Cerebral hemi atrophy as a consequence of cerebral insult in infancy: Is it difficult to diagnose?

Sir,

There are a variety of causes that lead to cerebral hemi atrophy with resultant complications, ranging from poor physical growth, hormonal abnormalities, mental retardation, seizures, and hemiplegia. Dyke, Davidoff, and Masson published a series of 9 cases in 1933 presenting with hemiparesis, seizures, facial asymmetry, and mental retardation and attributed to cerebral asymmetry on plain skull radiographs and pneumatoencephalographs. They showed thickening of calvarium, dilatation of frontal and ethmoid sinuses, and elevation of greater wing of sphenoid and petrous ridge. This condition is known as Dyke-Davidoff-Masson Syndrome (DDMS). Subsequent to this publication, several reports followed explaining the different types of cerebral atrophy and the distinction between atrophy and hyperplasia. Some case reports in children as well as in adults too were reported. Brain insult from a variety of causes from foetal stage to early infancy can result in cerebral hemi atrophy. The causes are either congenital, i.e. idiopathic (Primary) and intrauterine vascular injury or acquired, i.e. perinatal intracranial hemorrhage, infection (Encephalitis), trauma, vascular abnormalities (Sturge-Weber’s Syndrome), ischemia, hypoxia, etc. When the cerebral volume reduces in early stages, it may lead to changes in the skull, which is apparent in skull radiographs. Magnetic resonance imaging (MRI) reveals changes in the brain parenchyma with thinning of grey matter, reduced volume of white matter, enlarged lateral ventricle, reduced size of cerebral peduncles (ipsilateral), and reduced size of cerebellar hemisphere. We report a case of recurring seizures in a young 22-year-old female who presented to psychiatry outpatient in a tertiary care hospital and was diagnosed with cerebral asymmetry that could be traced to a cerebral insult in infancy.

A 22-year-old female was brought to the psychiatric OPD by her mother with the complaint of behavioral problems, recurring seizures, and weakness of right upper and lower limbs. Detailed history revealed that the patient was born at full term through vaginal delivery without any trauma/birth asphyxia. Birth weight recorded was 2300 g. At 3 months of age, the child suffered high-grade fever, had convulsions, and was admitted to the hospital for a month. She was suspected to be a case of encephalitis. Although the child recovered from fever and febrile convulsions, the milestones were delayed and speech was affected. Later, an IQ assessment showed moderate mental retardation (IQ 30-49). She had to be cared for her daily needs and had to be constantly attended to. As she grew up, seizures returned at regular intervals and she was put on anticonvulsants by the neurophysician. She started showing behavioral changes in 2004 (at 14 years of age) in the form of irritability, aggressiveness, and violence while on anticonvulsants Tab Sodium Valproate 300 mg BD and Tab. Phenytoin sodium 100 mg three times a day. She was put on low doses of atypical antipsychotic tab Risperidone 1 mg once a day and small dose of benzodiazepine (Clonazepam 0.25 mg/day). She remained symptom free. Frequency of seizures reduced considerably to once in 3-4 months. General physical examination was normal. Her gait was hemiparetic type with motor power grade III in right upper and lower limbs. All hematological and hormonal (T3, T4, TSH, FSH, LH) investigations revealed no abnormality. MRI brain done on 12/10/2012 revealed thickened skull vault with thickened diploic spaces. Mild prominence of the sylvian fissure sulcal spaces on the left side indicated hemi atrophy of the brain. Mild prominence of left lateral ventricular system was noted. Midline structures were maintained. Cerebellum and brain stem appeared normal [Figure 1].

