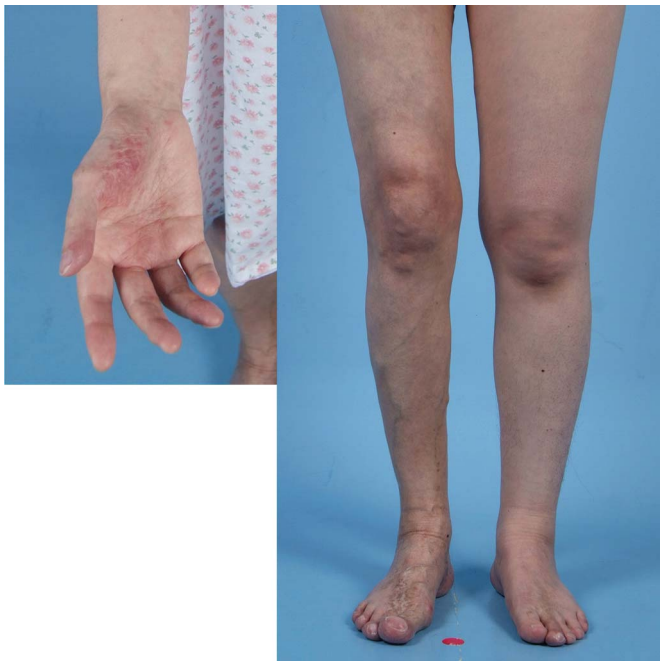
Teaching Neuro*Images*: Multiple giant intracranial aneurysms in Klippel-Trenaunay syndrome

Yong Woo Kim, MD
Namju Kim, MD
Jeong-Min Hwang, MD
Ho-Kyung Choung, MD
Sang In Khwarg, MD

Correspondence to Dr. Kim: resourceful@hanmail.net

Figure 1 Clinical features of Klippel-Trenaunay syndrome



Cutaneous capillary malformation on the right hand, hemihypertrophy of the left lower extremity, bony and soft tissue hypertrophy of right toe, and varicose vein on right leg confirmed the diagnosis of Klippel-Trenaunay syndrome.

A 40-year-old woman presented with horizontal diplopia and right proptosis for 1 month. She had esotropia and right abducens nerve palsy. Pupils were small and equal and there was no relative afferent pupillary defect. A cutaneous capillary malformation, hypertrophy of bone and soft tissue, and varicose veins on lower extremities were consistent with the diagnostic triad of Klippel-Trenaunay syndrome (KTS) (figure 1).[1] Orbital CT revealed multiple giant intracranial aneurysms (figure 2).

The mesodermal abnormality in KTS is due to somatic mutation, especially in hemangioblastic, lymphoblastic, and osteoblastic processes during embryogenesis.[2] Vascular malformations combined with KTS usually originate from slow flow systems, such as capillary, lymphatic, and venous systems.[1] However, arterial aneurysms are reported in the literature, though only 8 cases of intracranial aneurysms have been reported so far.[2] Our patient had mild ophthalmic symptoms associated with potentially life-threatening multiple giant intracranial aneurysms, unlike previous cases.

The present case emphasizes the importance of prompt cerebrovascular imaging in patients with KTS with neurologic or ophthalmic symptoms.

AUTHOR CONTRIBUTIONS

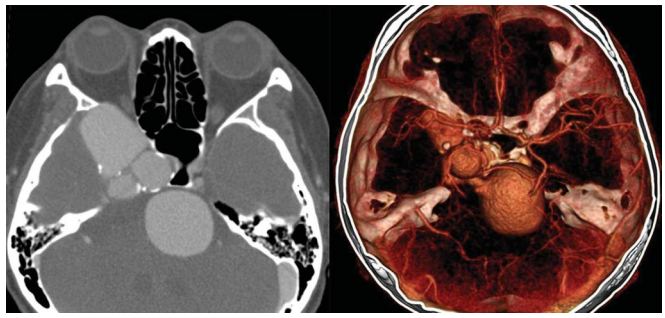
Yong Woo Kim, MD: data collection, interpretation of data, drafting and revision of the manuscript. Namju Kim, MD: data collection, interpretation of data, drafting and revision of the manuscript. Jeong-Min Hwang, MD: interpretation of data, drafting and revision of the manuscript. Ho-Kyung Choung, MD: interpretation of data, drafting and revision of the manuscript. Sang In Khwarg, MD: interpretation of data, drafting and revision of the manuscript.

Download teaching slides: www.neurology.org

From the Department of Ophthalmology (Y.W.K., S.I.K.), Seoul National University College of Medicine, Seoul National University Hospital, Seoul; the Department of Ophthalmology (N.K., J.-M.H.), Seoul National University College of Medicine, Seoul National University Bundang Hospital, Seongnam; and the Department of Ophthalmology (H.-K.C.), Seoul National University College of Medicine, SMG-SNU Boramae Medical Center, Seoul, Korea.

Go to Neurology.org for full disclosures. Funding information and disclosures deemed relevant by the authors, if any, are provided at the end of the article.

Figure 2 Orbital CT images



Orbital CT demonstrates multiple giant fusiform intracranial aneurysms, maximal diameter of 34 mm, from bilateral internal carotid artery, basilar artery, and right posterior cerebral artery.

STUDY FUNDING

No targeted funding reported.

DISCLOSURE

The authors report no disclosures relevant to the manuscript. Go to Neurology.org for full disclosures.

REFERENCES

1. Cohen MM, Jr. Klippel-Trenaunay syndrome. Am J Med Genet 2000;93:171–175.
2. Star A, Fuller CE, Landas SK. Intracranial aneurysms in Klippel-Trenaunay/Weber syndromes: case report. Neurosurgery 2010; 66:E1027–E1028; discussion E8.

Teaching Neuro*Images*: Multiple giant intracranial aneurysms in Klippel-Trenaunay syndrome

Yong Woo Kim, Namju Kim, Jeong-Min Hwang, et al.

Neurology 2013;81;e17-e18
DOI 10.1212/WNL.0b013e31829bfd4c