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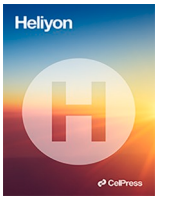

Case report

A case report on atypical presentations of Dyke-Davidoff-Masson syndrome

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ABSTRACT

Dyke-Davidoff-Masson Syndrome (DDMS) is a rare neurological disease with an unknown incidence. The manifestations of DDMS are variable, while typical symptoms are seizures, hemiparesis, and mental retardation. Here, we present a case involving a 19-year-old male patient who presents with headaches, mood changes, and a history of seizures during childhood. Based on the neuroimages, a diagnosis of DDMS was established. The application of sertraline hydrochloride as a therapeutic intervention has alleviated the symptoms. This case report illustrates the importance of understanding the clinical features of DDMS based on imaging.

1. Introduction

Typical neuroimaging findings of DDMS include partial or diffuse cerebral hemiatrophy accompanied by compensatory changes in the skull, such as thickening of the ipsilateral calvarium, nasal sinus hyperpneumatization, and elevation of the temporal bone [1]. The disease is mostly characterized by contralateral hemiparesis or hemiplegia and epilepsy. It is rare for DDMS to exhibit psychiatric symptoms as the primary manifestation. In this case, headaches and psychiatric symptoms were described as manifestations of DDMS. The demographic information of this patient is shown in Table 1.

2. Case presentation

2.1. History of present illness

A 19-year-old male presented to our hospital with a headache, which was described as blurred vision when he heard a noise, followed by intermittent tingling at the top of the head, lasting about 5 minutes and occurring 5–6 times a day. This first happened five months ago and has continued until now. In addition to the above symptoms, the patient has no other discomfort.

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Table 1
Demographic information.

Age	Nineteen
Sex	Male
Nation	The Han Nationality
Educational Level	Middle school
Occupation	Unemployed

2.2. Past medical history

He had a high fever for a week when he was five due to encephalitis. Then four years after that, the patient experienced a seizure. We couldn't find out more details about his epilepsy from his parents. They went to Shanghai for progressive therapy and he did not experience a seizure after treatment for three years. He is currently taking magnesium valproate 0.25g bid, and seizures have been well-controlled, with no occurrences over the past seven years. However, due to the long-term family environment and other social factors, the patient became irritable and even had suicidal intentions a year ago. Therefore, the patient's parents took him to the psychology department of the local hospital, where he was diagnosed with anxiety disorder and treated with buspirone 5mg bid and duloxetine 20mg bid. The patient's disease progression is shown in the timeline diagram (Fig. 1).

The patient was born at term but was delivered at home. There is no evidence that birth trauma occurred. The patient had normal physical growth and development, with a height of 172.4 cm and a weight of 97.3 kg. He has a biological sister, and the family members are healthy. There was no history of mental illness or related genetic disorders in the family.

2.3. Physical examination and auxiliary examination

This boy is right-handed and physical examination revealed no abnormality. The face and limbs on both sides are symmetrical. Also, there was no obvious abnormality in the walking gait. His mini-mental state examination (MMSE) score was 21 indicating mild mental retardation. He hasn't gone to school since he graduated from junior.

2.4. Diagnosis and treatment process

During the hospitalization, the patient did not have seizures, and the main symptoms were headache and depression. Combined with the patient's past medical history, we performed valproate concentration determination, cranial computerized tomography (CT), paranasal sinuses CT, cerebral magnetic resonance imaging (MRI), Electroencephalogram (EEG) and other relevant examinations. The concentration of valproate was slightly lower, and the test result was 48.79mg/L. The cranial CT revealed right cerebral hemisphere atrophy and encephalomalacia in the right front and temporal lobe (Fig. 2A and B). Additionally, the compensatory hypertrophy of the skull was shown in the temporal bone (Fig. 2A and B). The paranasal sinuses CT demonstrated no obvious abnormality. The MRI results (Fig. 3A–D) were consistent with the cranial CT results. The EEG recording showed an interhemispheric asymmetry with increased slow-wave activity in the right temporal area, with no epileptiform activity (Fig. 4). According to these, we then proposed a diagnosis of DDMS. Considering that the patient's headache may be mood-related, we requested a psychological consultation for further evaluation. After the communication between the psychiatrist and the patient, and the evaluation of the relevant scales, the diagnosis