From the foregoing, it is apparent that the patient suffered a febrile illness, which was labeled as encephalitis at a very vulnerable age of 3 months and affected the growth of the brain. The extent of the brain damage was revealed only through the MRI brain [Figures 2 and 3] when she presented with behavioral problems. It showed the reason for right-sided weakness and hemi paretic gait, seizure disorder, mental retardation, and difficulty in speech due to atrophy and reduced volume of brain with thin grey matter and reduced volume of white matter. Since her IQ fell in the range of moderately mentally challenged group, not much thought was given to investigate the real reason for this
The MRI findings of frontal lobe hemiatrophy together with seizures and mentally challenged state resembled the condition described by Dyke, Davidoff, and Masson’s series of 9 cases in 1933. The appearance in these cases showed thickening of calvarium, dilatation of frontal, and ethmoid sinuses, and elevation of greater wing of sphenoid and petrous ridge. The MRI findings in the above case almost conform to this condition. Solomon⁴ and Parker⁵ while reporting on the case studies explained the mechanism behind thickening of the skull vault. It is a fact that the brain grows to half of its adult size by the first year and reaches 75% of the adult size by three years. Any insult during these years will result in stunted growth of the brain and the skull vault grows inwards resulting in thickening of the vault, enlargement of frontal sinuses, and increased width of diploic spaces and greater wing of sphenoid and petrous ridge on the affected side. This is possible when the brain damage is sustained in the first few months of infancy, as in this case. This shows that DDMS although not a very common condition, it can present in varying degree of severity. What is important is to document a proper clinical history, a detailed clinical examination and investigations (CT, MRI, and Hormonal) to confirm the diagnosis of DDMS. Cerebral insult very early in infancy due to febrile illness (Encephalitis) seems likely to be the cause in this case.

Daniel Saldanha, Bushan Chaudhari, Suresh Kumar Mehta¹, Archana Narender Javadekar, Amit Kharat²

Department of Psychiatry, ¹Consultant in Psychiatry, ²Associate Professor Radiodiagnosis, Padmashree Dr. Dnyandeo Yashwantrao Patil Medical College and Research Centre, Pimpri, Pune, Maharashtra, India

Address for correspondence: Dr. Daniel Saldanha,
Flat No 1102, N Block, Grevillea, Magarpattacity,
Pune - 411 013 Maharashtra, India.
E-mail: d_saldanha@rediffmail.com

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Could do away with terms like ‘suicidal inclination’ or ‘morbid ruminations’ for suicide intent and ideation, respectively. Having these terms used or read widely in contemporary research encourages clinicians and trainees to use them interchangeably with little understanding. Alternatively, in my opinion, it may be beneficial to prepare a list of minimum essential elements for each suicidal construct. This is because suicide as a behaviour is studied and reported by many disciplines and each field may assign differential weightage to different components of suicidal behaviour. Hence, it may be more meaningful to have a list of common minimum elements that would serve as building bricks for definitions in suicidology. To give an example, any definition of intent would have to include intended outcome (death), agency (self) and subjective expectations about a chosen method resulting in intended outcome. Contemporary assessment guidelines for suicidal behaviour do not provide much clarity on how to assess these individual constructs, many of which play a role in determining future suicide risk.

A standardized evaluation scheme could be developed for suicide attempters with adequate provisions for individual variations along the lines of what has been developed for catatonia. A strong nomenclature would improve the quality of clinical documentations and data collection leading to meaningful suicide research, improved clinician communication, and better extrapolation of findings due to increased validity.

Vikas Menon
Department of Psychiatry, Jawaharlal Institute of Post Graduate Medical Education and Research, Puducherry, India

Address for correspondence:
Dr. Vikas Menon,

Sir,

Suicide is a multi-dimensional human behaviour that may have many determinants. Due to this complexity, perhaps, one can find at least 15 commonly referenced definitions of suicide in literature. With regard to the issue of definitions for attempted suicide, the waters are even muddier with a plethora of terms such as ‘parasuicide’, ‘non-fatal suicidal behaviour’ and ‘deliberate self-harm’ being used synonymously by researchers. In an attempt to clarify this aspect, Silverman et al. state that unless the intent to end one’s life is established in an episode of self-injurious behaviour, clinicians should desist from calling it a suicide attempt. It is questionable, though, to what extent these recommendations have penetrated among clinicians as well as the non-medical community, many of whom interact with suicidal clients. Often, we encounter scenarios where definitions related to suicide are used without rigour or differently interpreted according to the author of the paper under consideration. One possible reason for this confusion is the difficulty one faces in assessing the individual elements of contemporary suicide definitions, especially the intent. These may include the ambivalence of the individual in talking about the act and the need to magnify or minimize intent so as to meet some objective. Additionally, as most suicide attempts are assessed retrospectively, reliance on patient self-report and the judgement of the clinician in establishing suicide intent is much greater. As a result of these practical difficulties, it is often observed that episodes of self-harm end up getting mislabelled as suicide attempts without due diligence in establishing the intent and lethality involved in the act.

There are few suggestions that can be considered, to modify this existing scenario. For example, we Silverman revisited: A relook at some of the pitfalls and challenges in suicide nomenclature and few suggestions