Sir,

We read the article by Ola et al on Dyke-Davidoff-Masson syndrome published in January 2014 issue of the journal and found it very interesting. Highlighting such cases is important as it leads to increased understanding of this rare syndrome. We would like to report a similar case with few interesting variations from the case described in the journal.

A 16 year old Hindu male was admitted in our institute for evaluation of altered sensorium and abnormal body movements in the form of generalised tonic-clonic seizures. Initial examination revealed increased tone, exaggerated tendon reflexes and extensor plantar response on the left side of body with normal sensory, cranial nerve and cerebellar functions bilaterally. Examination of cranium and spine did not show any abnormality. Past history was suggestive of seizure disorder since the age of 6 years and the patient failed to respond to adequate antiepileptic drug therapy. Birth and developmental history was significant and suggestive of birth complicated by meconium aspiration syndrome and delayed motor and psychosocial milestones. Patients’ mother also gave history of recurrent episodes of violent behaviour, bizarre posturing followed by few depressive episodes and frequency of these events were increased during last few months. MRI scan of the brain showed asymmetry of anterior cranial fossa with thickening of calvarium and hyperpneumatisation of frontal sinus, cerebral atrophy and ventricular dilation all on the right side. All the findings favoured diagnosis of Dyke-Davidoff-Masson syndrome.

Significant contrasting feature in our patient than the patient described in the journal was the presence of left sided cerebellar atrophy (Figure 1). Dyke-Davidoff-Masson syndrome associated with crossed cerebellar atrophy (CCA) has been previously described in some patients. There are two schools of thought regarding occurrence of CCA, (1) the underlying pathogenesis of CCA may involve damage to the cortico-ponto-cerebellar tracts or centrolobular cerebellar sclerosis due to effects of repeated episodes of seizure, (2) but according to a survey done in patients with unilateral precocious destructive brain insults, the extent of the supratentorial lesion and the

![Fig. 1: Showing atrophy of the left cerebral and right cerebellar hemispheres known as crossed cerebellar atrophy (CCA).](image-url)
antecedent history of status epilepticus but not the recurrent seizures are related to the development of CCA.²

Another rare feature seen in our patient was history of abnormal behaviour consistent with schizoaffective disorder which was refractory to psychotropic medications. This is one of the rarest complications associated with this syndrome.

Another unusual but interesting finding in our patient was seen on EEG brain which showed focus of seizure activity from left cerebral hemisphere. The only available explanation of this finding in the standard medical literature is by Teixeira RA el al.³ They studied 51 patients with precocious brain insults and found that 26 had background EEG abnormality, 5 patients showed discoradance in EEG localisation. They also performed ictal SPECT and confirmed that the finding of false localisation was due to background EEG abnormality. The EEG discoradance in localisation of seizure focus on normal side seen in our case could also be explained by the presence of background EEG abnormality rather than skull changes.

Furthermore, minor structural alterations of the brain are important as they can alter the clinical picture of these patients. Therefore, an attempt should be made to describe the presence of such findings in a patient of Dyke-Davidoff-Masson syndrome. Shen et al. depicted three MR imaging patterns of cerebral hemiatrophy: MR imaging pattern I corresponds to diffuse cortical and subcortical atrophy; pattern II corresponds to diffuse cortical atrophy coupled with porencephalic cysts: and pattern III corresponds to previous infarction with gliosis in the middle cerebral artery (MCA) territory.⁴ CT angiography in our patient showed decreased perfusion in right middle cerebral artery territory. Accordingly, in our patient, pattern III was present which corresponds to early development of hemiparesis. Our patient developed hemiparesis before 2 years of age. Presence of early hemiparesis portends poor prognosis in these patients. On the contrary, our patient is doing fairly well on antiepileptic drugs.

References

Reply from Author
BS Ankit*, AK Gadhwal*, P Sirohi**, RP Agrawal***
Sir,

I am highly thankful to Dr Bharat Bhushan Sharma for showing keen interest in our article. The uncommon presentation of Dyke-Davidoff-Masson syndrome (DDMS); association of crossed cerebellar atrophy and Shizoaffactive disorder brought about by him in the letter is appreciated.

Classical radiological findings of DDMS asymmetry of cerebral hemispheric growth with atrophy on one side, ipsilateral osseous hypertrophy and hyperpneumatisation of sinuses are present in variable degrees according to the extent of disease. Age of presentation depends on time of neurologic insult and characteristic changes may be seen only in adolescence. These changes can occur only when brain damage is sustained before 3 years of age; however, it may become evident as early as 9 months after the injury. Both sexes and any of the hemisphere may be affected, but male gender and left side involvement are more common probably of vascular origin involving left middle cerebral artery. Clinical presentations of DDMS include variable degree of facial asymmetry, seizures, contralateral hemiparesis, mental retardation, learning disabilities, impaired speech, etc. Seizures can be focal or generalised. Complex partial seizure with secondary generalisation is very rare.
Children with intractable disabling seizures and hemiparesis before 2 years of age indicating poor prognosis, are the potential candidates for hemispherectomy with a success rate of 85% in carefully selected cases.

Recently, we encountered a patient of DDMS associated with mesial temporal sclerosis (MTS).

An 18 year old female already on antiepileptics presented to us with 8 episodes of seizures of mixed types (GTCS and Complex Partial Seizure with secondary generalisation). In history, the patient born of non-consanguineous marriage with uneventful perinatal history, had normal motor milestones and delayed learning skills and social developmental milestones with poor school performance. She was asymptomatic till 12 years of age when she had her first episode of GTCS. She had facial asymmetry, left sided brisk deep tendon reflexes, and extensor plantar response. Vision and hearing were normal and cranial nerves were intact. MRI of brain revealed right cerebral hemiatrophy, asymmetry of Bilateral lateral ventrical with right ventrical prominence, Asymmetry of Bilateral Hippocampus region with relatively smaller and hyperintense Left sided hippocampus suggestive of Dyke-Davidoff- Masson Syndrome (DDMS). MRI Brain also showed asymmetry of Bilateral Hippocampus region with relatively smaller and hyperintense Left sided hippocampus suggestive of Mesial Temporal Sclerosis with B/L cerebellar atrophy. All these features lead to the diagnosis of Dyke-Davidoff- Masson Syndrome with Mesial Temporal sclerosis (Figure 1).

Association of DDMS with MTS is extremely rare. Co-occurrence of these two diseases has not been documented. Although the etiology of MTS remains controversial, there is now a considerable amount of evidence demonstrating that MTS is both a result and a cause of seizures. Clinical studies suggest that prolonged seizures or complicated febrile seizures may result in MTS. The mechanism of the lesions is due to excessive excitability secondary to release of excitatory amino acids, primarily glutamate. Glutamate, acting at a number of subreceptors on the postsynaptic membrane, leads to prolonged depolarization of neurons and results in the entry of cytotoxic amounts of calcium.