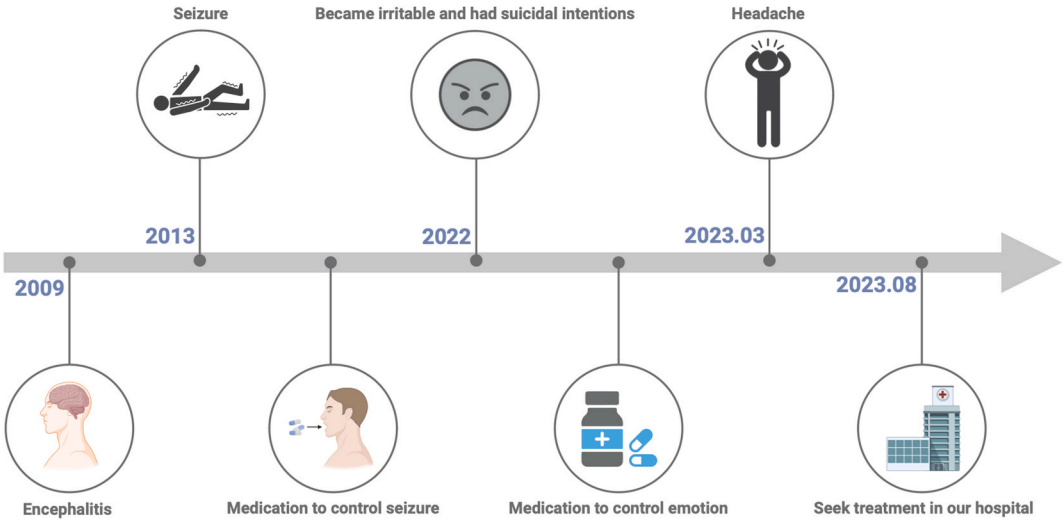

Fig. 1. A timeline of disease progression.

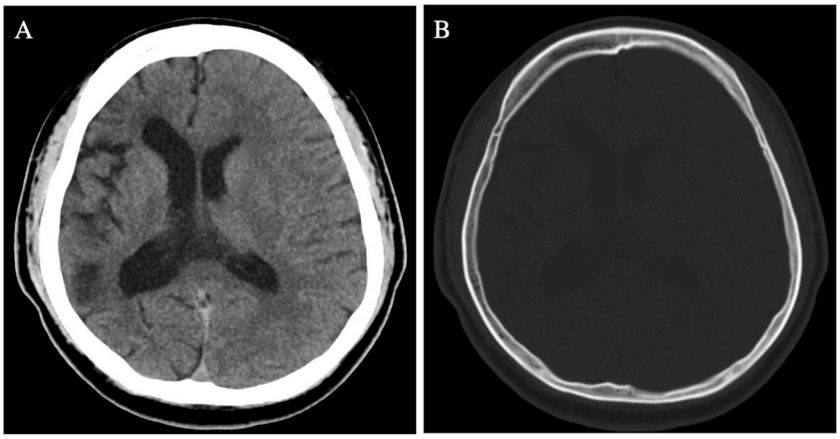

Fig. 2. The brain window (A), the bone window (B); A. The right cerebral hemisphere atrophy and encephalomalacia in the right front and temporal lobe; B. Compensatory hypertrophy of right temporal bone.

