Dyke-Davidoff-Masson Syndrome: Time to Revisit Case Series

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Sir

We read a case report by Manghera et al (JAPI, Vol 62 page No. 76-67), which was in response to an earlier case report by Ola et al.¹ Here we would like to share our experience and views as under-

The authors in the correspondence have said that crossed cerebellar atrophy is an unusual and rare finding. We in our study of 28 patients of DDMS, have found cerebellar atrophy in nine patients along with cerebral atrophy.² Out of nine patients with cerebellar atrophy three patients had diffuse bilateral atrophy, only one patient had unilateral cerebellar atrophy which was controlateral to left cerebral hemiatrophy (CHA). This is similar to the findings of the authors and the patho-physiology quoted by authors is worth appreciation. Other parenchymal changes observed in our study were cerebral peduncle atrophy in three patients, and thalamic atrophy with lentiform nucleus hypoplasia in 11 patients. Seven cases of CHA were associated with ipsilateral large schizencephalic cleft with absence of the septum pellucidum whereas two had porencephaly. Five patients had left-sided hippocampal sclerosis (HS), four were concordant and one was discordant.

The EEG disconcordance in localization of seizure focus on normal side seen in the case under correspondence could be explained by the presence of background EEG abnormality rather than skull changes, as we have observed background EEG abnormality in about two-third of our patients there was disconcordance in EEG findings, so probably it requires further insight.

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

The authors of earlier case report have observed mesial temporal sclerosis in an 18 year old female of DDMS, which was described to be extremely rare and not documented earlier. We in our case series found hippocampal sclerosis in five patients as mentioned earlier, so probably it is not extremely rare, but an overlooked entity. As authors have mentioned, that the etiology of MTS remains controversial, and there is now a considerable amount of evidence demonstrating that MTS is both a result and a cause of seizures, which explains it’s incidence.

We observed some other findings like dystonia, hemiparkinsonism, mirror movement and Dandy Walker syndrome (DWS).

In our study we also observed that phenytoin adverse effects were significantly (P < 0.03) associated with DDMS/HHE patients.

We concluded our study with remarks that, DDMS/HHE can present protean clinic radiological manifestation, cerebellar atrophy, hippocampal sclerosis and phenytoin intolerance.

References
