ACTA RADIOLÓGICA PORTUGUESA
September-December 2026 | Vol 38 nº 3 | e46246 https://doi.org/10.25748/arp.46246

Radiological Case Report / Caso Clínico

Dyke–Davidoff–Masson Syndrome in Adulthood

Síndrome de Dyke–Davidoff–Masson na Idade Adulta

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Received: 26/04/2026
Accepted: 13/08/2026
Published:02/09/2026

Abstract

Dyke–Davidoff–Masson syndrome is a rare neurological condition characterized by unilateral cerebral atrophy with compensatory thickening of the cranial vault, usually resulting from early brain injury. The diagnosis is typically established in childhood, with its identification in adulthood being uncommon. We describe the case of a 49-year-old male patient with a reported history of meningitis during childhood and a two-year history of recurrent headaches in the right temporoparietal region. Neurological examination revealed mild right hemiparesis and hemihypoesthesia, associated with ipsilateral conjunctival hyperemia and lacrimation. Based on these findings, the hypothesis of short-lasting unilateral neuralgiform headache attacks was initially considered. Neuroimaging revealed left cortico-subcortical encephalomalacia, ipsilateral cerebral hemiatrophy, and compensatory thickening of the cranial vault, findings compatible with Dyke–Davidoff–Masson syndrome. This case highlights the importance of neuroimaging in patients with atypical headaches and subtle focal neurological deficits.

Keywords

Brain Atrophy; Encephalomalacia; Hemiparesis; Magnetic Resonance Imaging; Headache.

Resumo

A síndrome de Dyke–Davidoff–Masson é uma condição neurológica rara caracterizada por atrofia cerebral unilateral com espessamento compensatório da calote craniana, geralmente resultante de lesão cerebral precoce. O diagnóstico é habitualmente estabelecido na infância, sendo a sua identificação na idade adulta pouco frequente. Descreve-se o caso de um doente do sexo masculino, de 49 anos, com antecedentes referidos de meningite na infância e história de cefaleias recorrentes na região temporoparietal direita com evolução de dois anos. O exame neurológico revelou hemiparesia e hemihipoestesia direitas ligeiras, associadas a hiperemia conjuntival e lacrimejo ipsilaterais. Com base nestes achados, foi inicialmente equacionada a hipótese de cefaleia neuralgiforme unilateral de curta duração. A neuroimagem revelou encefalomalácia córtico-subcortical esquerda, hemiatrofia cerebral ipsilateral e espessamento compensatório da calote craniana, achados compatíveis com a síndrome de Dyke–Davidoff–Masson. Este caso destaca a importância da neuroimagiologia em doentes com cefaleias atípicas e défices neurológicos focais subtis.

Palavras-chave

Atrofia cerebral; Encefalomalácia; Hemiparesia; Ressonância magnética; Cefaleia.

Introduction

Dyke–Davidoff–Masson syndrome (DDMS) is a rare neurological condition first described in 1933 by Dyke, Davidoff and Masson.[1] It is characterized by cerebral hemiatrophy associated with compensatory osseous changes of the skull and may clinically manifest with contralateral hemiplegia or hemiparesis, seizures, facial asymmetry, speech disturbances and cognitive impairment.[2,3]

The syndrome usually results from an early cerebral insult, either congenital or acquired, occurring during critical periods of brain development. Reported etiologies include ischemic events, trauma, central nervous system infections and intracranial hemorrhage, which lead to neuronal loss and interruption of normal cerebral growth. As a consequence, hemispheric atrophy occurs together with ipsilateral compensatory changes, particularly thickening of the calvarium and hyperpneumatization of the paranasal sinuses.[4]

In most cases, the diagnosis is established during childhood or adolescence, when more evident neurological manifestations such as epilepsy or persistent motor deficits become apparent. In contrast, identification of the syndrome in adulthood is uncommon and usually occurs as an incidental finding during neuroimaging performed for the investigation of other neurological complaints.[1,5]

We present the case of an adult patient in whom investigation of recurrent headaches revealed imaging findings consistent with Dyke–Davidoff–Masson syndrome.