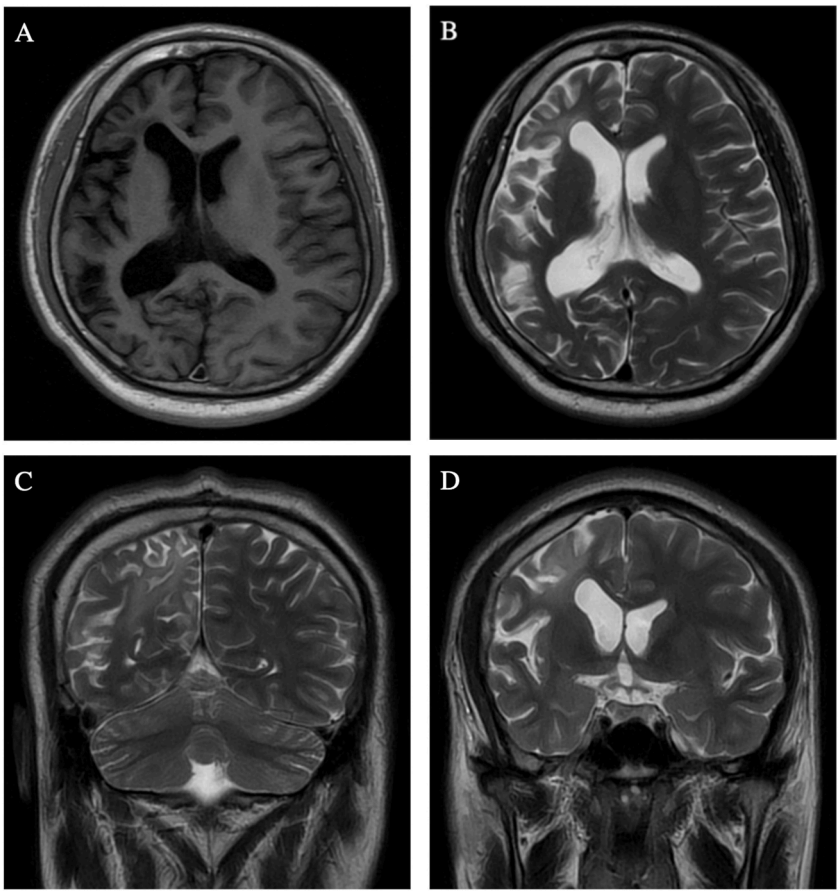

Fig. 3. T1 FLAIR (A), T2WI horizontal (B), T2WI coronal (C and D). Head MRI showed atrophy of the right cerebral hemisphere and encephalomalacia of the right frontal and temporal lobe.

of depressive state was given. It is also recommended to suspend the use of duloxetine and buspirone and to give a daily dose of 50mg sertraline hydrochloride to improve symptoms of depression and anxiety. The patient had no discomfort after taking sertraline, and the depression was better than before. Subsequently, the dosage was escalated to 100 mg after 4 days and remained at that level. No any adverse and unexpected events have occurred during the treatment. Then the patient was discharged after his headache symptoms improved.

