

Late diagnosis of acquired Dyke–Davidoff–Masson syndrome after childhood meningoencephalitis in a 34-year-old man: a case report

Abstract

Background: Dyke–Davidoff–Masson syndrome (DDMS) is a rare condition characterized by cerebral hemiatrophy with compensatory ipsilateral calvarial thickening and hyperpneumatization of the paranasal sinuses and mastoid air cells, clinically associated with seizures, contralateral hemiparesis and intellectual disability.

Case presentation: A 34-year-old man presented with focal clonic seizures of the left upper limb, occurring about once a week and sometimes evolving to bilateral tonic-clonic seizures, despite carbamazepine and phenobarbital. Born at term with an uneventful perinatal period and normal psychomotor development until the age of 3 years, he was then admitted to the intensive care unit for status epilepticus revealing meningoencephalitis, and was discharged with residual left hemiparesis and epilepsy. Examination showed left spastic hemiparesis, facial asymmetry, and cognitive impairment (MMSE 18/30). MRI revealed right cerebral hemiatrophy with ex vacuo ventricular dilatation, ipsilateral calvarial thickening and sinus hyperpneumatization, and contralateral cerebellar atrophy. EEG showed right-sided slowing with right temporal interictal discharges activated by sleep. The diagnosis of DDMS had never been made and was established only at the age of 34 years, 31 years after symptom onset. After phenobarbital was tapered off and replaced by lacosamide, seizure frequency decreased from about one per week to one per month.

Conclusion: This case illustrates the acquired form of DDMS after childhood meningoencephalitis and emphasizes the long diagnostic delay that may occur in this syndrome. Neuroimaging should be performed in any patient with childhood-onset hemiparesis and epilepsy, as early diagnosis allows timely recognition of drug resistance and optimization of management.

Keywords: Dyke–Davidoff–Masson syndrome, cerebral hemiatrophy, crossed cerebellar atrophy, delayed diagnosis, meningoencephalitis, drug-resistant epilepsy

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Najoua Mouloudi, Amine Reggabi, Houda Alloussi, Amal Satte

Department of Neurophysiology, Mohammed V Military Teaching Hospital, Morocco

Correspondence: Najoua Mouloudi; Department of Neurophysiology, Mohammed V Military Teaching Hospital, Rabat, Morocco; Tel 212648065133

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Introduction

Dyke–Davidoff–Masson syndrome was first described in 1933 by Dyke, Davidoff and Masson, who reported nine patients with hemiparesis, seizures, facial asymmetry and intellectual disability, in whom plain skull radiographs showed cerebral hemiatrophy with homolateral hypertrophy of the skull and sinuses.¹ The syndrome is rare and its prevalence is unknown.

DDMS may be congenital or acquired. In the congenital form, the insult occurs in utero and is mostly vascular. The acquired form results from an injury in the perinatal period or early childhood, such as birth asphyxia, trauma, infection, ischemia, hemorrhage or prolonged febrile seizures.² Compensatory skull changes develop only when the injury occurs before the brain has reached most of its adult size, usually in the first years of life.^{3,4}

Clinical manifestations include contralateral hemiparesis, epileptic seizures, facial asymmetry, intellectual disability and speech disorders.^{5,6} Although DDMS is usually diagnosed in childhood, it may be recognized in adolescence or adulthood.^{2,4,7,8} We report a case of acquired DDMS following meningoencephalitis at the age of 3 years, in which symptoms began at that age and the diagnosis was established only at the age of 34 years, and we discuss the reasons for and consequences of this diagnostic delay.

