC5241322.