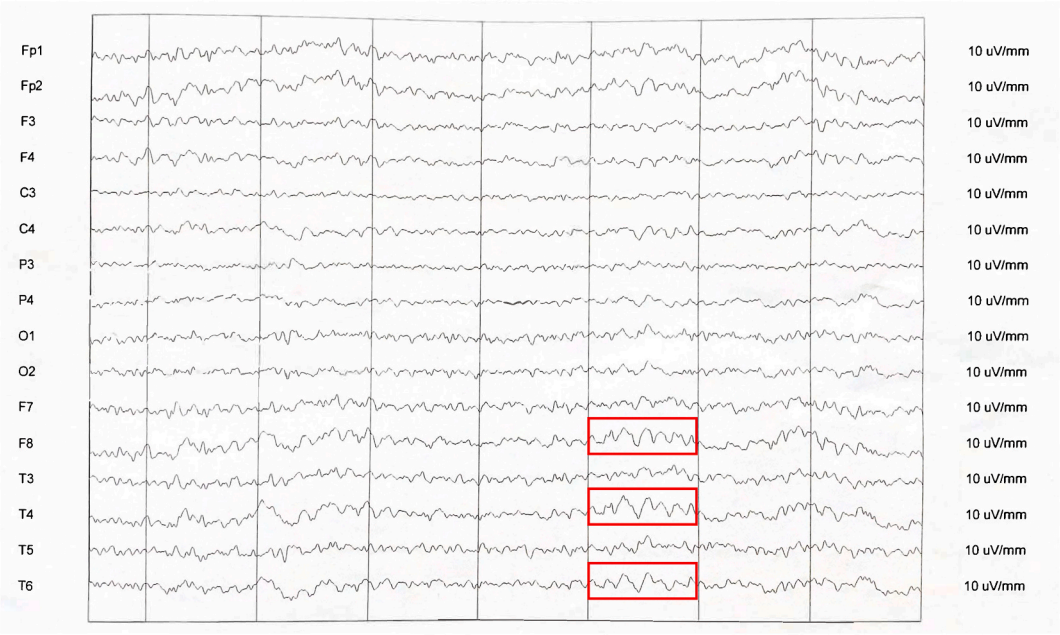

Fig. 4. The EEG showed an interhemispheric asymmetry with increased slow-wave activity in the right temporal area, with no epileptiform activity.

2.5. Follow-up visit

We knew that he has consistently taken his prescribed medications since his discharge. Headache symptoms are better than before, as the frequency is reduced to once a few months, the pain is also less than before. We will continue to follow this patient regularly to monitor his headaches, mood, seizures, and other changes in his condition.

3. Discussion

This case demonstrates an example of DDMS wherein psychiatric symptom and headache serve as the primary presentation. There are few reported cases of DDMS co-occurring with psychiatric symptoms in DDMS. As the incidence of DDMS is uncertain, our case report can provide a reference. After the first report of DDMS by Dyke in 1933 [2], many articles described this disease. DDMS is usually characterized by seizures, hemiparesis, mental retardation, and cerebral hemiatrophy in CT or MRI. There are few reports with psychiatric symptoms as the main manifestation, such as schizophrenia [3], anxiety, irritability [4], depression [5], and behavioral problems [6]. Most DDMS cases were identified after psychiatric symptoms developed.

The symptoms of DDMS are varied, so neuroimaging is a crucial part of the diagnosis process. Usually, atrophy of the left cerebral hemisphere is thought to be more common in DDMS patients [7,8]. However, the small sample size has a significant impact on this conclusion [9]. Insufficient cerebral development leads to the inward expansion of adjacent anatomical structures, thus elucidating the enlargement of the frontal sinus and the augmented breadth of the diploic space [10].

What distinguishes this case from other reported cases of this disease with concurrent psychiatric symptoms is that the chief complaint of the patient was headache. Headache is a non-specific clinical symptom. In addition, this case does not present hemiplegia or bilateral asymmetrical development of typical DDMS. Meanwhile, imaging shows atrophy of the right cerebral hemisphere. Although he suffered seizures at an early age, it was controlled well after pharmacotherapy. Unfortunately, the inability to ascertain the specific type of epileptic seizures was attributed to a dearth of clinical data. Also, another limitation of this case is that we did not take a cerebral CT or MRI angiography which may provide a basis for diagnosis of the cause. During the early stages of infancy, clinicians may employ cerebral MRI angiography as a diagnostic tool for identifying this uncommon medical condition [11]. The timely identification and diagnosis of the syndrome are imperative in facilitating the comprehensive development of the child's mental and physical well-being.

