

Dyke-Davidoff-Masson Syndrome as an Incidental Finding During Evaluation for Decompensated Heart Failure: A Case Report

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Abstract

Background: Dyke-Davidoff-Masson syndrome (DDMS) is a rare cause of cerebral hemiatrophy, most often diagnosed in childhood or adolescence. Fewer than two dozen cases have been described in which the diagnosis was first established in adulthood, and in nearly all of these the diagnostic route has been a seizure, a neurology consultation, or a neurological complaint such as gait disturbance.

Case presentation: We describe a 52-year-old woman with a four-decade history of right-sided hemiparesis, attributed only to an unverified childhood cerebrovascular event and an unconfirmed secondhand history of poliomyelitis, together with untreated childhood-onset epilepsy. She was admitted with one week of progressive pedal edema and acute dyspnea, on a background of hypertension, coronary artery disease with a prior anterior-wall myocardial infarction, and known congestive cardiac failure. Examination showed findings of decompensated heart failure alongside a chronic right spastic hemiparesis, global aphasia, and measurable right-sided limb hemiatrophy. Neuroimaging obtained during this admission — prompted by the neurological examination rather than by the presenting cardiac complaint — showed left cerebral hemiatrophy with ipsilateral calvarial thickening and frontal sinus expansion, a pattern consistent with DDMS.

Conclusion: DDMS can remain unrecognized for decades and may first come to clinical attention as an incidental finding during evaluation of an entirely unrelated illness. This case adds to the small adult literature by illustrating discovery through a cardiac, rather than neurological, route, and underscores the value of neuroimaging in adults with longstanding, unexplained hemiparesis whenever the opportunity arises.

Keywords: Dyke-Davidoff-Masson syndrome; cerebral hemiatrophy; incidental diagnosis; heart failure; hemiparesis; adult presentation

Introduction

Dyke-Davidoff-Masson syndrome (DDMS) is a rare cause of cerebral hemiatrophy first described in 1933 by Dyke, Davidoff, and Masson in a series of nine patients who showed hemiparesis, facial asymmetry, seizures, and characteristic skull radiographic changes.[1] The syndrome's radiological triad — atrophy of one cerebral hemisphere, ipsilateral compensatory thickening of the calvarium, and hyperpneumatization of the frontal and ethmoid sinuses — has since become its diagnostic hallmark.[2] Two etiological subtypes are recognized: a congenital form resulting from an intrauterine vascular insult, and an acquired form following perinatal or early-childhood trauma, infection, or ischemia.[2]

DDMS is overwhelmingly a diagnosis of childhood. A systematic review of the published literature identified 223 studies and reported a mean patient age of 19.44 years, with a slight male predominance (55.3%).^[3] Adult presentations are recognized but distinctly uncommon: several recent case reports place the number of previously published adult cases at around 21, out of roughly 100 cases described since the original report.^[4,5] Within this small adult literature, the route to diagnosis has been fairly consistent — a new-onset seizure,^[4] longstanding epilepsy or hemiparesis brought for neurological evaluation,^[6,7] or, in at least one report, cerebral hemiatrophy found incidentally on brain imaging obtained for a neurological complaint such as gait difficulty.^[5] A small number of reports have also emphasized just how long the diagnosis can be delayed, including one adult case explicitly built around a fifty-year gap between symptom onset and diagnosis.^[8]

We report a case that fits within this adult, delayed-diagnosis literature but differs from every comparable report we identified in one respect: the diagnosis was not prompted by any neurological event at all. It surfaced as a secondary finding during hospitalization for decompensated heart failure, in a woman whose right-sided hemiparesis and hemiatrophy had gone unexplained for roughly four decades. We present this case, with its considerable documentation gaps made explicit, to add a further, less-described route by which this rare syndrome can come to light in adulthood.

Case Presentation

The patient was a 52-year-old woman, a homemaker, who presented with a one-week history of progressive pedal edema and one day of severe dyspnea. The edema began insidiously at the ankles and had extended to the level of the knees over the preceding two days. The dyspnea was of sudden onset, severe (grade 4), worse on lying flat, and unrelieved by rest or her usual medications. She denied cough, chest pain, palpitations, or syncope, but described intermittent episodes of paroxysmal nocturnal dyspnea over the preceding two months.

Her past history included similar, milder episodes of the same complaints on previous occasions; hypertension for eight years, on unspecified regular treatment; and a three-year history of coronary artery disease with an anterior-wall myocardial infarction and congestive cardiac failure, also on unspecified regular treatment. She had right-sided upper- and lower-limb weakness dating back to approximately age 10, recorded only as a possible cerebrovascular event of unknown cause, and childhood-onset epilepsy that had never been treated with antiepileptic medication. The patient's attender additionally gave a history of poliomyelitis in childhood; this was secondhand and was not corroborated by any medical record. There was no family history of similar complaints. She followed a mixed diet with a normal appetite, regular bowel and bladder habits, and no history of substance use. Her birth and immunization history could not be obtained.

