Risk Factors for Pediatric Cerebral Sinus Vein Thrombosis

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INTRODUCTION

- Cerebral sinus vein thrombosis (CSVT) is a type of stroke consisting in thrombosis of the dural sinus and/or cerebral veins. The impaired venous drainage may result in venous infarcts and possible intracranial hypertension. Up to 60% of affected children have focal brain abnormality related to venous congestion resulting in ischemia and/or hemorrhage
- The estimated incidence in children is 0.6 per 100,000 children per year however the symptoms and presentations are variable among all age ranges raising the concern for missed diagnosis.
- Poor outcomes, including death, happen in 9 to 29% of patients. In addition, neurologic deficits, primarily cognition and behavior, are seen in 50% of affected children. Despite new progress on pediatric stroke, CSVT is under-recognized and its risk factors are not well established.

METHODS

- We conducted a single-center review of pediatric patients diagnosed with CSVT between 2008 and 2018, seen by the Hematology service at Cook Children's Medical Center (CCMC). Neonates (birth to 28 days) and children (29 days to 18 years) with radiographically confirmed CSVT were identified through the Cook Children's database.
- Through chart review, we recorded the age, birthday, sex, comorbidities, symptoms, neurological deficits and anticoagulation regimen. We collected the results of imaging done at diagnosis, 5-14 days, 3, 6 and 12 months from diagnosis. In addition, we noted hemoglobin levels at diagnosis and Prothrombotic laboratory values including antithrombin III, protein C and protein S activities, homocysteine, lupus anticoagulant with anticardiolipin antibodies, Factor V Leiden and Prothrombin gene mutations.

RESULTS

- There were 87 patients meeting inclusion criteria
- Tables 1, 2, and 3: neurologic deficits, symptoms, and diagnoses
- Table 4: thrombophilia evaluation
- Figure 1: hemoglobin values

Table 1. Neurologic deficits at diagnosis

Neurologic Deficit	N (%)
Altered mental status	8 (9%)
Increased intracranial pressure	8 (9%)
Focal weakness/hemiparesis	5 (6%)
Cranial nerve palsy	4 (5%)
Diplopia	3 (3%)
Papilledema	3 (3%)
Ataxia	2 (2%)

Table 2. Symptoms at diagnosis

Symptoms	N (%)
Fever	25 (29%)
Headache	28 (32%)
Vomiting/Nausea	29 (33%)
Neurologic Deficits	25 (29%)

Table 3. Comorbidities at diagnosis

Diagnosis	N (%)
Mastoiditis*	23 (26%)
Otitis media*	21 (24%)
Lemierre's syndrome	9 (10%)

* 16 (18%) had both mastoiditis and otitis media at diagnosis

Figure 1. Hemoglobin level at diagnosis

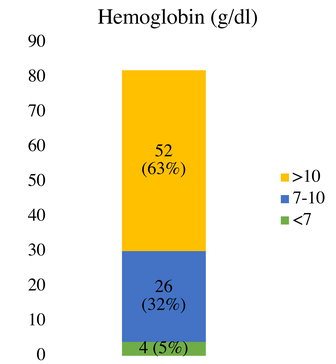

Table 4. Laboratory results by number tested

Lab Test	Abnormal N	Normal N	No Lab N
Antithrombin III	4	35	51
Protein C	5	27	58
Protein S	5	26	59
Lipoprotein A	1	2	87
Homocysteine	0	26	64
Lupus Anticoagulant	4	22	64
Anticardiolipin Antibodies IgM	4	32	54
Anticardiolipin Antibodies IgG	1	37	52
Beta-2 Glycoprotein Antibodies IgM	2	24	64
Beta-2 Glycoprotein Antibodies IgG	4	24	62
Prothrombin Gene Mutation	2	30	58
Factor V Leiden	2	29	59

CONCLUSION

- Infectious causes such as otitis media, mastoiditis and retropharyngeal abscess can be common contributors of pediatric CSVT. Almost 20% of our cases of CSVT did have concurrent mastoiditis and otitis media at diagnosis (Table 3). The median age for presenting cases was 5 years which is consistent with higher incidence of these infections among younger children.
- In our series, a quarter of the cases at presentation had a noted neurologic deficit not limited to altered mental status (Table 1). We suggest that if these infections are diagnosed, there should be a high index of suspicion for CSVT if any neurological symptoms are present.
- Anemia is a little known independent risk factor for thrombosis, especially CSVT. In our series, anemia (defined as hemoglobin of less than 7 gr/dl), presented in 5% of our patients. In addition, 32% of patients had a hemoglobin between 7 and 10 gr/dl. (Figure 1).
- Not all patients with CVST underwent a complete thrombophilia evaluation. It was usually not done if the CSVT was believed to be provoked by local inflammation/infection. Although this creates a limitation in our study, we found that in our group, thrombophilia was rather rare (Table 4).

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