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Cerebral Venous Sinus Thrombosis (CVST): Long-Term Single-Center Experience

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Background

CVST is a rare location of thrombosis involving Dural/ cerebral venous sinuses. It affects around 5-10 people per million population annually.

It is an uncommon but life-threatening form of stroke affecting younger individuals. Therefore, identifying and treating in a timely manner is critical.

Rarer thrombotic disorders like paroxysmal nocturnal hemoglobinuria (PNH) or Janus Kinase 2 (JAK2) mutation positive myeloproliferative neoplasms (MPN) can rarely present with CVST. It can also present during pregnancy for the first time.

Diagnosis is often established by Computed Tomography (CT) or Magnetic Resonance Imaging (MRI).

Infections, certain medication use (asparaginase or birth control pills) could lead to CVST. Patients often present with headaches, seizures or neurological deficits.

Management is often with systemic anticoagulation despite intraparenchymal hemorrhage. Reducing intracranial pressure by invasive approaches is sometimes needed.

Methods

We conducted a retrospective study of the patient population with CVST who were seen in the UNMC Hematology Clinic from 2010 onward and followed up for at least 3 months following the diagnosis.

After IRB approval, medical records of 51 patients who met this criteria were reviewed carefully. Data was collected regarding demographics, risk factors, health history, CVST presentation, associated diseases, any acquired/inherited thrombophilia, laboratory results at earliest available date from diagnosis, and treatments (antithrombotic therapy) received.

Descriptive statistics are analyzed.

Patient Demographics

Variable	Range/percentages	Median
Age	18-72 years	40 years
Gender	31 Females (60.78%) 20 Males (39.22%)	N/A
Ethnicity	Caucasian: 43 (84.31 %) African American: 4 (4.35%) Asian: 2 (4.35%) Hispanic: 1 Other: 1	N/A
Follow-up	Alive (N = 44): 0-299 months	70 months
Death	6 (86% survival rate)	N/A

Table 1. Descriptive statistics for patients with CVST at UNMC.

Results

Descriptive statistics with medians, quartiles, frequencies, and percentages are reported. We also provided the various anticoagulants that patients received. Provoking risk factors when identified are listed by frequency.

Table 1.

Associated Disease	Number of Patients (N = 51)	Percentages
Myeloproliferative neoplasms (MPNs) Polycythemia vera (PV) or Post-PV Myelofibrosis (MF) (2/51) Essential Thrombocythemia (ET) or Post-ET MF (1/51)	3	6%
Paranasal Sinus Disease	23	45.1%
Paroxysmal Nocturnal Hemoglobinuria	0	0%
Antiphospholipid Antibody Syndrome	5	9.8%
Sarcoidosis	1	1.9%
Vaccine-induced thrombotic thrombocytopenia (VITT) Ad26.COV2.S vaccine (Johnson & Johnson/Janssen)	2	3.9%
Idiopathic	31	60.8%

Table 2. Primary overview of CVST in patients at UNMC.

Variable	Valid	Data Missing	Range (%)	Mean	Median
Presenting Symptom	51 (100%)	0	Headache: 32 Nausea/Vomiting: 15 Seizure: 19 Body Weakness: 16 Vision Impairment: 10 Cognitive Difficulty: 6	N/A	N/A
Cancer history	51 (100%)	0	Positive: 13 Negative: 38	N/A	N/A
Smoking	51 (100%)	0	Yes: 10 No: 41	N/A	N/A
Obesity	51	0	Yes: 25 No: 26	N/A	N/A
Hormone Treatment	31	ONLY FEMALE	OC Pills: 15 Pregnant: 2 N/A: 16	N/A	N/A
Hemorrhage	15 (100%)	4 (50%)		N/A	N/A
Seizure	51 (100%)	0	Yes: 19 No: 32	N/A	N/A
Hemoglobin	51 (100%)	0	7.5–18.7 gm/dL	13 gm/dL	13 gm/dL
Platelet count (10^3)	51 (100%)	0	9–1202/uL	259/uL	239/uL
WBC (10^3)	51 (100%)	0	0.1–22/uL	10/uL	10/uL
Protein C Activity	27 (53%)	24 (47%)	28–187%	108%	105%
Protein S Activity	27 (53%)	24 (47%)	35–208%	98%	93%
Factor V Leiden mutation	41 (80%)	10 (20%)	Positive: 6 Negative: 35	N/A	N/A
Janus Kinase 2 V617F (JAK 2) Mutation	21 (41%)	30 (59%)	Positive: 3 Negative: 18	N/A	N/A
Duration of Anticoagulation	51 (100%)	0	Long Term: 45 Short Term: 5 None: 1	N/A	N/A

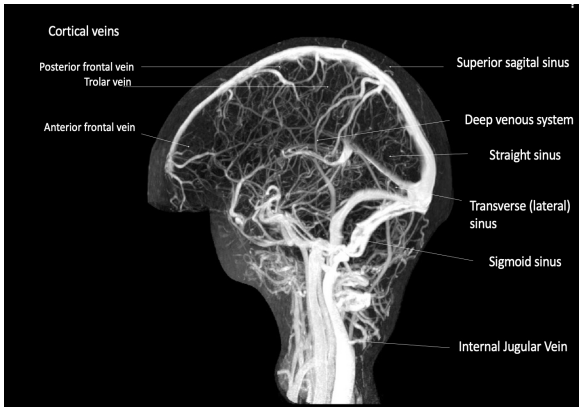

Figure 1. Picture showing various cerebral venous sinuses[1]

Conclusions and Limitations

- CVST was more common in females and the most common symptom at presentation is headaches.
- The average age group of affected individuals was 40 years.
- There were 2 patients who had vaccine-induced immune thrombotic thrombocytopenia (VITT) due to the J.J. COVID-19 vaccine.

The limitations of this study include it being retrospective; some records were incomplete/unavailable. For patients who did not have laboratory results at diagnosis available, we used results available from as close to diagnosis date as possible for analysis.

References and Acknowledgements

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