Cerebral Venous Sinus Thrombosis in Pediatric age group

Systematic Review

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Presented by

Aya Hussein Ibrahim Hussein

(M.B.B.Ch)

Under supervision of

Prof. Dr. Nahed Salah-Eldeen Ahmed

Professor of Neurology and Psychiatry

Faculty of Medicine – Ain shams University

Prof. Dr. Salma Hamed Khalil

Professor of Neurology and Psychiatry

Faculty of Medicine – Ain Shams University

Dr. Ahmed Ali Ibrahim El-basiouny

Assistant Professor of Neurology and Psychiatry

Faculty of Medicine – Ain Shams University

Faculty of Medicine

Ain Shams University

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الخثار فى الجيوب الوريدية الدماغية فى الأطفال

رسالة

توطئة للحصول على درجة الماجستير في طب المخ والأعصاب والطب النفسي

مقدم من الطبيبة

آيه حسين ابراهيم حسين

بكالوريوس الطب والجراحه

تحت إشراف

الأستاذ الدكتور/ ناهد صلاح الدين أحمد

أستاذ طب المخ والأعصاب والطب النفسى

كلية الطب – جامعة عين شمس

الأستاذ الدكتور/ سلمى حامد خليل

أستاذ طب المخ والأعصاب والطب النفسى

كلية الطب – جامعة عين شمس

الدكتور/ أحمد على إبراهيم البسيونى

أستاذ مساعد طب المخ والأعصاب والطب النفسى

كلية الطب – جامعة عين شمس

كلية الطب

جامعة عين شمس

٢٠١٥

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Aya Hussein Ibrahim Hussein

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Contents

List of Abbreviations

aCL	**:**	Anti-cardiolipin
AED	**:**	Anti Epileptic Drugs
AHA	**:**	American Heart Association
AP	**:**	Antroposterior
aPTT	**:**	Activated partial thromboplastin test
APC-R	**:**	Activated Protien C Resistance
APS	**:**	Anti-Phospholipid Syndrom
Apl	**:**	Anti-Phospholipid
AT	**:**	Anti thrombin
BV	**:**	Basal vien
bid	**:**	Bis in die
ß2GPI	**:**	Beta 2Glycoprotien I
CSF	**:**	Cerebro-Spinal Fluid
CT	**:**	Computerized topography
CTA	**:**	Computerized topography Arteriography
CTV	**:**	Computerized topography Venography
dMCV	**:**	Deep Middle Cerebral Vein
DST	**:**	Dural Sinus Thrombosis
DWI	**:**	Diffusion Weighed Imaging
FLAIR	**:**	Fluid Attenuated Inversion Recovery
FVL	**:**	Factor Five Leiden
GCV	**:**	Great Cerebral Vein
HHcy	**:**	Hyperhomocysteinemia
ICP	**:**	Intra Cranial Pressure

ICV	**:**	Internal Cerebral veins
IPS	**:**	Inferior Petrosal Sinus
LAC	**:**	Lupus Anti-Coagulant
LMWH	**:**	Low Molecular Weight Heparin
LP(a)	**:**	Lipoprotein (a)
OAT	**:**	Oral anticoagulant therapy
PC	**:**	Protein C
PS	**:**	Protein S
PT-M	**:**	Prothrombin Mutation
PCT	**:**	Phase Contrast Tequniques
SLE	**:**	Systemic Lupus Erythromatosis
SPaS	**:**	Sphenopairtal Sinus
SPS	**:**	Superior Petrosal Sinus
SSS	**:**	Superior Sagittal Sinus
SRS	**:**	Straight sinus
TCCS	**:**	Trans Cranial colored scan
TCD	**:**	Trans Cranial Doppler
MTHFR	**:**	Mthyl-Tetrahydrofolate Reductase
TOF	**:**	Time of flight
tPA	**:**	Tissue Plasminogen Activator
TS	**:**	Transverse sinus
UFH	**:**	Unfractionated Heparin
VKA	**:**	Vitamin K antagonist
VTE	**:**	Venous Thrombo Embolisim

List of Figures

List of Tables

Introduction

Cerebral veins contain about 70% of the total cerebral blood volume, but cerebral venous sinus thrombosis (CVST) occurs about thousand times less often than arterial stroke. Arterial and venous stroke cause different neurological deficits and occur in people of different ages. About half of the patients with an arterial stroke are older than 75 years; whereas CVST most frequently affects young adults and children. It is not a manifestation of atherosclerosis, but usually is associated with a prothrombotic state due to (inherited or acquired) thrombophilia, other blood disorders, dehydration, infectious diseases, cancer, or more rare causes ***(Ferro et al., 2004).***

