Multiple spinal cavernous malformations in Klippel-Trenaunay-Weber syndrome

Mnogie naczyniaki jamiste rdzenia kręgowego w zespole Klippla-Trenaunaya-Webera

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Neurologia i Neurochirurgia Polska 2012; 46, 5: 496-500
DOI: 10.5114/ninp.2012.31362

Abstract

Klippel-Trenaunay-Weber syndrome (KTWS) is a rare, congenital vascular disorder characterized by cutaneous haemangiomas, venous varicosities, and hypertrophy of the osseous and soft tissue. Various vascular anomalies of the central nervous system have been described in this syndrome. Two previous associations between KTWS and spinal cord cavernous malformations have been reported in the English literature. In this report, we present a patient in whom multiple cavernous malformations located in the conus medullaris region and cauda equina were associated with KTWS. General physical examination as well as neuroradiological and operative findings are described.

Key words: spinal cord, cauda equina, cavernous malformation, Klippel-Trenaunay-Weber syndrome.

Streszczenie

Zespół Klippla-Trenaunaya-Webera (ZKTW) to rzadkie, wrodzone schorzenie naczyniowe, w którym stwierdza się naczyniaki skóry, żylaki oraz przerost kości i tkanek miękkich. W przebiegu zespołu obserwowano różnorodne nieprawidłowości naczyniowe ośrodkowego układu nerwowego. W piśmiennictwie anglojęzycznym opisano wcześniej dwukrotnie związek ZKTW z obecnością naczyniaków jamistych rdzenia kręgowego.
W niniejszej pracy przedstawiono przypadek pacjenta, u którego w przebiegu ZKTW występowały mnogie naczyniaki jamiste okolicy stożka rdzeniowego i ogona końskiego. Omówiono wyniki badania klinicznego, badań obrazowych i nieprawidłowości stwierdzone w czasie leczenia chirurgicznego.

Słowa kluczowe: rdzeń kręgowy, ogon koński, naczyniak jamisty, zespół Klippla-Trenaunaya-Webera.

Introduction

Klippel-Trenaunay-Weber syndrome (KTWS) is defined as a congenital vascular disorder characterized by cutaneous haemangiomas, venous varicosities, and hypertrophy of the osseous and soft tissue. Vascular anomalies of the central nervous system, such as spinal and cerebral arteriovenous malformations (AVM), cavernous malformations, venous angioma, angiomyolipoma, venous varicosities and aneurysms, have been described in this rare syndrome [1]. Two previous associations between KTWS and medullary cavernomas have been reported in the English literature [1,2]. Cavernous malformations of the cauda equina are very rare, with only 13 reported cases in 2007 [3]. We present a unique case of multiple cavernous malformations located in the conus medullaris region and cauda equina associated with KTWS.

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Received: 25.07.2011; accepted: 25.01.2012

Case report

A 49-year-old man presented with low back pain radiating to both lower extremities. His symptoms had been increased progressively for three months. Family history did not show any significant finding. Neurological examination revealed left distal lower limb muscle power of 1/5. There was no genitourinary dysfunction or abnormal deep tendon reflexes. In general physical examination, multiple, grape-like, skin-coloured, reddish or purple, firm subcutaneous nodules on the dorsum of the foot were noted. Similar purple nodules with bleeding ulcers and port-wine stains were also seen on the left calf. There were multiple reddish or purple nodules and port-wine stains on the left buttock (Fig. 1A-C). These typical findings were consistent with KTWS.

Spinal magnetic resonance imaging (MRI) revealed two lesions with a maximum diameter of 2 cm. The first lesion was located in the spinal cord at the Th12-L1 level. This lesion was moderately hyperintense on T2-weighted sequences with peripheral low signal (hemosiderin) (Fig. 2A). After injection of gadolinium, the lesion showed heterogeneous enhancement. The second lesion, located in the cauda equina at the L5 level, was hyperintense both on T1- and T2-weighted images (Fig. 2B). T2-weighted sequences showed a peripheral rim and there was intense enhancement after gadolinium injection. MRI studies of brain, cervical and thoracic spine were normal. Computed tomography (CT) scans of the thorax and abdomen did not show any lesion in visceral organs.