Case Presentation

A 34-year-old man, the only child of his parents, was referred to our department for recurrent seizures. He was born at term after an uneventful pregnancy and perinatal period, and his psychomotor development was normal until the age of 3 years. At that age, he was admitted to the intensive care unit for status epilepticus, which revealed meningoencephalitis. He remained in the intensive care unit for 20 days, received antibiotic therapy, and was discharged on carbamazepine and phenobarbital, with a residual left hemiparesis. The available historical records do not specify the infectious agent, cerebrospinal fluid findings, electroencephalographic findings, or neuroimaging results obtained during the acute episode. Since then, he has continued to experience focal clonic seizures of the left upper limb, occurring approximately once a week and occasionally progressing to bilateral tonic-clonic seizures, despite ongoing treatment with carbamazepine and phenobarbital. For more than three decades, his motor deficit and epilepsy were managed symptomatically as sequelae of this childhood illness, and no specific syndromic diagnosis had been made.

Neurological examination revealed left spastic hemiparesis with pyramidal signs, and mild left central facial palsy. Cognitive screening showed impairment, with a Mini-Mental State Examination (MMSE) score of 18/30. The rest of the examination was unremarkable.

Brain magnetic resonance imaging (MRI) (Figures 1 & 2) revealed right cerebral hemiatrophy with enlargement of the ipsilateral subarachnoid spaces and ex vacuo dilatation of the right lateral ventricle, predominantly involving the occipital horn. Thinning of the right cerebral peduncle and thickening of the diploë of the right cranial vault were also observed. In addition, hyperpneumatization of the right frontal and sphenoid sinuses and the right mastoid air cells was noted. The left cerebellar hemisphere showed atrophy with widened cerebellar sulci, consistent with crossed cerebellar atrophy. Electroencephalography (EEG) showed slowing of background activity over the right hemisphere, with right temporal interictal epileptiform discharges increasing during sleep.

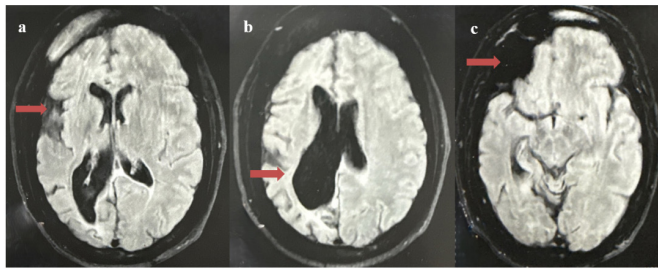


Figure 1 Axial FLAIR brain MRI showing right cerebral hemiatrophy (a), ex vacuo dilatation of the right lateral ventricle (b), and hyperpneumatization of the right frontal sinus (c).

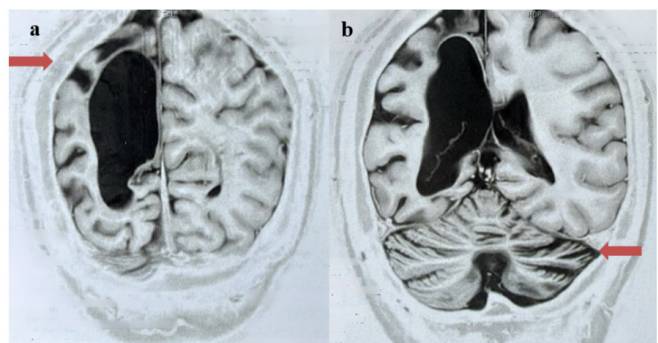


Figure 2 Coronal T1-weighted inversion-recovery brain MRI images showing (a) right-sided calvarial thickening, ipsilateral to the cerebral hemiatrophy, and (b) left cerebellar hemiatrophy with widening of the cerebellar sulci.

The diagnosis of acquired DDMS following childhood meningoencephalitis was thus established at the age of 34 years, 31 years after symptom onset. As seizures persisted despite two antiseizure medications administered at the maximum tolerated doses, the epilepsy was considered drug-resistant. Because of its potential adverse effects on cognition, phenobarbital was gradually tapered off and replaced by lacosamide, in combination with carbamazepine. The patient was also referred for physiotherapy, spasticity management and detailed neuropsychological assessment. At follow-up, he showed clinical improvement, with seizure frequency reduced from one per week to one per month.

