

Speech-Language Profile, Neurological Findings and Recovery Patterns Associated with Cerebrovenous Sinus Thrombosis

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Abstract

Cerebrovenous sinus thrombosis (CVT) is a rare neurological condition which is difficult to diagnose because of the varied neurological presentation. The pattern of speech-language findings and recovery patterns in CVT can be quite different from those seen associated with arterial stroke. In converse to arterial stroke, scarce information exists on the natural history and long-term prognosis of CVT. This is particularly true with respect to the pattern of speech and language deficits associated with this condition. The present study profiles the speech-language and recovery patterns in seven patients with cerebrovenous sinus thrombosis.

Key words: Speech-language, neurological, recovery, cerebrovenous sinus thrombosis

Introduction

Cerebro venous sinus thrombosis (CVT) is a rare and potentially deadly condition. Known conditions that increase the risk of CVT include hypercoagulable states, dehydration, adjacent infectious processes, low cerebral blood flow, oral contraceptives, hormone replacement therapy, pregnancy, and puerperium. Each of these conditions is associated with a higher risk of venous thrombus formation, but exactly why the cerebral venous sinus system is involved over other veins is unclear [1]. Diagnosis is difficult because of various and nonspecific clinical presentations. However, diagnosis must be established quickly in order to start anticoagulant treatment as early as possible. CVT is an infrequent condition that is extremely variable in its clinical presentation, mode of onset, imaging appearance, and outcome.

In contrast to the arterial stroke, which can be easily diagnosed clinically in majority of cases, CVT has no single pattern of presentation and it may be difficult to diagnose it on clinical grounds alone [2]. Clinical presentation may be acute (within 48 hrs), subacute (bt. 2 days and one month) and chronic (>30days). Subacute or chronic presentation are commoner than in arterial stroke and progress in gradual or stepwise fashion is common. When draining venous sinuses are occluded pressure must build up in feeding arteries to maintain perfusion and this results in oedema, infarct and haemorrhage leading to different focal neurological signs like hemiparesis, aphasia, ataxia, hemianopia, neglect etc. Haemorrhage may be bilateral if obstruction is in Superior sagittal sinus (SSS). Clinical profile is determined by a) underlying sinus/ venous system involved; b) mode of onset, i.e., acute, subacute or chronic; c) time interval between onset of disease and clinical presentation; d) nature of primary disease giving rise to CVT. Cortical deficits like aphasia, agnosia, apraxia, and cortical blindness are not uncommon but are fleeting in nature.

Speech disturbance has been mentioned in superior sagittal sinus thrombosis with a small venous infarct [3], occlusions of the lateral sinus present usually as an isolated intracranial hypertension, associated with aphasia when the left transverse sinus is occluded [4], sigmoid sinus thrombosis presenting with posterior alexia [5], neurological impairments such as limb paralysis and language disorders may or may not be present [6], acute micrographia and hypophonia as the sole manifestations of extensive deep venous sinus thrombosis [7], slurred speech and right sided clumsiness [8] slurring of speech, hoarseness, nasal twang of voice and difficulty in swallowing with nasal regurgitation, difficulty in moving tongue side-to-side, arching of palate to left side, reduced palatal movements on right side, tongue deviation to right side, tinnitus of two weeks and unilateral hearing problem [9], neurological deterioration with right hemiparesis, dysarthria, global aphasia, and depressed levels of consciousness [10]. Few studies have looked at the long-term speech and language outcomes in patients with CVT. Studies using various neuropsychological batteries aimed at assessing long-term cognitive outcome have reported cognitive impairment in approximately 35% of the patients in different cohorts usually in the form of non-fluent aphasia, working memory deficits and depression [11,12].

Although CVT causes stroke far less frequently than arterial pathologies [13,14], it has a predilection for younger individuals, particularly women of childbearing age. Its clinical evolution seems to be different from the other stroke subtypes and is highly variable between the studies [15]. The extent of functional recovery in survivors of CVT is better as compared to patients with arterial thrombosis in which the proportion of permanent dependent patients ranges between one third and two third of survivors²⁰ which is in contrast to CVT in which an independent survival of around 80% is commonly found.