The patient was operated on with laminectomy at the L5 level and after opening the dura mater, a 20 mm red-purple tumour was observed. The tumour originated from a nerve root in the cauda equina and was resected en bloc after cutting this root. There were also

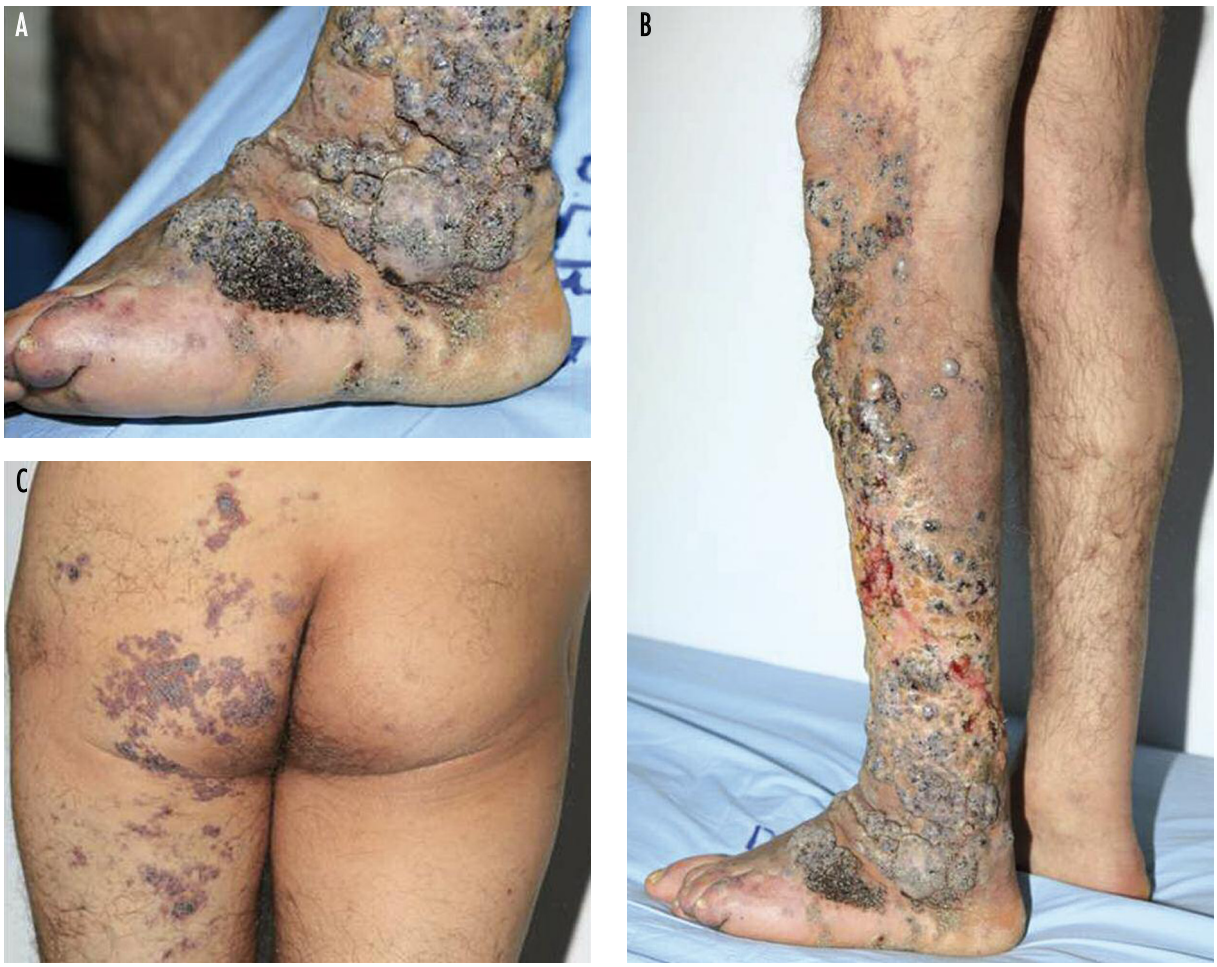

Fig. 1. Multiple, grape-like, skin-coloured, reddish or purple, firm subcutaneous nodules on the dorsum of the foot (A). Similar purple nodules with bleeding ulcers and port-wine stains on the left calf (B). Multiple reddish or purple nodules and port-wine stains on the left buttock (C)