Discussion

DDMS is defined radiologically by unilateral cerebral atrophy with compensatory ipsilateral changes of the skull: calvarial thickening, elevation of the petrous ridge and orbital roof, and hyperpneumatization of the frontal sinus and mastoid air cells.^{1,8} As the atrophic hemisphere no longer exerts outward pressure on the growing skull, the inner table grows inward, widening the diploë and enlarging the sinuses.^{1,2} In a review of 21 adult cases, Diestro et al. found cerebral hemiatrophy in all patients, and hemicalvarial

thickening and sinus hyperpneumatization each in 71.4%.⁷ The presence of these bone changes in our patient indicates an early-life insult, consistent with the history at the age of 3 years.

Our case corresponds to the acquired form of DDMS: the term birth, uneventful pregnancy and perinatal period, and normal psychomotor development until the age of 3 years argue against a congenital or perinatal origin. Central nervous system infections are among the recognized causes of acquired cerebral hemiatrophy;^{2,6} in the review by Diestro et al., three of the four adult cases with an identified etiology were attributed to encephalitis or meningoencephalitis,⁷ and childhood meningitis was the cause in two of seven adult patients reported by Ayas et al.⁹ Roy et al. described a similar patient who had meningoencephalitis with seizures at 28 months and later presented with right cerebral hemiatrophy, calvarial thickening, frontal sinus enlargement and crossed cerebellar atrophy.² In our patient, the episode at the age of 3 years is the only identified event in an otherwise normal early history, and the hemiparesis appeared at that time, which makes it the most likely origin of the hemispheric injury, although, in the absence of imaging from that period, this causal link cannot be formally proven.

The clinical picture of our patient is typical. In a systematic review of DDMS, contralateral hemiparesis or hemiplegia was present in 70% of patients and intellectual disability in 46%, and focal motor and focal to bilateral tonic-clonic seizures were among the reported seizure types.⁵ The same review found a slight male predominance (55.3%) and more frequent involvement of the left hemisphere; our patient thus has the less common right-sided form.

A striking feature of our case is the diagnostic delay of 31 years. Although DDMS is classically a pediatric diagnosis, many cases are recognized only in adolescence or adulthood: in the systematic review by Rondão et al., the mean age of reported patients was 19.4 years (range 0–83),⁵ and several adult series and case reports describe patients diagnosed only in adulthood.^{2,7,9} Recent reports describe diagnoses at 22, 38 and 39 years of age,^{4,8,10} sometimes in patients who had never undergone neuroimaging,^{4,8} and even an incidental imaging diagnosis in a 54-year-old woman.¹¹ Financial constraints and other unfavorable social determinants of health have been reported as contributors to such delays.¹⁰ In Rabat, Morocco, Junior Kessi et al. recently reported a 39-year-old man with left hemiparesis since childhood, born at term after a normal delivery without perinatal complications, in whom DDMS was diagnosed late in the absence of any prior radiological investigation⁸ — a situation very similar to ours. Several factors probably contributed to this delay in our patient: the deficit and seizures were attributed to the sequelae of the childhood meningoencephalitis; no records of childhood investigations were available, and any imaging that may have been performed at that time was not interpreted from a syndromic perspective; and the relative stability of the clinical picture did not prompt reassessment. This delay had practical consequences: the patient remained on the same two older antiseizure medications, including long-term phenobarbital, for about 30 years while continuing to have frequent seizures, so that drug resistance was not formally recognized and referral to an epilepsy center was postponed. Once the diagnosis was made, a change in antiseizure treatment was followed by a marked reduction in seizure frequency. This underlines the importance of brain MRI and periodic reassessment in any patient with childhood-onset hemiparesis and epilepsy, even when an apparent cause is known.