In converse to arterial stroke, scarce information exists on natural history and long-term prognosis of CVT. This is particularly true with respect to the pattern of speech and language deficits associated with this condition. Few preliminary reports often are limited by small numbers, their retrospective nature, and short follow up. These studies mention about the presence or absence of a speech disorder and /or aphasia without any further details regarding the nature and recovery patterns. There is dearth of studies on the nature and type of voice, speech and language disturbances associated with cerebrovenous sinus thrombosis. The objective of the proposed research is to profile the speech and language disorders associated with cerebrovenous sinus thrombosis in patients admitted to the stroke ward in department of Neurology at NIMHANS.

Clinical findings

Patient 1

A 42 year old man presented with h/o altered sensorium and headache since 7th August 2013 with two episodes of seizures. Paucity of movements on the right side. No detectable cranial nerve abnormality. Motor weakness improved over two days. Patients sensorium improved gradually and was verbalizing by 8th August 2014. He was a chronic alcoholic for last six years, but there were no symptoms of alcohol withdrawal. CT brain showed multiple haemorrhages including bilateral hemispheres and hyperdense lesion in right caudate subcortical region and left parietal region. MRI brain shows haemorrhages in right temporal and left parieto-cortical regions- resolving bleed in right caudate and left thalamic regions. He was managed with anticoagulants, antiedema measures. Showed significant improvement in sensorium and motor weakness within next two weeks. Oral mechanism examination on 14th August 2014 revealed normal structure and function. Western Aphasia Battery (WAB) score of 8.5 revealed anomic aphasia.

Language in India www.languageinindia.com ISSN 1930-2940 14:12 December 2014

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Patient 2

A 30y old female presented with four day history of generalized headache on 1st August 2013. One day h/o progressive decrease in sensorium associated with paucity of movements in left half of the body and left UMN facial palsy. On admission, patient could obey simple commands. Patient had significant improvement in sensorium and motor power by 3rd August 2013. CT brain revealed hypodensities in bilateral thalamus and hyperdense straight sinus, basal vein of Rosenthal, internal cerebral veins and thalamostriate vein. MRI brain revealed subacute thrombosis of right internal cerebral vein and basal vein of Rosenthal. Patient was diagnosed as having supratentorial lesion ? cerebral venous thrombosis ? mass lesion. The patient was managed with anteedema measures and anticoagulants. The MMSE score as on 4th August 2013: Orientation-7, registration-3, recall-2, calculation-1, naming-2, three step commands-3, reading-0, writing-1, copying-1, repetition-1.- Total 21. No significant difficulty in oromotor abilities and Western aphasia battery evaluation revealed absence of aphasia.

Patient 3

22 year old male reported on 31st July 2013 with history of headache, vomiting and seizures since three days. H/o altered sensorium and right sided weakness. CT brain revealed mixed density lesion of temporo-parietal region with mass effect and midline shift. MRI revealed extensive left temporoparietal hemorrhagic infarct with subacute thrombosis of left transverse sinus. A diagnosis of left temporoparietal CVT was made. Decompressive hemicraniectomy was done on 1st August 2013. Speech and language evaluation revealed mild dysarthria with Global aphasia. Follow up evaluation on 7th October 2013 revealed persisting minimal comprehension deficits and second follow up on 25th November 2013 reported mild naming difficulties.

Patient 4

30yr old lady postpartum on day five presented with history of irrelevant speaking in the form of asking for food after taking food. She also presented with left unilateral moderate headache with nausea and episodes of vomiting. Could not indicate needs and was not verbal. CT brain revealed mixed density lesion in the left frontotemporoparietal region with mass effect and 0.7cm midline shift and mild hydrocephalus. A diagnosis of left frontotemporal

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cerebral venous thrombosis was made. Left frontotemporoparietal craniectomy and lax duroplasty was done on 25th July 2013. Speech and language evaluation on 12th August 2013 revealed mild dysarthria with resolving global aphasia. Re - evaluation on 19th August 2013 revealed the presence of minimal naming difficulties.

Patient 5

20 year old female reported on 19th July 2013 with history of right hemiplegia after delivery two weeks back, headache, vomiting, altered sensorium and weakness of left side. CT brain showed moderate diffuse cerebral edema with right frontal hypodensities. A diagnosis of CVT postpartum stroke was made. Speech and language evaluation on 27th July 2013 revealed clinically normal speech and language status.

