SCRIPTA MEDICA (BRNO) – 77 (4): 207–212, *October 2004*

ABNORMAL DEVELOPMENT OF CEREBRAL DURAL SINUSES AND JUGULAR VEIN

DUFKOVÁ E.[1], PROCHÁZKA J.[2]

[1] Department of Paediatric Surgery, Orthopaedics and Traumatology, Faculty Hospital, Faculty of Medicine, Masaryk University, Brno

[2] Department of Paediatric Radiology, Faculty Hospital, Faculty of Medicine, Masaryk University, Brno

Received after revision September 2004

Abstract

Developmental abnormalities of the cerebral venous system occur due to stopped development of the venous system and continuing primitive embryonal veins draining into an abnormal draining vein. Association with occult vascular malformations was detected in patients with developmental venous anomalies in 40 %; it represents a risk of intracerebral bleeding. Therefore, further monitoring of the patients, preferably using the MRI method, is recommended. The case report presents an angiographic finding of anomalous sinistral cerebrocerebellar drainage and dysplasia of vena jugularis interna dextra with splitting and fusiform extension. This morphological finding is not currently demonstrated by clinically serious symptoms. Further dispensary care and a follow-up MRI examination are planned.

Key words

Developmental abnormality, Dural sinuses, Jugular vein, Cerebrocerebellar drainage, Angiography

INTRODUCTION

Developmental morphological anomalies in the cerebral dural sinuses, emissary veins, and the jugular bulb are closely related to the development of cerebral hemispheres, change in the postnatal type of blood circulation, and postural haemodynamic changes (*6*).

The embryonal development of the cerebral venous system can be summarised in several stages of development. Of the original undifferentiated primitive vascular plexus in the area of the head in an embryo with a size of 5 mm, three basic dural vascular plexuses are created. In a foetus with a size of 12 mm the frontal dural plexus is developed along with lateralisation of the future cerebral geminal veins. What follows then are anastomoses between the frontal and central plexus and the development of pial veins. At the stage of 16–21 mm, the sagittal, transverse, and

sigmoid sinuses develop. At the next stages of development, the prosencephalic vein appears temporarily and when the foetus grows to 40 mm, the deep cerebral venous plexus develops. At the size of 80 mm, the vena basalis and vena magna Galeni are formed. In the third month, the basic development of the foetal cerebral venous system reaches its final appearance (*10, 11*).

At that time the central cerebral vein and its branches are very well developed compared to the upper and lower cerebral veins. This condition changes between the 5th and 7th months, when there occurs a rapid development in the upper and lower cerebral veins along with a regression of the inflow of the central cerebral vein. In relation to further development of the cerebral hemispheres during the 7th prenatal month, the vein is then differentiated into insular, opercular, and convexity vein segments. At this stage, the anastomosing v. anastomotica superior and v. anastomotica inferior are visible, and later also the superficial v. cerebri media superficialis (*7*).

Increased blood flux from the rapidly developing cerebral hemispheres of the foetus leads to extending the sinus transversus while keeping the translucency between the sigmoid and jugular sinuses. Extension of the transversal sinus leads to the creation of the occipital, marginal sinus in the area of the foramen magnum and emissary veins. Creation of the jugular vein from jugular sinuses begins after the delivery along with the development of postnatal type of circulation and gradual postural effects (*6*).

CASE REPORT

A five-year-old patient was admitted for examination due to swelling on her neck on the right side during cough and cry observed from April 2002. The swelling was not painful, without any other local changes. Starting from February 2003, she suffered from a temporary uncharacteristic pain in the right limbs when lying down, which had disappeared before she was taken to hospital. The patient did not have any other subjective problems. The girl was born as the second child from the third hazardous pregnancy of her mother. The delivery was on time, with head first, via induction with preliminary loss of amniotic fluid. The childbirth weight was 3,100 g, length 49 cm. Psychomotoric development was slightly retarded in speech, without further alterations. For repeated infections of the upper respiratory system the patient underwent removal of adenoids in July 2000. She did not suffer from any other more serious illnesses, injuries or operations. She is orthopaedically checked for pedes planes and allergologically checked for allergic reactions to pollen and mites. She does not use any permanent medication except for antihistamine.

The clinical examination of the right half of the neck did not show any resistance or other pathological finding. During the Valsalva manoeuvre, a loosely bordered ventrolateral convexity on the right side was observed. Clinical neurological examination did not detect any focal symptomatology.