The annual incidence is currently estimated to be 0.67 in 100.000 per year, with a preponderance of neonates (43%), and over half (54%) being less than 1 year of age ***(deVeber et al., 2001).*** Interestingly, there appears to be a male predominance in this age group, with as many as 75% of neonates presenting with CVST being male. Beyond the neonatal period, the incidence of CVST is relatively evenly distributed among the age groups to 18 years, and averages between 2 and 4% per year ***(Golomb et al., 2003),*** While in another study on older children: the female to male ratio was 1.29:1 ***(Ameri and Bousser, 1999).***

Modern neuroimaging techniques have improved the diagnostic process and together with the increased awareness this will probably result in increased recognition in the future.

CVST has received far less attention in clinical research than arterial stroke. The small number of patients with CVST limits the performance of large epidemiological studies and clinical trials on a scale similar to that in patients with arterial stroke ***(Stam, 2005).***

Thrombosis of the cerebral veins may cause focal deficits due to local effects of venous obstruction, but also more generalized effects as a result of increased cerebrospinal fluid (CSF) pressure caused by blocking of the major sinuses. In the majority of patients, these two processes occur simultaneously ***(Stam, 2005).***

The course and clinical features of CVST are highly variable. It is only after the introduction of computerized tomography and particularly magnetic resonance imaging that verification of the diagnosis during life has become part of daily clinical practice. As a result, the clinical spectrum of the disease has enormously increased. It is well known that the clinical features of CVST are extraordinarily variable. Consequently, the diagnosis may be difficult. The average delay from the onset of symptoms to the diagnosis is 7 days ***(Ferro et al., 2004).***

Patients may present to an ear, nose and throat surgeon with recurrent ear infections and headache, to a pediatrician with headache, vomiting and vague symptoms in the context of a systemic disease, or to a neurologist

because of epileptic seizures, or to a neurosurgeon because of chronic intracranial hypertension. The variety of clinical features and the large number of specialists dealing with the care of patients with CVST have resulted in papers in peer-reviewed journals of many different disciplines, making a clear and concise overview difficult ***(Ferro et al., 2004)***.

Aim of work

The aim of this work is to highlight the recent insights of the pathogenesis, clinical presentation and diagnosis of cerebral venous sinus thrombosis in the pediatric age group also for better management and improving the quality of life of these patients.

Anatomy of Cerebral Veins and Sinuses

The cerebral venous system can be divided into a superficial and a deep system. The superficial system comprises sagittal sinuses and cortical veins, which drain superficial surfaces of both cerebral hemispheres. The deep system consists of the lateral sinus, straight sinus and sigmoid sinus along with draining deeper cortical veins. Both of these systems mostly drain into internal jugular veins (Fig.1) ***(Kïlïc et al., 2008).***

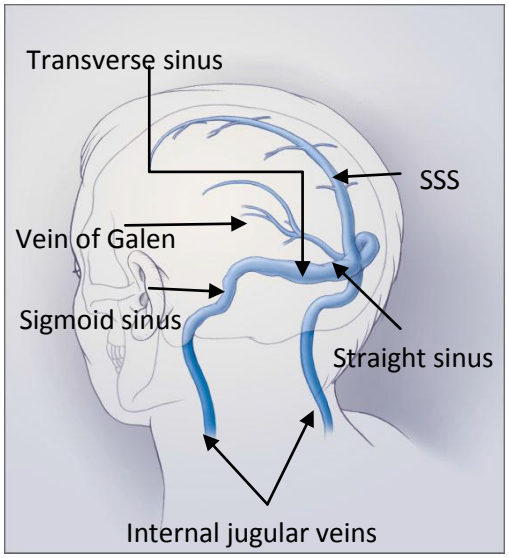

Fig. (1) Illustration of cerebral venous sinuses ***(Stam, 2005).***

Generally, venous blood drains into the nearest venous sinus or into deep veins. The superficial cerebral veins are interlinked with anastomotic veins of Trolard and Labbé (Fig.2). Thus, the superolateral surface of the hemisphere drains into the superior sagittal sinus while the posteroinferior aspect drains into the transverse sinus ***(Kïlïc et al., 2008).***