Not available: exact names and doses of the “regular medications” used for hypertension and for coronary artery disease/heart failure; any documentation of the presumed childhood cerebrovascular event or of the alleged poliomyelitis; seizure semiology, frequency, and the most recent seizure prior to this admission. These details could not be obtained and are disclosed here as a limitation rather than supplied.

Clinical Findings

On general examination, the patient was conscious and coherent (Glasgow Coma Scale E4V4M5 = 13; the reduced verbal component should be interpreted cautiously given the global aphasia described below, since standard GCS scoring assumes intact language function). She was moderately built and nourished, without pallor, icterus, cyanosis, clubbing, or lymphadenopathy. Bilateral pitting pedal edema extended to the knees. Vital signs were: temperature 98.2°F, pulse 92 beats per minute and regular, blood pressure 140/90 mmHg in the right upper limb (sitting), and respiratory rate 25 per minute. Jugular venous pressure was elevated 6 cm above the sternal angle. No neurocutaneous markers were noted.

Limb examination revealed anisomelia, with the right upper and lower limbs shorter than the left in every segment measured, and shortening affecting the proximal, middle, and distal segments alike (rhizomelic, mesomelic, and acromelic involvement). Circumferential measurements confirmed a uniform reduction in bulk on the right:

Segment	Right	Left
Upper arm	23 cm	27 cm
Forearm	15 cm	19 cm
Thigh	42 cm	46 cm
Leg	22 cm	25 cm

Cardiovascular examination revealed a third heart sound (S3) without murmurs; respiratory examination revealed bilateral crepitations in the scapular and infrascapular regions; the abdomen was soft with no organomegaly — a constellation consistent with decompensated heart failure.

Neurological examination showed global aphasia, precluding formal assessment of intelligence, orientation, or memory; cranial nerves were intact. Motor examination showed clasp-knife spasticity and weakness (grade 3–4/5 across shoulder, elbow, wrist, hip, knee, and ankle movements) in the right upper and lower limbs, with a right hand grip of 60% of the left. Deep tendon reflexes were exaggerated on the right (3+ versus 2+ on the left) and the right plantar response was extensor. Coordination, sensation, cerebellar function, and the autonomic examination were normal, and there were no meningeal signs.

Not available: formal cognitive assessment (precluded by aphasia); presence or absence of facial asymmetry, a classically described feature of DDMS that was not explicitly documented in either direction in the source material.

Diagnostic Assessment

Computed tomography of the brain showed left cerebral hemiatrophy with ipsilateral falcine shift, hyperostosis of the left calvarium, and expansion of the left frontal sinus. Magnetic resonance imaging confirmed left cerebral hemiatrophy with prominent sulcal spaces and ex-vacuo dilatation of the left lateral ventricle, together with ipsilateral calvarial thickening and left frontal sinus expansion. Magnetic resonance angiography additionally showed reduced caliber of the left internal carotid artery and narrowing of the left middle cerebral artery; the documentation available for this report did not specify a definitive interpretation of this vascular finding beyond noting it.

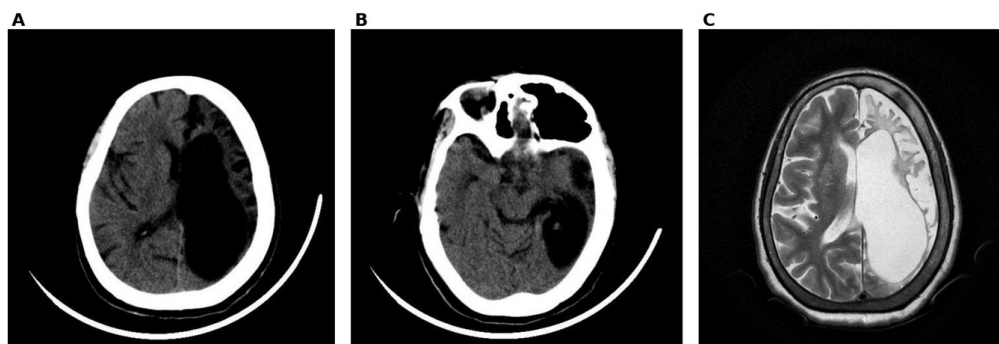


Figure 1. CT and MRI brain, axial views.

