ORIGINAL RESEARCH PAPER

Volume-8 | Issue-3 | March-2019 | PRINT ISSN No 2277 - 8179

INTERNATIONAL JOURNAL OF SCIENTIFIC RESEARCH

ACQUIRED DYKE-DAVIDOFF-MASSON SYNDROME PRESENTED WITH CROSSED NON-APHASIA

Neurology

Ritwik Ghosh — Post Graduate Trainee, Department of General Medicine, Burdwan Medical College and Hospital, Burdwan.

Arpan Mandal* — Associate Professor, Department of General Medicine, Burdwan Medical College and Hospital, Burdwan . *Corresponding Author

Devlina Roy — Post Graduate Trainee, Department of General Medicine, Burdwan Medical College and Hospital, Burdwan.

Subrata Biswas — Post Graduate Trainee, Department of General Medicine, Burdwan Medical College and Hospital, Burdwan.

ABSTRACT

Dyke-Davidoff-Masson syndrome (DDMS) is a rare disease which is clinically characterized by hemiparesis, seizures, facial asymmetry, and intellectual disability. The classical radiological findings are cerebral hemiatrophy, calvarial thickening, and hyperpneumatization of the frontal sinuses. This disease is a rare entity, commonly congenital and rarely acquired and it mainly presents in childhood.

"Crossed non-aphasia", a distinctly rare and interesting finding, is characterized by normal linguistic skills in a right-handed subject following damage to the left hemispheric linguistic cortex. However, these subjects show deficits in visuospatial skills, usually processed by the right hemisphere.

In this report, we discuss a subject who presented with "crossed non-aphasia", a clinical manifestation that often could go unnoticed, again in a rare case of DDMS with atypical radiological findings.

KEYWORDS

Dyke-Davidoff-Masson syndrome, cerebral hemiatrophy, Calvarial thickening, crossed non-aphasia, atypical radiological findings.

INTRODUCTION:

Dyke-Davidoff-Masson syndrome (DDMS) is a rare disease comprising hemiparesis, seizures, facial asymmetry, and mental retardation [1]. The classical findings including cerebral hemiatrophy along with calvarial thickening and hyperpneumatization of the frontal sinuses are only found if an insult to the brain occurs before 3 years of age [2]. The major concern of the disease remains the intractable seizures for which drug therapy is not sufficient in most of the cases, and a surgical approach is necessary. However, if the patient presents later in life, the disease manifestation may not be similar to that seen in childhood, and management changes accordingly.

Anomalous lateralization of linguistic functions is observed in a small group of right-handed subjects with unilateral brain damage as either crossed aphasia (aphasia after right-hemisphere damage) or crossed non-aphasia (left hemisphere damage without aphasia but with symptoms of right hemisphere damage such as visuospatial deficits). The incidence of crossed non-aphasia is reportedly far less than that of crossed aphasia. In this report, we discuss a subject who presented with "crossed nonaphasia," with an extensive left hemispheric lesion following an adult onset DDMS.

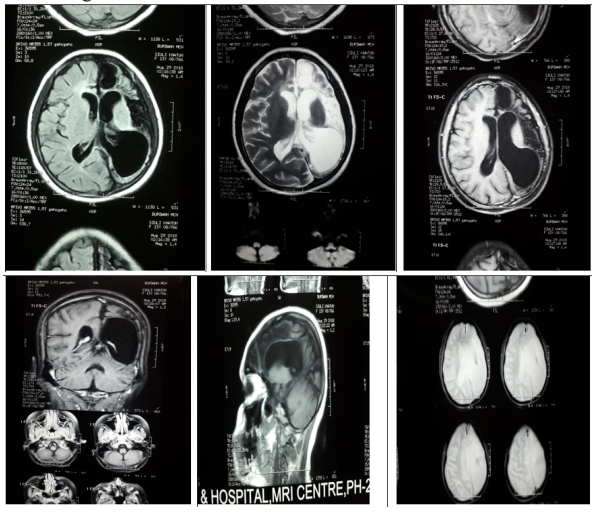
CASE REPORT:

A 13-year-old girl presented to the emergency department with a history of right focal motor seizures with occasional secondary generalization. After stabilization of vitals and controlling seizures, detailed history of the illness was taken which revealed she was born out of a nonconsanguineous marriage, without any antenatal or perinatal complications. She had normal developmental milestones till fifth year of life and always preferred to use her right hand over the left one while doing uni-manual tasks.

