Klippel Trenaunay Syndrome With Occipital Infarct

ÖMER İYİGÜN, ZEKİ ŞEKERÇİ, CEMİL RAKUNT, FAHrettİN ÇELİK

Department of Neurosurgery (Ö.I, Z.S, C.R, F.C) Ondokuz Mayis University Samsun, Türkiye

Abstract: Klippel-Trenaunay syndrome is a congenital angiodysplasia characterized by varicose veins, cutaneous haemangiomas and bony and soft tissue hypertrophy. This report describes a case of Klippel-Trenaunay Syndrome with dilated tortuous vessels in the retina and left occipital infarct.

Key words: Cerebral infarct, Klippel-Trenaunay Syndrome, Retinal Involvement.

INTRODUCTION

The Klippel-Trenaunay Syndrome is a congenital angiodysplasia characterized by a vascular nevus, varicose veins and bony and soft tissue hypertrophy as well as associated anomalies such as pes equinovarus, syndactyly, polydactyly, congenital dislocation of hips or shoulders, spina bifida, scoliosis or pelvic asymmetry.

In 1900, Klippel and Trenaunay (5) described this syndrome for the first time. Parkes Weber (10) described a similar triad of findings. Arteriovenous fistulas, vascular hyperplasia and bony hypertrophy occur in both syndromes. In Klippel-Trenaunay syndrome, the fistulas are small and numerous but in Parkes Weber syndrome they are large and few and may lead to circulatory disturbances. All these symptoms may become apparent at any time from birth to adulthood (12). Osteohypertrophy is frequently present at birth usually affecting the limbs. The congenital haemangioma varies in size and colour, frequently following a radicular distribution although the varices are congenital but may increase in size (1).

The etiology of Klippel-Trenaunay syndrome is unknown. It does not appear to be hereditary nor is there sex preference (2).

In this report we present and unusual case of Klippel-Trenaunay Syndrome with occipital infarct.

CASE REPORT

A 55-year-old female presented to our neurosurgical centre with a 15-day history of mild headache. There was an associated history of enlarged left arm and leg and accompanying erythematous lesion over the face and neck. There was no history of convulsions, vomiting, loss of consciousness or focal neurological deficit. Her family had noticed the erythematous lesions on her face and left upper and lower limbs which increased in size in proportion to her general growth. Her left hand and foot, however, became disproportionately large. She has four siblings none of whom is affected.

Physical examination revealed that there was evidence of portwine on her face and over the left forearm and left leg (Fig1). The left hand and foot were disproportionately large (Fig 2a-2b). The rest of the systemic examination did not reveal any obvious abnormality. Total and differential blood counts including platelet count were within normal limits.

Femoral angiography was proposed but the patient refused.

Ocular examination revealed a visual acuity of 20/20 in both eyes. The conjunctiva and iris did not reveal any angioma or other disorder. On fundus examination of the right eye, the optic disc was hyperaemic and minimally elevated in appearance. The retinal veins were markedly dilated and tortuous...
The clinical syndrome named after these investigators, includes haemangiomas, hypertrophy of the soft tissue and bone with overgrowth of the extremity and varicose veins. The vascular lesion in Klippel-Trenaunay syndrome is one of deep venous medicine.

**DISCUSSION**

The Klippel-Trenaunay syndrome is a related condition consisting of a triad of cutaneous haemangioma extending over the limbs, varicosities of the affected limbs and soft tissue and bony hypertrophy (4,6). In 1900 Klippel and Trenaunay (5) published an article entitled “Du nevus variqueux ostéohypertrophique” in archives Generales de...
Fig. 5: Cranial CT showed a hypodense well-circumscribed lesion at the left occipital region.

abnormality with insufficiency (9). Parker Weber (10) described a similar triad in 1907. In a further report in 1918 (11), he included arteriovenous fistulas as part of the syndrome. Since then the names Klippel-Trenaunay and Klippel-Trenaunay Weber have been used interchangeably and indiscriminately: Klippel-Trenaunay syndrome for patients with no arteriovenous fistula and Klippel-Trenaunay-Weber syndrome for those with a clinically apparent fistula popularized by Lindenauer in 1965 (7).

Neurovascular involvement in the Klippel-Trenaunay-Weber syndrome is very rare. Djindjian et al. (3) described the occurrence of spinal arteriovenous malformations in five patients with the syndrome.

In 1988, Oyesiku et al. described a true cerebral arteriovenous fistula in the Klippel-Trenaunay-Weber Syndrome. This is the first case to be reported (8).

A higher incidence of neurovascular anomalies in the Klippel-Trenaunay-Weber syndrome may become evident using cranial CT scan.

The occipital infarct probably due to an undiagnosed micro haemangioma of the occipital brain tissue.

If a case of Klippel-Trenaunay Syndrome is encountered angiography of cerebral, spinal and four extremities routine diagnostic tests of haematologic disorders must be performed.

Correspondence: Ömer Iyigün, M.D.
Ondokuz Mayıs Üniversitesi
Tıp Fakültesi
Nöroşirurji Anabilim Dali
Samsun TÜRKİYE

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