The etiology of DDMS can be classified as either congenital or acquired [2], nevertheless, the natural evolution of this disease is still unclear. It is common to see vascular abnormalities in congenital DDMS, which occurs during the perinatal period. The acquired type is developed during the perinatal period or later [12] and its etiology includes trauma, infection [13], and vascular disease [14,15], among others. Based on a comprehensive systematic review, it has been observed that seizures in the acquired type are typically focal, exhibiting improved control and prognosis. Conversely, in the congenital type, seizures tend to manifest as generalized tonic-clonic episodes in most instances [8]. Also, most dilated sulci and encephalomalacia cases were observed in acquired cases. However, the etiology of each case is not sufficiently clear in the literature reports, which compromises the precise discrimination between congenital and acquired cases. In this case, we can find dilated sulci and encephalomalacia in CT and MRI images. The prognosis of

seizures is controlled well. According to the above, we classify it as the acquired type. The central nervous system infection in early childhood is thought to be a possible etiological factor because dilated sulci and encephalomalacia may be the consequence of traumatic delivery or intracranial infection.

DDMS is more likely to have a favorable prognosis when symptoms appear after two years of age, according to Zawar et al. [16]. The main therapeutic approach for this disease is symptomatic treatment. It has been suggested that functional hemispherectomy might show efficacy in patients with refractory seizures [17].

4. Conclusion

In conclusion, it is imperative to conduct regular follow-up and brain CT or MRI examinations for individuals who develop epilepsy during childhood, regardless of the effectiveness of their current epilepsy management, to facilitate the diagnosis and treatment of uncommon medical conditions. Given that a subset of adolescent patients with DDMS may primarily exhibit mental symptoms upon seeking medical intervention, it is indeed very important for psychiatrists, neurologists, pediatricians, and radiologists to be familiar with this condition for its early diagnosis and treatment.

Ethical statement

The patient provided informed consent for the publication of his anonymised case details and images.

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Data availability

Data sharing is not applicable to this article as no datasets were generated or analyzed during the current study.

CRediT authorship contribution statement

Bingjie Yang: Writing – original draft, Conceptualization. **Shuqi Hu:** Writing – review & editing, Methodology, Conceptualization. **Yiru Jiang:** Writing – review & editing, Resources. **Song Shu:** Writing – review & editing, Resources. **Huixia Zhou:** Writing – review & editing. **Jiahui Zhu:** Writing – review & editing. **Hao Zhang:** Writing – review & editing, Supervision.

Declaration of competing interest

The authors declare that they have no known competing financial interests or personal relationships that could have appeared to influence the work reported in this paper.

Appendix A. Supplementary data

Supplementary data to this article can be found online at https://doi.org/10.1016/j.heliyon.2024.e35600.

References

[1] Z.Ö. Ayas, K. Asil, R. Öcal, The clinico-radiological spectrum of Dyke-Davidoff-Masson syndrome in adults, Neurol. Sci. 38 (2017) 1823–1828, https://doi.org/10.1007/s10072-017-3074-7.
[2] Cornelius Gysbert Dyke, Cerebral hemiatrophy with homolateral hypertrophy of the skull and sinuses, Surg Gyn Obstet (1933) 588–600.
[3] B. Amann, C. García de la Iglesia, P. McKenna, E. Pomarol-Clotet, M. Sanchez-Guerra, M. Orth, Treatment-refractory schizoaffective disorder in a patient with dyke-davidoff-masson syndrome, CNS Spectr. 14 (1) (2009) 36–39, https://doi.org/10.1017/s1092852900020034.
[4] B. Wang, W. Jiang, W. Yan, J. Tian, J. Xu, Y. Li, Y. Zhao, Y. Dai, G. Cheng, G. Hou, Clinical characteristics and neuroimaging findings of seven patients with Dyke Davidoff Masson syndrome, BMC Neurol. 21 (2021) 213, https://doi.org/10.1186/s12883-021-02242-4.
[5] J. Sordia-Ramírez, A. Infante-Valenzuela, I. de J. Hernández-Galarza, A. Costilla-Esquivel, Neuropsychiatric symptoms in a patient with Dyke-Davidoff-Masson syndrome and systemic lupus erythematosus: a case report, J. Med. Case Rep. 13 (2019) 111, https://doi.org/10.1186/s13256-019-2039-2.
[6] S.S. Bhandarı, S.J. Joseph, I.L. Sharma, G. Medhı, Dyke davidoff masson syndrome presenting with intellectual disability with behavioral problems and substance use disorder: a case report, Türk Psikiyatri Derg. 29 (2018) 291–294.
[7] O. Unal, T. Tombul, B. Cirak, O. Anlar, L. Incesu, M. Kayan, Left hemisphere and male sex dominance of cerebral hemiatrophy (Dyke-Davidoff-Masson Syndrome), Clin Imaging 28 (2004) 163–165, https://doi.org/10.1016/S0899-7071(03)00158-X.
[8] M.B.A. Rondão, B.R.R.H.S. Hsu, R.S. Centeno, P.H.P. de Aguiar, Dyke-Davidoff-Masson Syndrome: main clinical and radiological findings- systematic literature review, Seizure 110 (2023) 58–68, https://doi.org/10.1016/j.seizure.2023.04.020.

