

Cerebral haemorrhage in arteriovenous malformation associated with Klippel-Trenaunay syndrome

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Summary. The computed tomography, magnetic resonance imaging and angiographic findings are described in a patient with Klippel-Trenaunay syndrome, who also had a cerebral haemorrhage from an arteriovenous malformation. The resulting aphasia disappeared completely after resorption of the haemorrhage. In this syndrome, the occurrence of a cerebral angioma has not previously been mentioned in the literature.

Key words: Spontaneous intracerebral haemorrhage – Arteriovenous malformation – Klippel-Trenaunay syndrome – Neurocutaneous angiomatosis

Introduction

The Klippel-Trenaunay syndrome (KTS) consists of soft tissue and bony hypertrophy, usually unilateral, involving the lower extremities, ipsilateral cutaneous port wine haemangiomas and vascular naevi. The KTS is associated with venous anomalies [6] and the related Parkes-Weber syndrome of arteriovenous angiomas [13]; in addition lymphangiomas may be found [8]. These are different manifestations of this disease [2]. Usually all the signs are present at birth [10]. In our case cerebral haemorrhage revealed the underlying arteriovenous malformation.

Case report

A 34-year-old man, whose right leg was larger than his left leg at birth, suddenly suffered for half a day from visual loss and confusion. Five days later, he had a speech disorder and was brought to the hospital. The patient's right leg was longer and more voluminous than the left and displayed superficial lymphohaemangiomas, thromboses, and oedema, as well as onychogryphosis (Fig. 1). The patient was anxious and excited; he had a mild receptive aphasia and his right tendon reflexes had increased, but there was no paresis.

Computed tomography (CT) and magnetic resonance imaging showed a left cerebral haematoma, which extended from the basal temporal lobe to the third ventricle and displaced the adjacent structures. The haematoma was associated with a widespread vascular malformation extending into

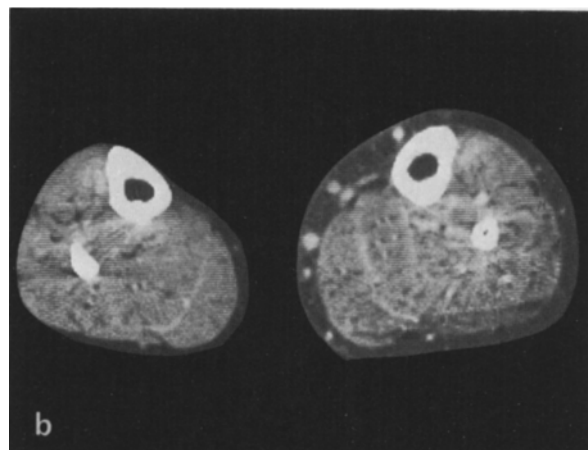
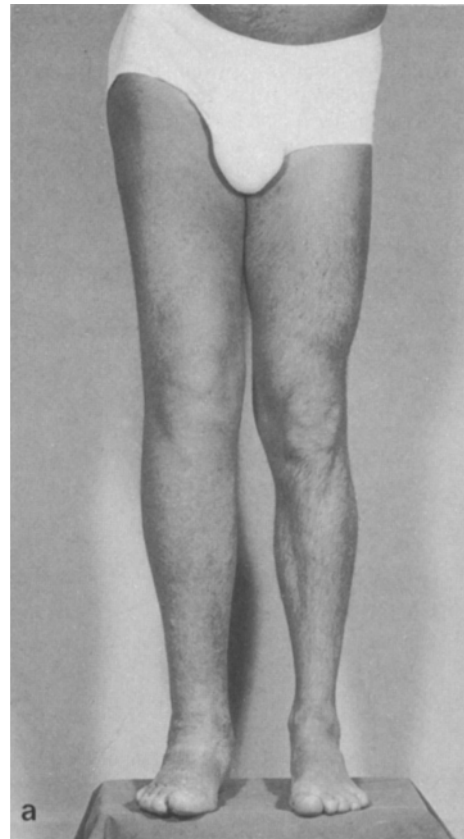


Fig. 1. **a** Hypertrophy of the right leg. **b** Transverse CT scan of the right calf, showing soft tissue hypertrophy and contrast enhancement of enlarged superficial veins