Case Report

A 49-year-old male presented to the Emergency Department in June 2024 due to recent worsening of long-standing right temporoparietal headache, with increased intensity during the previous three months. He reported a history of meningitis during childhood, with no available clinical documentation or previous neuroimaging studies allowing precise characterization of the episode. The attacks were described as intense stabbing pain lasting 10–20 seconds and occurring several times per day, associated with ipsilateral

autonomic signs, namely conjunctival injection, lacrimation and rhinorrhea.

At admission he was conscious, oriented and hemodynamically stable. The initial neurological examination did not reveal clear deficits, although mild right hemiparesis accompanied by hemihypoesthesia on the same side was later identified.

Neurological evaluation was requested. The patient reported episodes of severe pain (9/10) lasting approximately 20 seconds with progressively increasing frequency, reaching seven to eight episodes per day, with associated ipsilateral autonomic features.

Between episodes he remained asymptomatic. He denied loss of consciousness, visual disturbances, vertigo or newly developed neurological deficits.

The clinical presentation suggested a short-lasting trigeminal autonomic headache, and short-lasting unilateral neuralgiform headache attacks with conjunctival injection and tearing (SUNCT) was considered. Treatment with lamotrigine was initiated and a cranial computed tomography scan was requested.

Non-contrast cranial computed tomography demonstrated a left parietotemporal cortico-subcortical encephaloclastic lesion, associated with left cerebral hemiatrophy/hypoplasia and mild ipsilateral ventricular dilatation (Figure 1A). No acute parenchymal abnormalities, extra-axial collections or signs of mass effect were identified. A mucous retention cyst/polyp was noted in the left maxillary sinus, with otherwise preserved aeration of the paranasal sinuses and middle ears. The constellation of findings was suggestive of Dyke–Davidoff–Masson syndrome. For further characterization, brain magnetic resonance imaging was subsequently performed. Axial FLAIR imaging confirmed a left parietotemporal cortico-subcortical encephaloclastic lesion with marginal gliosis, associated with ipsilateral cerebral volume loss, interpreted as chronic sequelae of a probable old infarction in a partial territory of the left middle cerebral artery (Figure 1B). Three-dimensional time-of-flight MR angiography showed asymmetric caliber of the M1 and M2 segments of the middle cerebral arteries, smaller on the left, with reduced vascular density of the corresponding cortical branches.

Axial T2-weighted imaging further demonstrated left hemispheric atrophy with associated sulcal widening and mild ipsilateral ventricular enlargement (Figure 1C). Mild ipsilateral calvarial thickening was also noted, supporting compensatory osseous remodeling in the context of Dyke–Davidoff–Masson syndrome. No additional signal or morphological abnormalities of the remaining brain parenchyma were reported, and there were no areas of restricted diffusion or increased magnetic susceptibility. The available imaging reports did not describe thalamic atrophy, Wallerian degeneration or trigeminal nerve abnormalities.

During follow-up, lamotrigine was associated with partial improvement in the frequency and intensity of headache attacks, without documented new focal neurological deficits or seizures.

Discussion

Dyke–Davidoff–Masson syndrome is a rare entity associated with early cerebral insults leading to cerebral hemiatrophy and compensatory osseous changes. Classical imaging findings include unilateral cerebral volume loss, ipsilateral ventricular dilatation and thickening of the homolateral calvarium, frequently accompanied by hyperpneumatization of the paranasal sinuses.[1,7]

Computed tomography and magnetic resonance imaging represent the main diagnostic tools, allowing characterization of both structural alterations and associated parenchymal sequelae. Magnetic resonance imaging is particularly useful for identifying areas of encephalomalacia, gliosis and associated vascular abnormalities.