The laboratory results of screening biochemical and haematological examinations were within limits. Ultrasonographic examination of the neck showed gemination of the vena jugularis interna dextra, with communication in the area of the carotid bifurcation and in the area of the jugular vein. CT and MRI examination of the brain natively and after intravenous administration of a contrast medium did not show any pathological finding. On the basis of the ultrasonographic examination, an angiographic examination was made – both-sided angiography of cervical veins and cerebral angiography on the left with a finding: during sinistral carotid angiography the dextral sinuses were filled massively with the contrast medium, i.e. sinus transversus and sinus sigmoideus, filling subsequently

the v. jugularis interna dextra. Contrariwise, the sinus transversus sinister was by contrast shown only weakly and crevicularly, and the sinus sigmoideus sinister reached only approximately half the translucency compared to the right; as a result, the translucency of the v. jugularis interna sinistra was also lower, both relatively and absolutely. The other shown cervical veins on the left were filled normally. The cerebral carotid angiography on the left showed normal arterial phase; however, the phlebogram was abnormal. The enlarged veins on the convexity provide drainage leading to the sinus sagittalis superior, basally the v. basalis (Rosenthali), and temporo-occipitally the v. temporooccipitalis (Labbé). The sinus transversus was narrowed on the left and its contrast image was very weak, with a resulting reduction of translucency of the sinus sigmoideus. The v. jugularis interna dextra (VJID) developed quite abnormally, it was split into two parts in its caudal : the translucency of the medial part was almost 1/3 lower than the cranial VJID and the lateral part was fusiform aneurysmatically extended above double – triple the translucency of the cranial VJID (in dependence on the pulse phase). Thus, a highly reaching bulbus inferior v. jugularis ("high jugular bulb") was shown. However, the angiographic flow of the contrast medium was normal, no signs of stagnation of contrast in VJID were detected (*Figs. 1, 2*).

Furthermore, cardiological examination was conducted with a normal finding. Sonographic examination of the limb musculature showed a physiological finding. The EMG examination of lower limbs in the conduction study and needle EMG did not show any signs of peripheral neuropathy and myopathy, the SSEP examination of the n. tibialis and n. medianus was also within limits. The EEG record showed abnormal irregular basic activity with sinistral frequency and amplitude asymmetry with photostimulation and hyperventilation activation, no specific graphoelements were detected. The fundus oculi finding was normal.

DISCUSSION

Patients with venous anomalies in the area of head and neck may, to a larger extent, be also afflicted with cerebral vein anomalies (*3*). In patients with extensive superficial vein anomalies in the head and neck, a 20 % prevalence of developmental anomalies of the cerebral venous system is reported (*2*). These developmental abnormalities of the cerebral venous system occur due to stopped development of the venous system and continuing primitive embryonal veins draining into an abnormal draining vein (*9*). Therefore it is recommended that the presence of cerebral venous abnormalities should be specifically searched for in such patients (*2*).

Cerebral developmental venous anomalies as such are generally considered to be benign nosological units (*2, 5, 8*), in most cases without clinical signs. What is, however, important is their coincidence with venous malformation, mainly cavernoma. Association with occult vascular malformations was detected in patients with developmental venous anomalies in 40 % (*5*); this represents a risk of intracerebral bleeding. Therefore, further monitoring of the patients preferably using the MRI method is recommended as this method is considered to be an optional method imaging both the nosological units (*5*).

The morphological finding presented here is not currently demonstrated by clinically serious symptoms. Further dispensary care and a follow-up MRI examination are planned.