Patient 6

18 year old female reported on 8th June 2013 with weakness in right side and h/o vomiting and altered consciousness since a week. CT scan revealed hypodensities in bilateral thalami and MRI showed edema in bilateral thalami, caudate nucleus, left lentiform nucleus and bilateral deep white matter with early venous infarct in left thalamus. A diagnosis of left transverse sinus thrombosis causing vasogenic edema with early venous infarct in left thalamus was made. C/o weakness in the Rt side of the body since 08/06/13. Patient delivered a child on 30th May. She was apparently normal till 08/06/13 when she developed vomiting and altered consciousness. Taken to a local hospital where MRI: edema in bilateral thalami and caudate nucleus, left lentiform nucleus and bilateral deep white matter with early venous infarct in Lt thalamus. Features suggestive of Lt transverse sinus thrombosis causing vasogenic edema with early venous infarct in Lt thalamus. Speech and language evaluation on 10th July 2013 revealed the presence of global aphasia with oropharyngeal neurogenic dysphagia. WAB results on 15th July 2013 revealed recovery of language disturbances and the presence of anomic aphasia. Frenchay dysarthria evaluation revealed no significant oromotor difficulties.

Patient 7

32 year old female presented with history of hemicranial headache and vomiting since six weeks. CT brain revealed acute right temporoparietal bleed with perilesional edema, midline shift of 13.3mm and dilatation of temporal horn. A diagnosis of right temporoparietal

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hemorrhagic infarct and right parietal decompressive craniectomy was done on 27th July following which there was significant improvement in sensorium. Speech and language evaluation done on 29th July 2013 revealed Wernicke's aphasia with oropharyngeal neurogenic dysphagia. This resolved to anomic aphasia with near normal oromotor skills and absence of dysphagia in a weeks time.

Discussion

This study profiles the speech-language and recovery patterns in seven patients with cerebrovenous sinus thrombosis. Five out of seven patients had noticeable speech-language difficulties. In most of the patients the speech language difficulties recovered within a span of two weeks to one month after the onset of CVT. Near total recovery with respect to speech-language abilities was observed in all five patients. A younger age of onset could be observed in patients with CVT compared to patients with arterial stroke reported in literature.

The age range of the patients with CVT was 18-42 years. There were two males and five females with CVT. In two females, CVT developed postpartum, whereas in others it was associated with other neurological symptoms. The lesions varied from left frontotemporo-parietal region, left transverse sinus thrombosis to acute right temporo-parietal haemorrhagic infarct.

Recovery of speech-language skills happened within two weeks to one month following CVT in most of the patients. In majority of the patients with CVT and associated speech-language difficulties (Pt 1, Pt 3, Pt 4, Pt 6, Pt 7), the aphasia type was global immediately post CVT and then recovered to anomic pattern in an interval of two weeks to one month. Language disturbances have been frequently reported in patients with CVT [6, 10]. Dysarthria was present in the acute phase which recovered to near normal oromotor abilities in most of the patients during follow up evaluations after a month. Presence of slurred speech and oromotor difficulties associated with CVT is also frequently reported [8-10]. Only two patients (Pt 6 and Pt 7) with left transverse sinus thrombosis and right temporoparietal haemorrhagic infarct respectively had dysphagia during the acute phase. Both patients had neurogenic dysphagia including the oral as well as the pharyngeal phase. Dysphagia also recovered within a two week to 1 month interval. Detailed evaluation of reading and writing skills could not be done during the acute phase of evaluation.

Language in India www.languageinindia.com ISSN 1930-2940 14:12 December 2014

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Conclusion

Dysarthria, aphasia and dysphagia are commonly associated with CVT. In comparison to arterial territory strokes, the speech-language and swallowing difficulties subsequent to CVT, show a rapid recovery pattern with near total recovery within a span of two weeks to one month. Occasionally persisting difficulties with higher level language comprehension and dyslexias may persist. Follow up evaluation of these patients is necessary to identify subtle language difficulties in the later phase of the condition.

Acknowledgements

I thank the Director, National Institute of Mental Health and Neurosciences, Bangalore for all support and for providing necessary infrastructure. I thank all the participants in the study.

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Language in India www.languageinindia.com ISSN 1930-2940 **14:12 December 2014**

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