[9] J.D.B. Diestro, M.K.C. Dorotan, A.C. Camacho, K.T. Perez-Gosiengfiao, L.I. Cabral-Lim, Clinical spectrum of Dyke-Davidoff-Masson syndrome in the adult: an atypical presentation and review of literature, BMJ Case Rep. (2018), https://doi.org/10.1136/bcr-2018-224170 bcr2018224170, bcr-2018–224170, 2018.
[10] P. Malik, R. Garg, A.K.D. Gulia, J. Kario, Dyke-Davidoff-Masson Syndrome- a rare cause of refractory epilepsy, Iran. J. Psychiatry 9 (2014) 42–44.
[11] C. Caffarelli, F. Santamaria, A. Vottero, C.P. Dascola, V. Mirra, F. Sperli, S. Bernasconi, Progress in pediatrics in 2013: choices in allergology, endocrinology, gastroenterology, hypertension, infectious diseases, neonatology, neurology, nutrition and respiratory tract illnesses, Ital. J. Pediatr. 40 (2014) 62, https://doi.org/10.1186/1824-7288-40-62.
[12] P.H. Aguiar, C.W. Liu, H. Leitão, F. Issa, G. Lepski, E.G. Figueiredo, F. Gomes-Pinto, R. Marino Júnior, MR and CT imaging in the Dyke-Davidoff-Masson syndrome. Report of three cases and contribution to pathogenesis and differential diagnosis, Arq Neuropsiquiatr 56 (1998) 803–807, https://doi.org/10.1590/s0004-282x1998000500016.
[13] A. Zilkha, CT of cerebral hemiatrophy, AJR Am. J. Roentgenol. 135 (1980) 259–262, https://doi.org/10.2214/ajr.135.2.259.
[14] R.N. Sener, J.R. Jinkins, MR of craniocerebral hemiatrophy, Clin Imaging 16 (1992) 93–97, https://doi.org/10.1016/0899-7071(92)90119-t.
[15] K. Ono, K. Komai, T. Ikeda, Dyke-Davidoff-Masson syndrome manifested by seizure in late childhood: a case report, J. Clin. Neurosci. 10 (2003) 367–371, https://doi.org/10.1016/s0967-5868(03)00011-0.
[16] I. Zawar, A.A. Khan, T. Sultan, A.W. Rathore, Dyke-Davidoff-Masson Syndrome. An unusual cause of status epilepticus, Neurosciences 20 (2015) 385–387, https://doi.org/10.17712/nsj.2015.4.20150481.
[17] B. Shrestha, Acquired cerebral hemiatrophy: dyke-Davidoff-Masson Syndrome - a case report, Turk Neurosurg 23 (2013) 117–121, https://doi.org/10.5137/1019-5149.JTN.4283-11.1.