Metameric syndromes

To The Editor: I read with interest the case report by Nishino et al.1 (Nishino K, Ito Y, Sorimachi T, et al: Sturge-Weber syndrome associated with arteriovenous malformation in a patient presenting with progressive brain edema and cyst formation. Case report. J Neurosurg Pediatr 5:529–534, May 2010) in which the authors described a 3-year-old girl with a port-wine stain within the ophthalmic distribution of the left trigeminal nerve and an arteriovenous malformation (AVM) in the left temporoorbital region and basal ganglia. The AVM progressively enlarged with occulsive changes of the venous system and brain edema. The authors concluded that the abnormalities in the brain venous system (spare cortical veins, occluded left sigmoid and superior sagittal sinuses) were compatible with Sturge-Weber syndrome. They also stated that the engorged scalp vein connected to the superior sagittal sinus was consistent with sinus pericranii.

Reviewing the constellation of clinical and imaging features documented by the authors, I disagree with the diagnoses of Sturge-Weber syndrome and sinus pericranii.

The authors fail to distinguish between two different developmental entities of the craniofacial vasculature: cerebrofacial venous metameric syndromes (CVMSs) and cerebrofacial arteriovenous metameric syndromes (CAMSs). Wyburn-Mason (Bonnet-Dechaume-Blanc) syndrome is the prototype of CAMS, and Sturge-Weber syndrome is the prototype of CAMS.

Wong and colleagues2 identified three different types of CAMS. Type 1 involves the median (olfactory) prosencephalon (hypothalamus, corpus callosum, and hypophysis) and nose. Type 2 involves the lateral (optic) prosencephalon (optic nerve, retina, parietal-temporal-occipital lobes, and thalamus) and maxilla. Type 3 affects the rhombencephalic (otic) area (cerebellum, pons, and petrous bone) and mandible.

Similar to CAMS, CVMS can be divided into three types according to the same developmental anatomical landmarks. Note that these disorders can present with incomplete involvement of the expected pathway or a combination of more than one territory.2 The clinical sequelae of CAMS and CVMS are quite different. The former tends to be more aggressive and causes substantial morbidity. Sinus pericranii is transcalvarial communication between dural sinuses and scalp veins2 associated with scalp or facial venous malformation. Transcalvarial drainage of a hypertensive dural venous system caused by AVMS, a known phenomenon, is different from sinus pericranii. The latter refers to a pure venous phenomenon.

In view of this information, I believe that the diagnosis is likely to be extensive CAMS with incomplete facial features.

Ahmad I. Alomari, M.D., M.Sc., F.S.I.R.
Children’s Hospital Boston
Harvard Medical School
Boston, MA

Disclosure

The author reports no conflict of interest.

References


Response: No response was received from the authors of the original article.

Long-term outcomes of dorsal rhizotomy

To The Editor: We read with great interest the study recently published by Dudley et al.1 (Dudley RWR, Parolin M, Gagnon B, et al: Long-term functional benefits of selective dorsal rhizotomy for spastic cerebral palsy. Clinical article. J Neurosurg Pediatr 12:142–150, August 2013). The authors report on functional outcomes before and 1, 5, 10, and 15 years after selective dorsal rhizotomy (SDR) in children who had been entered prospectively in the McGill University Rhizotomy Database. The strength of this study lies in the use of standardized and validated outcome measures, including the Ashworth scale for muscle tone in the lower extremities, the Gross Motor Function Measure (GMFM) for gross motor function assessments, and the Pediatric Evaluation of Disability Inventory (PEDI) as an indication of the patient’s performance of activities in daily life. In addition, the authors

documented any additional orthopedic interventions, and they attempted to stratify the results per Gross Motor Function Classification System (GMFCS) levels and described the course of the functional outcomes with group-based trajectory modeling (GBTM) over time. Unfortunately, the data sets are incomplete, which in our opinion makes it difficult to state the long-term benefits of SDR as strongly as the authors conclude.

Between 1991 and 2001, 105 children underwent SDR, of whom 102 were included in this follow-up study. Sex and average age at surgery were reported for the entire cohort, but full information was available about the specific cerebral palsy (CP) diagnosis in 81 patients, the preoperative GMFCS levels in 52, and additional interventions such as orthopedic procedures and botulinum toxin injections in 88. There was also no demographic and background information reported on the 15-year follow-up study cohort, on which the authors’ major conclusions are based.

The study sample size also decreased considerably over time. Impressively, the original study cohort consisted of 102 children, but the number of participants decreased to 97, 62, 57, and 14 at the 1-, 5-, 10-, and 15-year follow-up studies, respectively. The last postoperative study included GMFM scores of 14 participants (14% of the original study cohort), while Ashworth and PEDI scores were only reported for 8 participants (8%). The stratification of the results by GMFCS levels in this last follow-up study was performed in 13 (13%) and 7 (7%) participants regarding GMFM and PEDI, respectively, but the stratification was not reported for the Ashworth scale scores. In addition, GBTM was based on follow-up studies up to 10 years after SDR (not 15 years), of which 50% of the data was missing, and none of the participants reported complete data of all trajectories. The association found from this (incomplete) GBTM data set was subsequently used to design a predictive tool (Predictive Index for Long-Term Ambulation after Rhizotomy [PILAR]).