Figure 1. Axial neuroimaging. **(A)** Non-contrast CT at the level of the lateral ventricles, showing left cerebral hemiatrophy with ex-vacuo dilatation of the left lateral ventricle and ipsilateral calvarial thickening. **(B)** Axial CT closer to the skull base, showing marked expansion and hyperpneumatization of the left frontal sinus. **(C)** Axial T2-weighted MRI confirming left cerebral hemiatrophy, prominent sulcal widening, ex-vacuo ventricular dilatation, and leftward midline shift. Exact slice levels and sequence parameters were not specified in the source documentation; images are reproduced from the original clinical study and contain no identifying features.

This constellation — cerebral hemiatrophy with ipsilateral calvarial and sinus changes, occurring on the side opposite the patient's hemiparesis — was considered characteristic of DDMS. A broad differential was entertained, including Rasmussen encephalitis; postictal hemiatrophy following prolonged or recurrent febrile seizures; infectious causes (herpes encephalitis, TORCH infections, HIV); other vascular or hematological syndromes producing crossed cerebral findings (Sturge-Weber syndrome, crossed cerebral-cerebellar diaschisis, hemiconvulsion-hemiplegia-epilepsy syndrome); ischemic injury; neoplasia (basal ganglionic germinoma); prior radiation exposure; phakomatoses such as neurofibromatosis; and several rarer syndromes associated with hemiatrophy (linear nevus syndrome, Fishman syndrome, Silver-Russell syndrome, infantile hemiplegia syndrome). The absence of neurocutaneous markers argued against the phakomatoses; the chronic, static, non-progressive nature of the deficit and the classic radiological triad favored DDMS over an active inflammatory or neoplastic process. We note, however, that several of these alternatives — particularly infectious and inflammatory causes — cannot be fully excluded on imaging alone, and that laboratory, cerebrospinal fluid, and electroencephalographic data that would ordinarily help exclude them were not available for this report.

Not available: complete blood count and metabolic panel; inflammatory markers; infectious/autoimmune serologies; electroencephalogram; echocardiogram, electrocardiogram, and chest radiograph relevant to the cardiac presentation; any cerebrospinal fluid analysis. None of these were obtainable from the source documentation.

Therapeutic Intervention

The pharmacological and supportive management provided during this admission, for either the decompensated heart failure or the underlying neurological condition, could not be obtained and is not available in the documentation reviewed for this report. This absence is disclosed as a limitation rather than inferred or filled in.

Follow-up and Outcomes

Length of stay, in-hospital course, complications, discharge status, and any subsequent follow-up were not documented in the source material and could not be obtained. This report is accordingly limited to the diagnostic pathway described above and does not include outcome data.

Timeline

Approximate timing	Event
Birth	Birth and immunization history not obtainable
~Age 10	Onset of right-sided hemiparesis, retrospectively labeled “?CVA”; poliomyelitis also alleged by history, unconfirmed
Childhood (age unspecified)	Onset of epilepsy; never treated
~8 years before admission	Hypertension diagnosed
~3 years before admission	Coronary artery disease with anterior-wall myocardial infarction and congestive cardiac failure diagnosed
~2 months before admission	Intermittent paroxysmal nocturnal dyspnea
1 week before admission	Pedal edema onset
2 days before admission	Edema extends to knees
1 day before admission	Acute severe dyspnea prompts hospital presentation
During admission	General, systemic, and neurological examination performed; CT, MRI, and MRA brain obtained; DDMS diagnosis proposed
After discharge	Not documented

Discussion

DDMS is, in the great majority of published cases, a diagnosis made in childhood, when seizures, hemiparesis, or developmental concerns bring a child to medical attention.[3] The pooled data from the largest available systematic review — 223 studies, mean age 19.44 years — illustrate this skew clearly, as does the finding that contralateral hemiparesis (present in roughly 70% of pooled cases), facial asymmetry (31%), and intellectual disability (46%) are all documented far more often in a pediatric-weighted population than an adult one.[3] Adult first-diagnosis, by contrast, is genuinely uncommon: recent case reports converge on a figure of about 21 previously published adult cases.[4,5] A single-center retrospective series has also described seven adults diagnosed with DDMS over a six-year period, indicating that adult diagnosis, while rare, is not confined to isolated single reports.[10] Our patient's age of 52 sits comfortably within, if toward the older end of, this small adult group, at least two members of which have been diagnosed at a similar age, in their fifties.[4,5]