In most reported cases, the diagnosis is established in childhood, when more evident neurological manifestations such as seizures, persistent motor deficits or developmental delay appear. Identification in adulthood is less frequent and generally occurs during the investigation of other neurological complaints.4 Some cases reported in the literature suggest that the syndrome may remain undiagnosed for decades and only be recognized later in life.[6]

In the present case, despite the presence of mild hemiparesis, the neurological deficit was subtle and apparently longstanding, and had not previously been recognized as a manifestation of structural brain pathology. Neuroimaging revealed findings typical of the syndrome, suggesting an old cerebral insult acquired early in life. The parietotemporal topography and vascular asymmetry on MR angiography may support a previous vascular event, while the reported history of childhood meningitis represents a plausible infectious insult. However, causality cannot be established retrospectively.

Recurrent headache has been reported in association with DDMS.6 However, the literature reviewed did not identify robust or consistent evidence supporting a specific association between DDMS and trigeminal autonomic cephalalgias. A characteristic trigeminal autonomic phenotype or causal relationship has therefore not been established.

The differential diagnosis includes other causes of unilateral cerebral hemiatrophy, such as sequelae of perinatal infarction, Rasmussen syndrome, Sturge–Weber syndrome, and post-traumatic or post-infectious sequelae. The presence of compensatory calvarial thickening and associated osseous alterations remains a characteristic imaging feature supporting Dyke–Davidoff–Masson syndrome.[7]

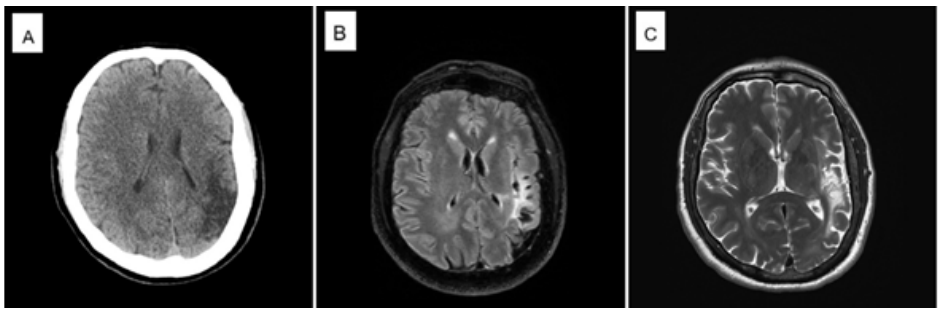

Figure 1 – (A) Axial non-contrast brain CT showing a left parietotemporal cortico-subcortical encephaloclastic lesion, associated with left hemispheric volume loss and mild ipsilateral ventricular dilatation. (B) Axial FLAIR MRI demonstrating marginal gliosis associated with the left parietotemporal encephaloclastic lesion. (C) Axial T2-weighted MRI confirming left hemispheric atrophy with associated sulcal widening, ventricular enlargement and mild ipsilateral calvarial thickening.

Conclusion

Dyke–Davidoff–Masson syndrome should be considered in the differential diagnosis of unilateral structural brain abnormalities identified in neuroimaging studies. Although it is typically diagnosed during childhood, it may remain unrecognized until adulthood and be incidentally identified during the investigation of other neurological conditions. Correct interpretation of the imaging findings, supported by careful clinical history, is essential to avoid misinterpretation as recent lesions or active pathological processes.

Ethical Disclosures / Divulgações Éticas

Conflicts of interest: The authors have no conflicts of interest to declare.

Conflitos de interesse: Os autores declaram não possuir conflitos de interesse.

Financing Support: This work has not received any contribution, grant or scholarship.

Suporte financeiro: O presente trabalho não foi suportado por nenhum subsídio ou bolsa.

Confidentiality of data: The authors declare that they have followed the protocols of their work center on the publication of data from patients.

Confidencialidade dos dados: Os autores declaram ter seguido os protocolos do seu centro de trabalho acerca da publicação dos dados de doentes.

Protection of human and animal subjects: The authors declare that the procedures followed were in accordance with the regulations of the relevant clinical research ethics committee and with those of the Code of Ethics of the World Medical Association (Declaration of Helsinki).

Protecção de pessoas e animais: Os autores declaram que os procedimentos seguidos estavam de acordo com os regulamentos estabelecidos pelos responsáveis da Comissão de Investigação Clínica e Ética e de acordo com a Declaração de Helsínquia da Associação Médica Mundial.

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