Klippel Trenaunay Syndrome With Occipital Infarct

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Abstract : Klippel - Trenaunay syndrome is a congenital angiodysplasia characterized by varicose veins, cutaeous 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 Sydrome, 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 bifidia, scollosis 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 hypertropy occur in both syndromes. In Klippel - Trenaunay syndrome, the fistulas are small and numerous but in Parkes Weber sydrome they are large and few and may lead to circulatory disturbances. All these symptoms may become apperent 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 Syndrom with occipital infarct

CASE REPORT

A 55-year-old female presented of 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.

Physial 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 conjuctiva and iris did not reveal any angioma or other disorder.On fundus examination of the right eye, the optic disc was hyperaemic and minimaly elevated in appearance. The retinal veins were markedly dilated and tortuous

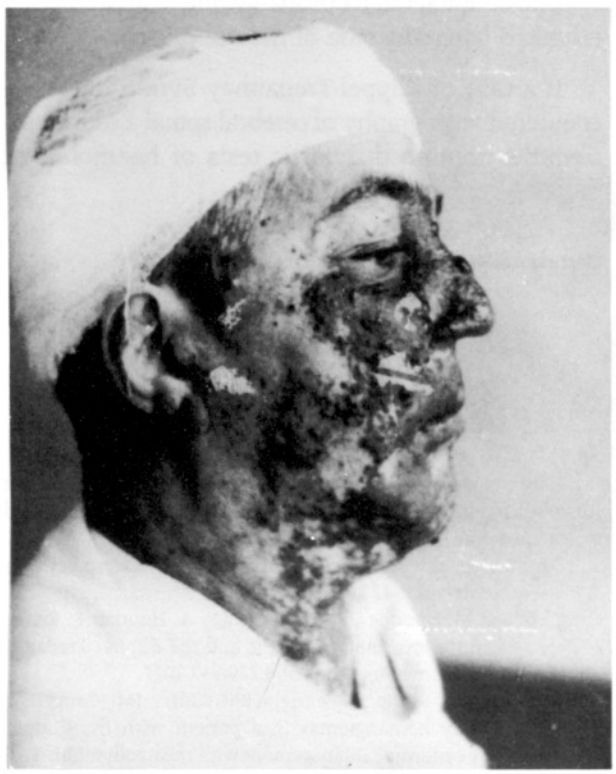

Fig 1 : Massive erythematous lesions are seen on the face and neck.

over the disc. No definitive choroidal angioma was visualized. Fluorescent angiography confirmed the presence of dilated, tortuous vessels in the right eye but no leakage of dye could be exhibited (Fig 3-4).

Cranial CT showed a hypodense, well circumscribed lesion with no contrast enhancement at the left occipital region (Fig 5) which was thought to be a low grade astrocytoma or infarct.

Occipital craniotomy was undertaken in june 1990. Immediately beneath the dura the lesion was seen over the occipital pole. The lesion was removed subtotally. The operative course was uneventful. Histologic a diagnosis proved to be typical necrotic material.

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 nevaus variqueux osteohypertrophique" in archives Generales de

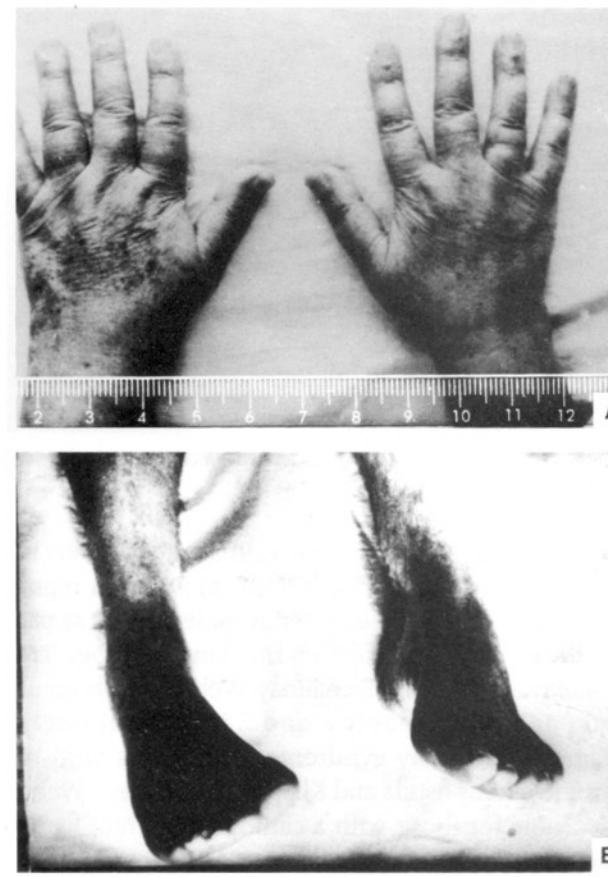

Fig. 2a - 2b : Minimal erythematous lesion and hypertrophy of left hand and leg of a 55-year-old woman with klippel-Trenaunay syndrome

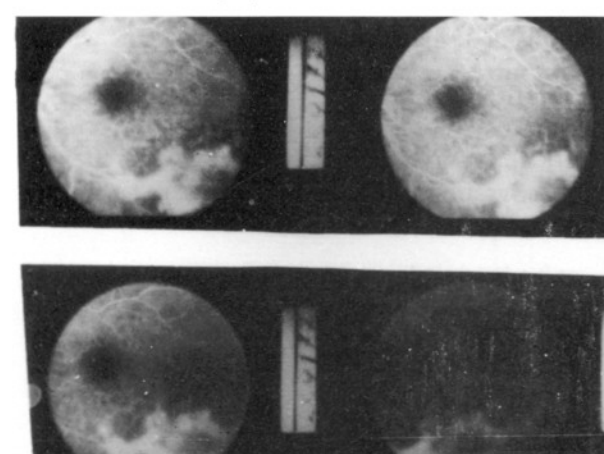

Fig. 3 : Fluoresent angiography showed the presence of dilated, tortuous vessels in right eye but no leakage of dye.

medicine. The clinical sydrome named after these investigators, includes haemangiomas, hypertrofy 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

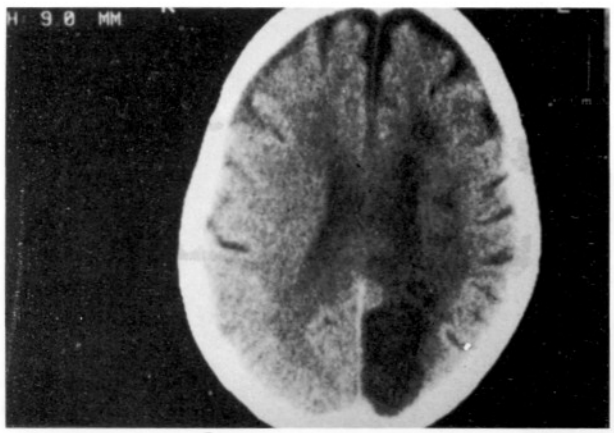

Fig. 5 : Cranial CT showed a hypodense well-circumscribed lesion at the left occipital region.

abnormality with insufficieny (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 sydrome for those with a clinically apparent fistula popularized by lindenauer in 1965 (7)

Neurovascular involvement in yhe Klippel-Trenaunay Weber syndrome is very rare. Djindjian at al (3) described the occurence 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 sydrome may became evident using cranial CT scan.

The occipital infarct probably due to an undisclosed 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 haemotologic disorders must be performed.

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