The long-standing hemispheric lesion also had remote effects along the connected pathways. Cerebral peduncle atrophy, noted in seven of 19 children by Atalar et al., is consistent with Wallerian

degeneration of the corticospinal tract.¹² Atrophy of the contralateral cerebellar hemisphere represents crossed cerebellar atrophy, attributed to interruption of the cortico-ponto-cerebellar pathway by a long-standing supratentorial lesion. In the series of Tien and Ashdown, patients with crossed cerebellar atrophy usually had significant contralateral supratentorial hemispheric atrophy and were younger at the time of injury than those without atrophy,¹³ which supports the early and chronic nature of the process in our patient.

The main differential diagnoses of cerebral hemiatrophy are Sturge–Weber syndrome, characterized by a facial port-wine birthmark, leptomeningeal angiomatosis and tram-track cortical calcifications;^{4,14} Rasmussen encephalitis, an immune-mediated disease with progressive unilateral hemispheric atrophy, progressive neurological deficit and intractable focal seizures;¹⁵ hemimegalencephaly, in which the affected hemisphere is enlarged; and sequelae of cerebrovascular lesions, inflammatory processes or head trauma, which may themselves represent the cause of acquired DDMS.⁶ The absence of a cutaneous angioma or a progressive neurological course, together with the presence of compensatory skull changes, favored DDMS in our patient.

The EEG findings — right-sided background slowing with right temporal interictal discharges activated by sleep — are concordant with the side of the structural lesion and with the seizure semiology, supporting a structural focal epilepsy of the right hemisphere. Persistent weekly seizures despite carbamazepine and phenobarbital raised concern for drug-resistant epilepsy.¹⁶ Adjunctive lacosamide increases the likelihood of a 50% or greater reduction in seizure frequency in drug-resistant focal epilepsy, as shown in a Cochrane review;¹⁷ in our patient, replacement of phenobarbital with lacosamide was followed by a reduction in reported seizure frequency from approximately one seizure per week to one per month. As seizures persist, follow-up in a specialized epilepsy center remains warranted. Epilepsy surgery may be considered in selected patients with DDMS who have disabling drug-resistant seizures, following comprehensive presurgical evaluation.^{4,6,18} In our patient, the suitability of a surgical approach would need to be assessed through such an evaluation.

Long-term use of phenobarbital and carbamazepine also deserves attention: both are enzyme-inducing antiseizure medications, with possible chronic impairment of attention, executive function, processing speed and learning, a risk increased by polytherapy and pre-existing neurological disease.¹⁹ Together with the hemispheric lesion and the high seizure burden, these effects may contribute to the cognitive impairment observed. For this reason, phenobarbital was progressively withdrawn and replaced by lacosamide, a newer antiseizure medication with a more favorable cognitive and interaction profile. Management is usually symptomatic and multidisciplinary, combining antiseizure medications, physiotherapy, muscle relaxants for spasticity, speech and occupational therapy, and psychiatric support when needed.^{2,4} A more favorable prognosis is associated with the onset of hemiparesis after 2 years of age and the absence of prolonged or recurrent seizure.^{4,8}

Conclusion

Dyke–Davidoff–Masson syndrome should be considered in any patient with contralateral spastic hemiparesis, seizures and intellectual impairment, particularly when there is a history of early-childhood brain injury such as meningoencephalitis. As our case shows, the diagnosis may be delayed for decades when symptoms are simply attributed to a known childhood insult. MRI is the key investigation, showing cerebral hemiatrophy with ipsilateral calvarial

thickening and sinus hyperpneumatization, which indicate an early-life insult. Associated ipsilateral cerebral peduncular thinning and crossed cerebellar atrophy further characterize the extent of the chronic structural changes. Epilepsy in DDMS is often drug-resistant, and early referral to an epilepsy center, optimization of antiseizure treatment, and multidisciplinary rehabilitation are essential to improve quality of life.

Ethical statement

Written informed consent was obtained from the patient for the publication of this case report and the accompanying images. Patient identity has been anonymized.

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Conflicts of interest

The authors declare that there are no conflicts of interest.

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