The authors acknowledge the decrease in sample size as a limiting factor of their study, but they defend this with the following clarification: “The decrease in the number of patients presented at each time point was, for the most part, not because of patient attrition, but rather that patients had not yet reached the 10-year, and particularly the 15-year, follow-up.” This is somewhat at variance with the emphasis of the title, which refers to long-term follow-up.

If the authors’ interest is indeed the long-term outcome, they may be interested to refer to a number of studies published from Cape Town, including 20-year follow-up studies and cross-sectional studies including outcomes from 17 to 26 years after SDR. One of the challenges of true long-term follow-up studies is changes in the outcome measures used. Indeed, when the original Cape Town studies were initiated in the early 1980s, current standardized outcome measures such as the GMFM and PEDI did not exist. However, there are similarities between assessment tools such as the 5-point muscle tone and functional movement scales and the Life-Habit questionnaire and older measures such as the Ashworth scale, GMFM, and PEDI. As with the present study, the absence of a control group was a limiting factor in our 20-year follow-up study, but this is a common problem in this type of research. Last but not least, we acknowledged our small sample size (n = 14) but nevertheless were able to track down, assess, and report on each of these 14 participants (100%) 20 years after SDR, which provides a comprehensive picture of the functional status of the entire study cohort in the long term.

In 2015 we plan to conduct a 30-year follow-up study and hope to see the positive results described in our former studies maintained in the adults with spastic CP. The functional benefits we have shown are comparable to the admitted incomplete outcomes of the 15-year follow-up studies as published by Dudley et al. We look forward to seeing the long-term follow-up data from the Montreal group but would encourage the authors to attempt to report on complete study cohorts in order to ensure the integrity of their results.

Nelleke G. Langerak, Ph.D.
Christopher L. Vaughan, Ph.D., D.Sc.
Jonathan C. Peter, M.B.Ch.B., F.R.C.S.
A. Graham Fieggan, M.B.Ch.B., M.S.C., M.D., F.C.S.
University of Cape Town
Cape Town, South Africa
Warwick J. Peacock, M.B.Ch.B., F.R.C.S.
University of California at San Francisco
San Francisco, CA

Disclosure
The authors report no conflict of interest.

References

Response: No response was received from the authors of the original article.
Angiocentric glioma

To THE EDITOR: I read with interest the article by Shakur et al. (Shakur SF, McGirt MJ, Johnson MW, et al: Angiocentric glioma: a case series. Clinical article. J Neurosurg Pediatr 3:197–202, March 2009). The authors reviewed the literature on angiocentric glioma in children and reported 3 of their cases. They stated “The median age at surgery for patients among the 25 cases was 6.5 years, compared with 9.9 years for children in the Göteborg epilepsy surgery series.” This statement erroneously indicates that patients with angiocentric glioma underwent surgery more than 3 years earlier than other children who underwent surgery for seizures, because when reviewing Table 1 it became clear that the 5 cases included from the study of Wang et al. were recorded using the patients’ ages when their seizures began, not their ages at the time of resection and histopathological diagnosis. Similarly, while recording the pediatric cases from the study of Preusser et al. the authors used the ages at which the seizures began, not when tissue diagnoses were obtained. In Table 1, it is stated that the ages given were the ages at the time of the operation. Additionally, the authors elected to exclude 1 patient from the study of Wang et al. and 3 patients from the study of Preusser et al. because these patients underwent surgery in adulthood although their seizures began in childhood. Using the same references in Table 1 and reported ages at the time of the surgery, I compute the median age as 10 years in children who underwent resection of angiocentric glioma. This is almost identical to 9.9 years, which was reported in the Göteborg epilepsy surgery series.

KORGUN KORAL, M.D.
University of Texas Southwestern Medical Center
Dallas, TX

Disclosure

The author reports no conflict of interest.

References


RESPONSE: We would like to thank Dr. Koral for his astute observations regarding our recent article on angiocentric glioma. Although age was not the focus of our case series, we acknowledge that the median age at surgery for patients included in the 25 cases studied was more similar to the median age of children in the Göteborg epilepsy surgery series than we initially published. Nonetheless, our conclusion that angiocentric glioma is a distinct clinicopathological entity among seizure-associated lesions in children remains steadfast. Specifically, postoperative seizure freedom in our review was 100% with gross-total resection and 56% with subtotal resection at 2.3–168 months of follow-up, whereas low-grade gliomas accounted for 8% of the histopathological diagnoses in the Swedish study and were associated with 66.7% seizure freedom by the 2-year follow-up. Angiocentric glioma may therefore have a more favorable seizure prognosis compared with other supratentorial gliomas characterized by seizures in children, and our clinical analysis affirms that the extent of resection may play a key role in determining seizure-free survival in children with brain tumors.

Our compilation and assessment of the case reports published since the original description of angiocentric glioma yielded the following key findings that have remained unchanged since initial publication: 1) seizure is the most common symptom at presentation; 2) MRI demonstrates a supratentorial, nonenhancing, T1-hypointense, T2-hyperintense lesion; 3) the tumors have characteristic pathological features; and 4) the outcome following treatment with gross-total resection of the tumor is excellent.

SOPHIA F. SHAKUR, M.D.
The University of Chicago
Chicago, IL

GEORGE I. JALLO, M.D.
Johns Hopkins University Medical Center
Baltimore, MD

MATTHEW J. McGIRT, M.D.
Vanderbilt University Medical Center
Nashville, TN

Please include this information when citing this paper: published online October 11, 2013; DOI: 10.3171/2010.1.PEDS09527. ©AANS, 2013