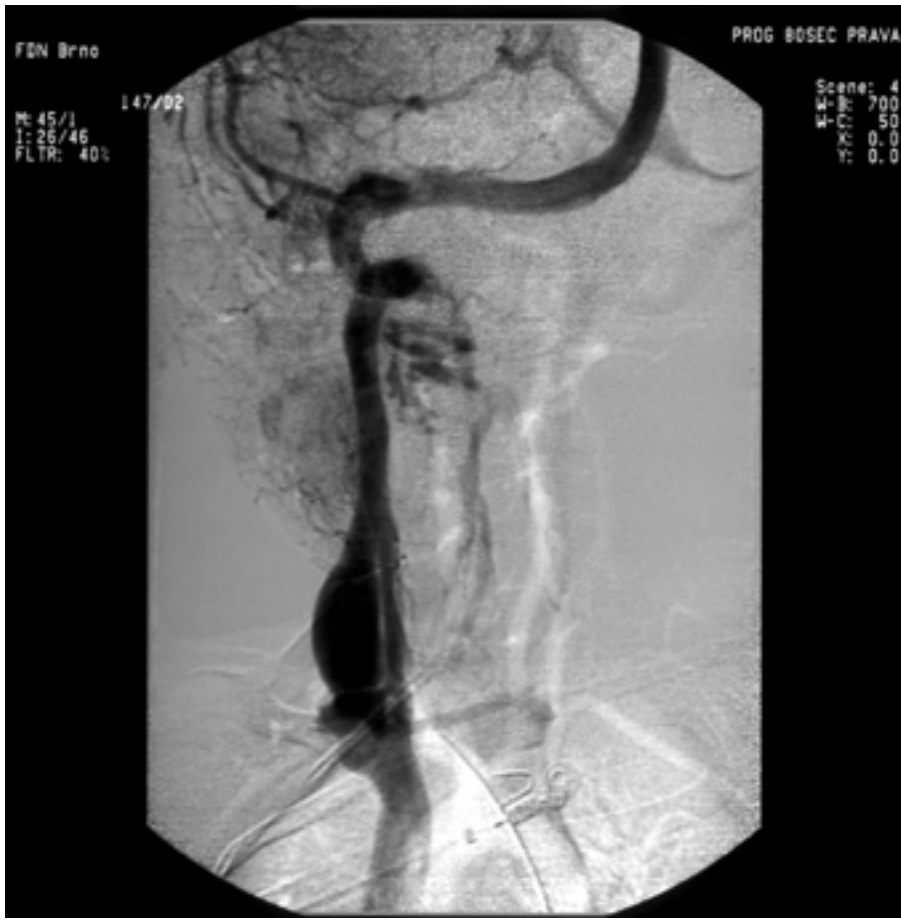

Fig. 1
Angiographic finding of split caudal part of vena jugularis interna dextra

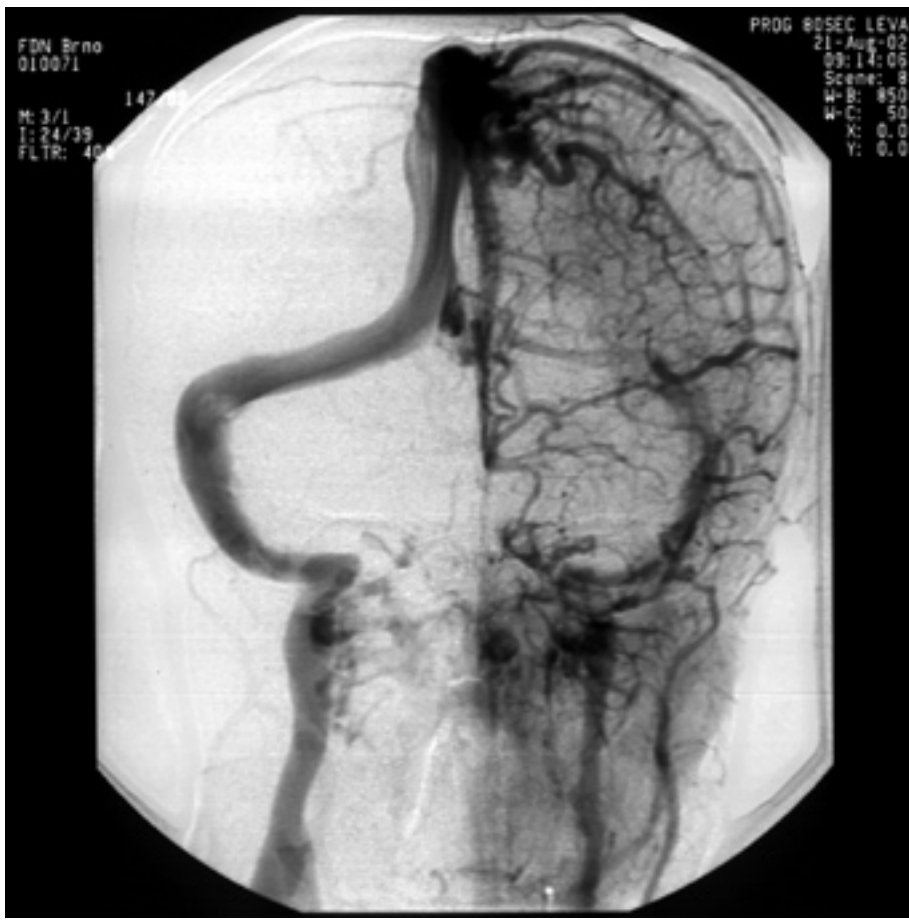

Fig. 2
Angiographic image of the sinistral cerebrocereberal vein drainage

VÝVOJOVÁ ANOMÁLIE MOZKOVÝCH ŽILNÍCH SPLAVŮ A JUGULÁRNÍ ŽÍLY

Souhrn

Vývojové abnormality mozkového žilního systému vznikají zastavením vývoje žilního systému a přetrváváním primitivních embryonálních vén drénujících do abnormální odvodné žíly. Asociace se skrytou vaskulární malformací byla u pacientů s vývojovou žilní anomálií nalezena až ve 40 % a představuje riziko intracerebrálního krvácení. Je proto doporučováno další sledování pacientů, nejlépe metodou MRI. Kazuistika popisuje angiografický nález anomální levostranné cerebrocerebelární drenáže a dysplazii vena jugularis interna dextra s rozštěpením a vřetenovitým rozšířením. Tento morfologický nález se v současné době neprojevuje klinicky závažnými příznaky. Je plánována další dispenzární péče a kontrolní MRI vyšetření.

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