What is more informative than her age is how the diagnosis was reached. Every comparable adult case we identified was diagnosed through a neurological route: a new-onset seizure prompting emergency evaluation, [4] longstanding epilepsy or hemiparesis brought to a neurology clinic,[6,7,9] or, in the case closest to ours, cerebral hemiatrophy found incidentally on a CT scan obtained for weakness and gait difficulty — itself still a neurological complaint.[5] We were unable to find a published case in which the diagnostic imaging was prompted by an entirely different organ system's decompensation, as occurred here: this patient's neuroimaging followed directly from the neurological examination performed as part of a general medical workup for acute heart failure, not from any complaint referable to the nervous system itself. We would frame this cautiously — as a discovery pathway not represented in the cases we found, rather than as one that has never occurred — but it is, to our knowledge, a genuine point of difference from the existing adult DDMS literature. This comparison rests on structured but non-systematic searches of PubMed-indexed and Google Scholar-indexed literature rather than a formal, protocol-driven systematic review, so the observation should be read as provisional rather than exhaustive.

The four-decade interval between symptom onset and diagnosis is also worth situating honestly. Long diagnostic delays are not unique to this case; one prior report is built specifically around a fifty-year gap between the onset of symptoms and the eventual diagnosis of DDMS in adulthood,[8] and another explicitly describes a patient who presented very late despite symptoms dating to birth.[6] What may be somewhat more specific to our patient is the nature of the intervening explanations she was given — an unconfirmed “?CVA” and a secondhand, unverified history of poliomyelitis — neither of which appears to have prompted neuroimaging at the time. Distinguishing DDMS from the sequelae of poliomyelitis or of a discrete childhood stroke is a recognized diagnostic challenge, since both can produce longstanding, static limb weakness and asymmetry; the distinguishing radiological triad of DDMS is not something either of those alternative diagnoses would produce, but it can only be identified if imaging is actually obtained. That imaging occurred here largely as a byproduct of an unrelated hospitalization is, we think, the case's central teaching point: a longstanding, unexplained hemiparesis in an adult may warrant brain imaging even when the presenting complaint lies elsewhere, since the diagnostic window does not close with age.

Global aphasia was a prominent feature in this patient and is worth commenting on specifically, since it complicated both her general assessment (the Glasgow Coma Scale verbal component) and, more importantly, precluded any formal evaluation of cognition. Language and speech disorders are recognized, if less commonly reported, features of DDMS — present in roughly 15% of cases in the largest pooled review[3] — so aphasia of this severity sits within the described phenotype rather than outside it, though we cannot say from the available documentation whether this represents a longstanding deficit or an acute change coinciding with this admission. This uncertainty is not unique to our case: intellectual impairment, while part of the classical triad, is not invariable in adult-diagnosed DDMS, and at least one recent report explicitly described a phenotype without cognitive impairment or childhood seizures.[11]

This case has substantial limitations, most of which stem from the nature of the source documentation available to us rather than from the clinical course itself. No laboratory investigations, electroencephalogram, echocardiogram, electrocardiogram, or chest radiograph were available for review, all of which would ordinarily support both the cardiac and neurological aspects of the presentation and help exclude alternative diagnoses more rigorously. Treatment, hospital course, and outcome data were entirely unavailable, which limits this report's usefulness for informing management and is a significant departure from CARE reporting expectations. The history of poliomyelitis rests on an unverified secondhand account. Formal cognitive testing could not be performed because of aphasia, so we cannot confirm or refute intellectual disability traditionally described as part of the DDMS triad in this patient. As a single case, this report cannot establish incidence or generalize about presentation patterns; its contribution is limited to the specific, narrow observation about discovery pathway described above.

Conclusion

This case illustrates that Dyke-Davidoff-Masson syndrome can go undiagnosed for decades and may ultimately surface not through a seizure or a neurology referral, but as an incidental finding during evaluation of an unrelated acute illness — here, decompensated heart failure. For clinicians encountering an adult with longstanding, unexplained hemiparesis, this case is a reminder that a definitive structural diagnosis may still be within reach, whenever the opportunity to image the brain presents itself.

Patient Perspective

The patient's own account of her diagnosis and its lifelong impact was not captured in the source documentation.

Informed Consent

Written informed consent for publication of this case, including the clinical and imaging findings reported here, was obtained from the patient.

Figure 1 (the CT/MRI panels) contains no facial or other identifying features. The source material separately contains two clinical photographs that clearly show the patient's face; consistent with standard de-identification practice, these have not been used anywhere in this manuscript.

Declarations

Consent for publication: Obtained from the patient — see Informed Consent above.

Conflict of interest: The authors declare no conflicts of interest.

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Author contributions: Midhun Kumar Maganti, Hafsah Arif Kundi, Nithin Prabhakar, and Rakhi Bai contributed to the preparation and review of the manuscript and approved the final version.

Data availability: Not applicable; no additional dataset beyond what is reported here.

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