After that, she had severe febrile illness with seizures at the age of 5 years, for which she was treated in a local hospital as a case of meningoencephalitis, and she was discharged subsequently after 3 weeks on antiepileptic medications. However, she continued to have seizures. Besides, the parents also noted subtle learning difficulties, slurred speech, facial deviation, and progressive right sided weakness. Her seizures did not respond to several antiepileptic medications in different combinations, and, at the time of presentation, the patient was on triple antiepileptic drugs without any episode of breakthrough seizures for the past three years. Notwithstanding, the patient's hemiparesis had worsened gradually with increased stiffness in the right extremities and poor dexterity of the right hand. The patient had also complained of diminished vision of the right eye 2–3 years previously, which was later established as homonymous hemianopia of the right eye field on clinical examination and visual field charting.

On detailed neurologic examination, she was found to be conscious, alert, and oriented to person and time, but not to place. She cooperated well with the examination procedure. The cranial nerve examination revealed a right upper motor neuron facial palsy. Remaining cranial nerves were normal. The motor examination showed right hemiparesis. The muscle strength in the upper and lower limbs was decreased (UL weaker than LL). There were no sensory deficits. The handedness was assessed by means of the Edinburgh Handedness Inventory (Oldfield, 1971) and it revealed a strong right-handedness (100%). She did not have any left-handed primary relatives.

A magnetic resonance imaging (MRI) of the brain was subsequently done, which revealed (see images below) almost complete atrophy of the left cerebral hemisphere without an enlarged frontal sinus and thickening of the calvarium on the same side.



Subsequent to the above investigations, though the patient as well as the caregivers did not report any apparent linguistic disturbances, we decided to examine her linguistic skills as she was right-handed and

exhibited an extensive left-hemispheric lesion, to find out any subtle linguistic deficits. A detailed speech and language examination was carried out at. For linguistic examination, the "Bangla" adaptation of the Western Aphasia Battery (WAB) (Kertesz, 1982) and, for speech evaluation, the Frenchay Dysarthria Assessment (Enderby, 1983) was used. The linguistic evaluation showed clinically normal language skills. The scores of the Western Aphasia Battery (Aphasia Quotient—AQ) on various subtests were determined and praxis subsection of the WAB was administered to find out the presence of apraxia. However, she could perform all the tasks flawlessly. The administration of FDA revealed mild deficits at the lip and tongue sections. The speech intelligibility was fair at the word level and minimally imprecise at the conversational level. The minimal dysarthria observed were due to the minimal imprecision while producing the tongue-tip phonemes such as /t/, /d/, /s/, /ʃ/, /n/, and /l/. Considering the site of lesion, the dysarthria in P was unilateral upper motor neuron (UUMN) type.

On assessment of the visuospatial skills, she exhibited difficulty drawing the overlapping pentagons, the wire cube, and the face of a clock showing time, 10 past five.

Her examination did not reveal any neurocutaneous markers. We accordingly kept a diagnosis of Crossed non-aphasia in adult onset DDMS and managed her conservatively with muscle relaxants like baclofen and physiotherapy as well as anti-epileptics and counseling.

DISCUSSION

Dyke, Davidoff, and Masson first reported the condition way back in 1933. They described plain skull radiographic changes in 9 patients who presented with seizures, facial asymmetry, hemiparesis, and mental retardation [1]. Brain imaging reveals subtotal or diffuse hemiatrophy, prominent cortical sulci, dilated lateral ventricles and cisternal space, calvarial thickening, ipsilateral osseous hypertrophy with hyperpneumatization of the sinuses (mainly frontal and mastoid air cells), and an elevated temporal bone [2]. The clinical features include contralateral hemiparesis with an upper motor neuron type of facial palsy, focal or generalized seizures, and mental retardation along with learning disabilities [3].

Out of the two identified types of cerebral hemiatrophy, the acquired type results from various causes like birth asphyxia, prolonged febrile seizures, trauma, tumor, infection, ischemia, and hemorrhage eventually leading to reduced brain derived neurotrophic factors (BDNF) production [4, 5].

The radiological findings in our patient were somewhat atypical as there were no associated calvarial thickening or hyperpneumatized sinuses and near complete left hemispheric atrophy. The classical MRI changes of this disease is found only if the brain insult has occurred before 3 years of age because the maximum growth of a child's head occurs in the early years due to outward pressure of the enlarging human brain on the bony skull table which reaches three fourths of the adult size by the end of 3 years [6, 7, 8]. Therefore, only when brain damage is sustained before 3 years of age, other structures overlying the brain grow inward, thus resulting in an increased width of the diploic spaces, enlarged sinuses, and an elevated orbital roof, characteristic of this disorder [6]. Again, this explains the atypical radiologic findings, as the primary insult was sustained after five years of age, in this case.

Chronic hemiatrophy of the brain in our case acted like 'functional hemispherectomy' explaining the gradually progressive loss of dexterity and paucity of seizure episodes after years of intractable seizures [9, 10].

