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Dural Sinus Thrombosis in Rwanda: An Observational Descriptive Study

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ABSTRACT

BACKGROUND: Dural sinus thrombotic (DST) events are common emergencies globally, yet awareness, even among health professionals, is limited. DST is devastating when the diagnosis is missed and/or the patients are inadequately treated. If the diagnosis is made and the patient is correctly treated, the results are generally good.
METHODS: We performed a retrospective descriptive study of a prospectively compiled database documenting management and outcome of 20 adult patients treated with DST at King Faisal Hospital (KFH) and Centre Hospitalier Universitaire de Kigali (CHUK) from March 2012 to March 2017.
RESULTS: Headache was the most common presenting symptom occurring in 14 patients (70%). The mean duration from presentation to diagnosis was 6 days (range: 2 to 21 days). There was one death with the 13 remaining patients (95%) responding well to treatment.
CONCLUSION: The delay in diagnosis of five days suggests that awareness of this condition is still low in Rwanda and that the diagnostic tools necessary to confirm this diagnosis continue to not be available in most medical facilities in the country. In order to achieve optimal results referral of patients suspected of having DST to a hospital with the imaging modalities is needed to confirm the diagnosis along with specialists who can treat this condition.

Keywords: Dural Sinus Thrombosis, Anticoagulation, Stroke, Venous Infarction, Africa, Rwanda

INTRODUCTION

Dural sinus thrombosis (DST) is a rare cerebral thrombotic event that represents diagnostic challenges [1]. Epidemiological data of this condition are lacking in Rwanda and in many other developing countries. In a nation-wide series from Portugal, Europe, DST was estimated to occur with an incidence of 0.22/100,000 annually (95% CI: 0-0.47) [2]. A report from Iran had a much higher rate of DST-- 1.23/100,000 per year [3]. Pregnancy seems to greatly increase the risk of DST [4].

The largest study currently available regarding DST is the prospective International Study on Cerebral Vein and Dural Sinus Thrombosis (ISDST), a cohort of 624 adults with DST. In this study, women comprised 75% of the 624 adult patients in the cohort, and they had a better prognosis compared to men. Compared with cerebral arterial events, DST occurs in younger patients with the mean age of patients in the ISDST study of 39 years [5].

In up to 85% of DST cases, a risk factor can be identified. Prothrombotic conditions, either genetic or acquired, are common in patients with DST. Head trauma, pregnancy and puerperium, infections, and dehydration are sometimes considered risk factors [5,6]. DST is often overlooked in many clinical settings due to its highly variable clinical presentation. Missing this diagnosis may result in devastating consequences. An autopsy study performed in the 1970s found a relatively high prevalence of DST (9%) [6]; but this might reflect the high mortality associated with untreated cases.

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Early recognition and intensive management of DST is crucial to improve morbidity and mortality. Three to five percent of these patients are likely to die in the first few hours of onset in the acute phase and 21% of patients are likely to deteriorate several days after admission [7]. Due to the recent availability of modern imaging modalities in Rwanda: Magnetic Resonance Imaging (MRI) and computed tomography (CT), DST is now more frequently diagnosed. Because of its myriad of causes and clinical symptoms, patients suffering from DST will initially be seen by internists, oncologists, hematologists, obstetricians, pediatricians, family practitioners and traditional healers before they are referred to a specialist service. Awareness and a high level of suspicion for this condition are of paramount importance for our patients.

Aims: This study sought to highlight the clinical challenges encountered in the diagnosis and management of patients with DST in Rwanda, with the specific objectives being to identify the unique clinical presentation of patients with DST; The diagnostic challenges; The management challenges and finally the outcomes of patients with DST over a period of 5 years from March 2012 to March 2017.

METHODS

Study design: We performed a retrospective descriptive study

Study setting: Patients treated at two referral hospitals in Kigali, Rwanda, from March 2012 to March 2017.

Inclusion and exclusion criteria: Inclusion criteria: Inpatients of all pediatric and adult age groups with Confirmed diagnosis of DST by imaging (MRV and /or contrasted CT brain); Exclusion criteria: Patients with cerebral arterial thromboembolic events and/or other causes of stroke other than DST.

Recruitment/enrolment of participants: All patients diagnosed with dural sinus thrombosis during study period were enrolled in the study.

How were outcomes measured: A questionnaire was used to collect data on outcomes at 6 months and 1 year after treatment. Headache severity using VAS score, Neurological function found on physical examination, recanalization of dual sinuses on MRI.

Data collection tools employed: A questionnaire was used to collect data about clinical presentation, treatment given to and outcome of patients.

Data collection: Data of a prospectively compiled database documenting the management and outcome of 20 patients with DST.

Patient follow-up: The patients were followed up for a mean period of 6 months after diagnosis. Magnetic resonance imaging (MRI) with MR angiography and venography was used in all patients for follow up.

Statistical analysis: SPSS was used to analyze data, by computing frequencies of presenting signs and symptoms, risk factors, imaging modality used, treatment given, complications and outcome

RESULTS

Participants: For the period of March 2012 to March 2017, 20 patients met the diagnostic criteria for cerebral vein and dural sinus thrombosis in our center. Among the 20 patients, 11 were female (55%), and all were adult patients (15 years and older) (Table 1). The mean age in our series was 35.