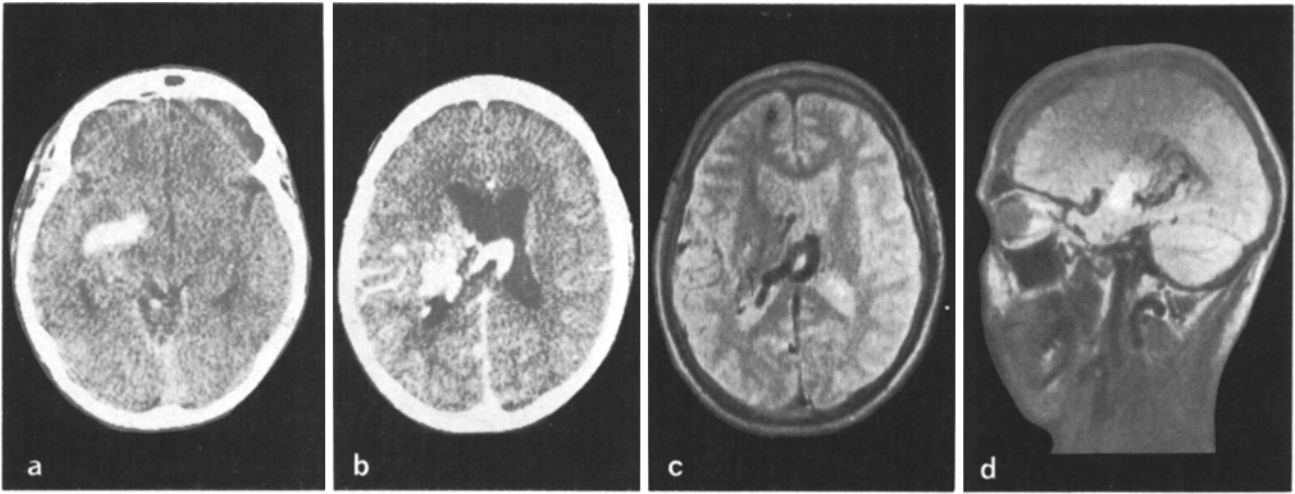


Fig. 2. **a** CT scan without contrast, demonstrating the temporal haematoma plus anomalous vascular structures and sinus drainage. **b** CT scan with contrast enhancement of the vascular malformation. **c** Dark-pictured vascular malformation in magnetic resonance scan. **d** Left parasagittal magnetic imaging scan illustrating the temporal haematoma and the adjoining vascular malformation

the lateral ventricles (Fig.2). Angiography demonstrated a large arteriovenous angioma in the left basal ganglia, the paraventricular white matter and the left brain stem. The angioma was partially fed by enlarged lenticulostriate arteries from the left middle cerebral artery and was also supplied by the left vertebral artery via the superior cerebellar artery and thalamic perforating arteries (Fig. 3).

Owing to the location and extension of the haemorrhage and the arteriovenous angioma, surgical evacuation and resection were not attempted. After treatment with dexamethasone

the neurological disturbances resolved. Three weeks later, CT scans indicated that the haematoma had turned into a hypodense defect. The patient felt well and went directly back to work.

Discussion

The clinical features of the KTS include different skin disorders, bone deformities and angiectasias, as well as in some cases mental deficiency and epileptic seizures [1, 2, 7, 10]. In

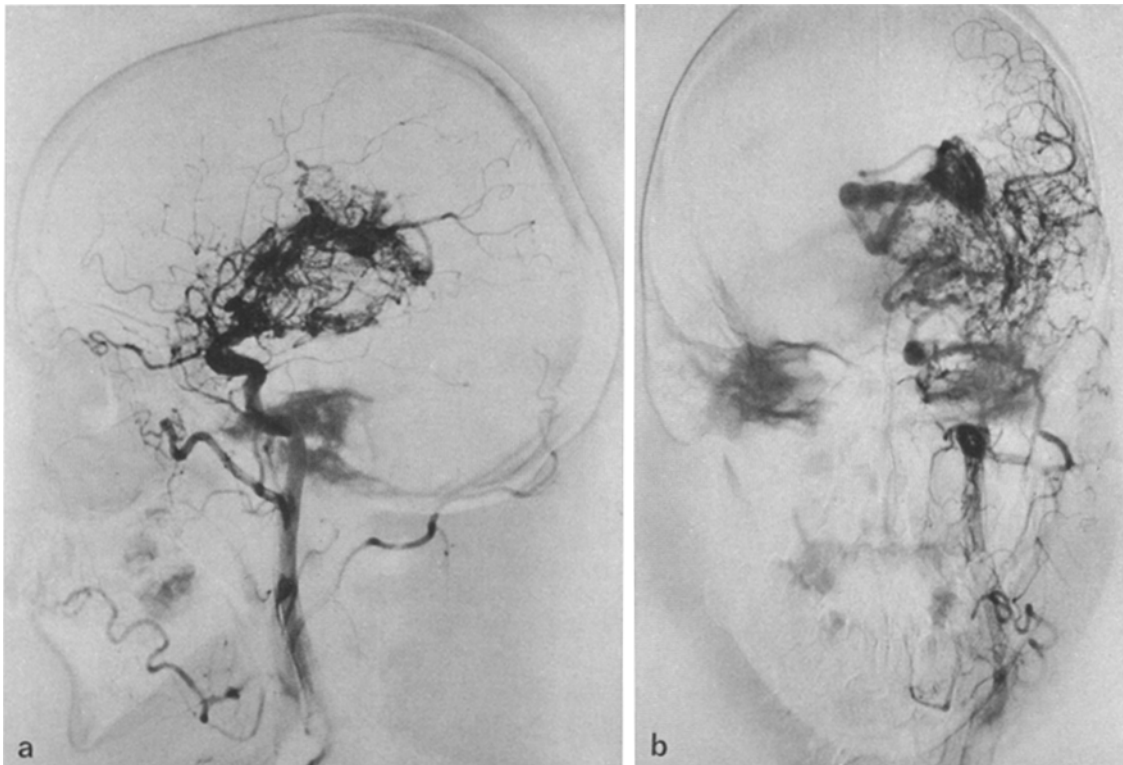


Fig. 3a–d. Left carotid and left vertebral angiograms. The large arteriovenous angioma is supplied by branches of the middle cerebral artery (**a, b**) and by cerebellar and thalamoperforate arteries (**c, d**)