Most striking feature in this case is the clinical profile of our right-handed subject showed apparently normal linguistic skills despite a crucial extensive lesion in the linguistic areas of the left hemisphere, leading to a label of "crossed nonaphasia." Fischer and colleagues reported that if the language is lateralized to the right hemisphere in a right-handed subject, the visuospatial functions could be lateralized to the left hemisphere. The visuospatial deficits could be taken as evidence of this skill being located in the left hemisphere. Therefore, the processing of left hemispheric function (language) by the right hemisphere and the right hemisphere function (visuospatial skills) by the left hemisphere reveals a "mirrored cross-cerebral dominance," a rare clinical manifestation. The clinical profile exhibited by this patient was not an exception to the claims of Alexander and Annett, who reported that crossed nonaphasia was always a "mirror image" type. In addition, the absence of apraxia has also been supported by the earlier findings of these authors as they claimed that subjects with crossed nonaphasia have only a 50% chance of coexisting verbal apraxia [11, 12].

The differential diagnosis of this syndrome includes Sturge-Weber syndrome, Rasmussen encephalitis, Silver-Russell syndrome, basal ganglia germinoma, Fishman syndrome, and linear nevus syndrome. Closest differential being the Rasmussen encephalitis was excluded from detailed history, clinical examinations and negative neuroviruses panel tests by PCR. Most of the differentials, however, can be differentiated by performing a thorough clinical examination and by neuroimaging [13, 14].

To conclude, DDMS itself is a rare disease and adult presentation is even rarer. On top of that, this paper presented a right-handed teenage girl who exhibited apparently normal linguistic functions following colossal damage to the left hemisphere linguistic areas of the brain. The absence of aphasia in a right-handed subject following insult over left hemisphere language-dominant areas should never be ignored. Rather, such subjects with anomalous cerebral linguistic representation (such as crossed non-aphasia—as reported in our case and crossed aphasia) should serve as eye-openers to our long-held fallacies on brain-language-handedness relationships.

REFERENCES

1. Dyke CG, Davidoff LM, Masson CB. Cerebral hemiatrophy and homolateral hypertrophy of the skull and sinuses. Surg Gynecol Obstet. 1933;57:588–600.
2. Unal O, Tombul T, Cirak B, Anlar O, Incesu L, Kayan M. Left hemisphere and male sex dominance of cerebral hemiatrophy (DDMS) Clin Imaging. 2004;28:163–165.
3. Afifi AK, Godersky JC, Menezes A, et al. Cerebral hemiatrophy, hypoplasia of internal carotid artery and intracranial aneurysm. Arch Neurol. 1987;44:232–235
4. Sener RN, Jinkins JR. MR of craniocerebral hemiatrophy. Clin Imaging. 1992;16:93–97
5. Stred SE, Byrum CJ, Bove EL, Oliphant M. Coarctation of midaortic arch presenting with monoparesis. Ann Thorac Surg. 1986;42:210–212
6. Solomon GE, Hilal SK, Gold AP, Carter S. Natural history of acute hemiplegia of childhood. Brain. 1970;93:107–120
7. Graham A, Molnar Z. Development of the nervous system. In: Standring S, editor. Gray's Anatomy. ed 40. London: Churchill Livingstone, Elsevier; 2008. p. 385.
8. Lee JH, Lee ZI, Kim HK, Kwon SH. A case of Dyke-Davidoff-Masson syndrome in Korea. Korean J Pediatr. 2006;49:208–211.
9. Van Empelen R, Jennekens-Schinkel A, Buskens E, Helders PJ, van Nieuwenhuizen O, Dutch Collaborative Epilepsy Surgery Programme Functional consequences of hemispherectomy. Brain. 2004;127:2071–2079.
10. Devlin AM, Cross JH, Harkness W, Chong WK, Harding B, Vargha-Khadem F, et al. Clinical outcomes of hemispherectomy for epilepsy in childhood and adolescence. Brain. 2003;126:556–566
11. Alexander, M. P., & Annett, M. (1996). Crossed aphasia and related anomalies of cerebral organization: case reports and a genetic hypothesis. Brain and Language, 55, 213–239.
12. Fischer, R. S., Alexander, M. P., Gabriel, C., Gould, E., & Milione, J. (1991). Reversed lateralization of cognitive functions in right handers. Brain, 114, 245–261.
13. Rao KC. Degenerative diseases and hydrocephalus. In: Lee SH, Rao KC, Zimmerman RA, editors. Cranial MRI and CT. New York: McGraw-Hill; 1999. pp. 212–214.
14. Zilkha A. CT of cerebral hemiatrophy. Am J Roentgenol. 1980;135:259–262
15. Narain NP, Kumar R, Narain B. Dyke-Davidoff-Masson syndrome. Indian Pediatr. 2008;45:927–928