Table 1: Stable 1: Compelling Results of all patients

Clinical information		Frequency (n=20)	Percentage
Sex	Female	11	55%
	Male	9	45%
Mean Age		35 years	
Mean time for Diagnosis		5.75 days	
Signs/Symptoms	Headache	14	70%
	Raised ICP	7	35%
	Photophobia	6	30%
	Seizures	5	25%
	Hemiparesis	5	25%
	Cranial nerve palsy	3	15%
	Loss of consciousness	3	15%
Imaging used for diagnosis	MRV	19	95%
	CT- Scan	3	15%
Risks factors	Pregnancy and puerperium	4	20%
	Contraceptives pills	2	10%
	Coagulopathy	1	5%
	Severe dehydration	2	10%
	Brain trauma	4	20%
	No risk factor identified	8	40%
Treatment	LMWH and warfarin	16	80%
	Factor X inhibitor (Xarelto)	3	15%
	LMWH	1	5%
Outcome	Good recovery	19	95%
	Death	1	5%
Complications	Retinal detachment	1	5%
	Tissue necrosis	1	5%

Presenting symptoms: common presenting symptoms identified in our participants: Signs of raised ICP in seven patients (35%), photophobia in six patients (30%), seizures in five patients (25%), hemiparesis in five patients (25%), diplopia in three patients (15%) and low

level of consciousness in three patients (15%). There was a mean delay of 5.8 days from the time the patient sought medical help until the diagnosis was made (range 2 to 21 days).

Risk Factors: Eleven of our 20 patients were women. Four (20%) of the female patients in this study were either pregnant or post-partum and two of them were taking oral contraceptives. Head trauma was identified in 4 patients (20%) as a risk for DST. Patients' genetic and/or acquired pro-thrombotic conditions (Table 1) and oral contraceptive drugs were the 2 most common risk factors in the ISDST.

Investigations: The diagnosis of DVT is difficult to make on clinical grounds. Contrast brain CT scan was the first diagnostic imaging test in all cases. It showed features suggestive of sinus thrombosis in only 3 (15%) cases. MRV was diagnostic in all 19 cases in which it was use. MRV was not performed in one patient because of difficulties in positioning due to pregnancy.

Management: Anticoagulation therapy with LMWH and warfarin was the management in 16 patients (80%). A Factor X inhibitor rather than LMWH plus warfarin was used in three patients (15%), and one patient was treated with LMWH only. Most patients (except 1) did not have any complications associated with this treatment. Only one patient had a complication related to anticoagulation. This male suffered intramuscular hematomas and muscle necrosis—a rare complication of warfarin [8].

Early recognition and prompt management of DST is generally associated with a good outcome; 19 of our 20 patients (95%) had a favorable outcome. All patients were followed for a period of 12 months and only 1 had an early recurrence of symptoms.

DISCUSSION

Our mean age of 35 was the same as in the ISDST study [5]. Headache is the commonest symptom in all series of DST. It was present in 89% of patients in the ISDST study [7] and 70% of our patients. One of our patients presented with a headache as his only symptom and even though he presented late he had a good outcome. Patients who present late with isolated headaches tend to do well [9].

The diagnosis of DST is challenging. Our experience highlights the challenge of diagnosing DST in a resource limited setting. Clinical and brain CT findings are not sufficient to reliably diagnose DST and MRV is infrequently available.

Tests for protein C and S deficiency as well as factor V Leiden and antithrombin III deficiency are not available in Rwanda. However, for all patients where other risk factors were not apparent, blood samples for these tests have been sent to Nairobi. In addition, 2 of our patients had severe dehydration, a risk factor for venous thrombosis. In 40% of our patients no risks factors were identified.

Paradoxically deterioration and worsening of symptoms is more frequent in patients diagnosed early (23%) compared to those diagnosed late (8.3%). The relatively less severe course in late presenters is attributed to the relatively benign nature of DSTs that are associated with mild symptoms [9]. In our series, one patient, with an ''early'' (2 days from symptom onset) DST diagnosis deteriorated. Her deterioration was mainly due to acute raised intracranial pressure caused by an intracerebral hemorrhage (pontine bleed) causing an obstructive hydrocephalus. Her neurological symptoms initially improved on surgical management), but she died from septic shock.

Limitations: This study has several limitations. Most importantly it reflects only patients seen at two of the teaching hospitals in Rwanda that possess one advanced diagnostic modality (magnetic resonance venography). We therefore postulate that this condition is underdiagnosed in Rwanda. Other reports indicate that DST is more common in children than in adults and this study did not include children. Despite these limitations, once the diagnosis of DST is made the treatment is straightforward and the outcome is generally good [10].

CONCLUSION

Dural Sinus Thrombosis is rarely diagnosed in countries with limited resources. An increased awareness of the possibility of this condition and prompt referral to institutions with advanced diagnostic capabilities and specialists (neurologists and neurosurgeons) experienced in managing these patients are needed to optimize outcomes.

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