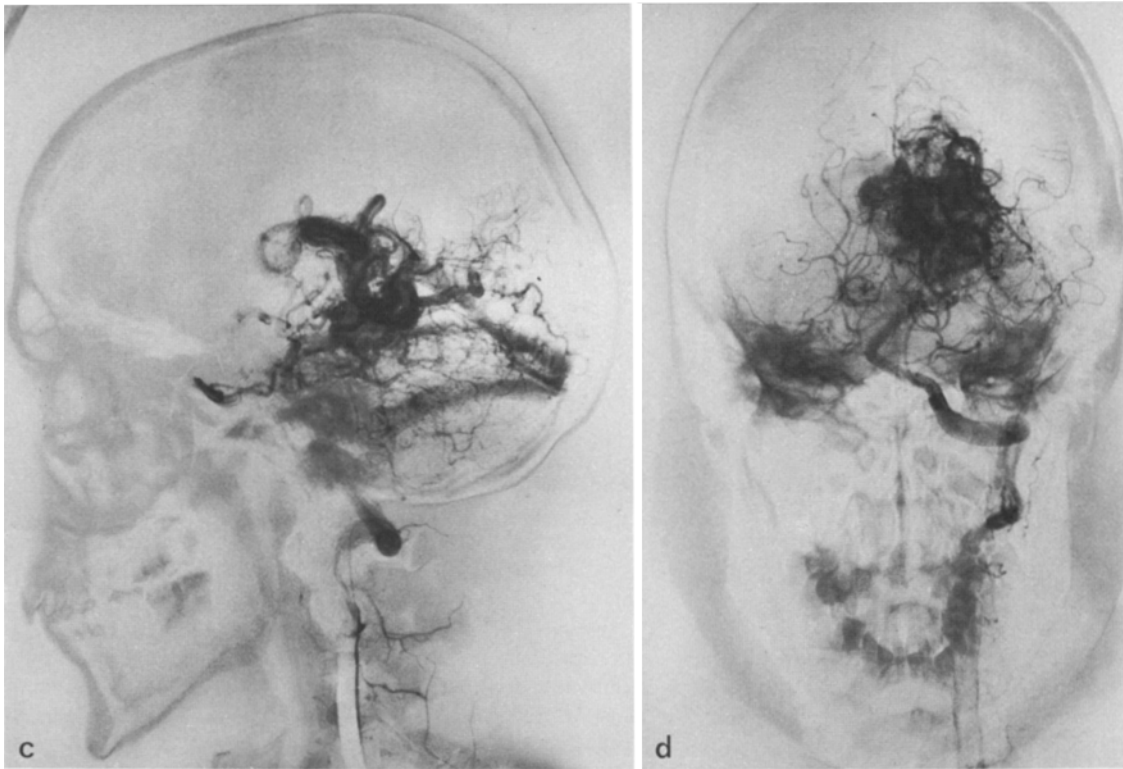


Fig. 3c, d

addition to vascular anomalies of the skin, haemangiomas of the abdominal organs [5], the vertebral column and the spinal meninges have been described [1, 3]. Angiography of the lower extremities has revealed haemangiomas, enlargement of the femoral artery, aplasia of the deep veins, varicosities of the superficial veins and thromboses [11]. Goldstein et al. [4] reported a case with hypertrophy of the left arm, naevi on the left face, and blindness in the left eye, as well as aplasia of the left internal carotid artery and an unusual collateral circulation to the left middle cerebral artery territory. Choroidal angiomas in KTS have been reported by O'Connor and Smith [9]. In some patients, KTS was associated with other phakomatoses: Sturge-Weber syndrome, tuberous sclerosis, neurofibromatosis, and von Hippel-Lindau disease [1, 2, 7].

However, an increased frequency of these additional disorders in KTS has not been demonstrated compared with the general population [2]. Therefore, it is difficult to decide whether a sign is specific to KTS or not. To our knowledge, the additional cerebral arteriovenous angioma seen in our patient had not been previously reported in this syndrome, but neurocutaneous dysplasias are typical in phakomatoses and cerebral angiomas is well known in some of these syndromes [12].

Furthermore, hypertrophy and arteriovenous disorders in KTS have been possibly related to abnormalities involving the brain stem, hypothalamus and diencephalon [2]. For this reason, we conclude that CT, magnetic resonance imaging and angiography may be likely to reveal more frequent cerebral vascular disorders in this syndrome.

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Received July 11, 1985 /

Received in revised form September 23, 1985 /

Accepted September 26, 1985