



Classical oral manifestations of Dyke-Davidoff-Masson syndrome: a case report with review of the literature

Ritesh Kalaskar¹, Ashita Ritesh Kalaskar²

*Departments of¹Pediatric & Preventive Dentistry and²Oral Medicine & Radiology,
Government Dental College & Hospital, Nagpur, India*

Abstract (J Korean Assoc Oral Maxillofac Surg 2018;44:198-203)

Dyke-Davidoff-Masson syndrome is a non-inherited rare condition that presents during childhood and is characterized by seizures, hemiplegia, mental retardation, cerebral hemiatrophy, calvarial thickening, and hyperpneumatization of the frontal sinuses. The present article highlights a case of a 12-year-old male child with additional clinical findings of café-au-late pigmentation and ocular lipodermoid. This is the first case report of Dyke-Davidoff-Masson syndrome to describe oral manifestations, such as unilateral delayed eruption of teeth, hypoplasia, and taurodontism, which could be unique and characteristic of this condition. Oral health care providers and physicians should be aware of these oral observations as dental referrals could warrant early dental prophylactic care and can be useful in diagnosing the possible time of injury and type of Dyke-Davidoff-Masson syndrome.

Key words: Cerebral atrophy, Enamel hypoplasia, Open bite, Impacted deciduous tooth, Seizures

[paper submitted 2017. 6. 17 / accepted 2017. 8. 16]

I. Introduction

Dyke-Davidoff-Masson syndrome (DDMS) is a rare condition that is characterized by hemi-cerebral atrophy, calvarial thickening, hyperpneumatization of the frontal sinuses, and facial asymmetry¹. It is usually secondary to trauma to the developing brain in utero or in early childhood². Trauma to the developing brain results in loss of neurons that comprise the growth of the developing brain, leading to mental retardation, seizures, and learning disability³. Presentation of classically observed characteristics depends on the age of trauma to the developing brain². Childhood presentation may differ from that of adult if the trauma to the brain presents later in life. The majority of cases reported in the literature showed that

this condition is predominant in male patients and in the left hemisphere¹. The etiology of DDMS is still controversial, but trauma, infection, intracranial bleeding, congenital vascular anomalies, and peritoneal hypoxia are the leading etiology of this syndrome. Since 1933, less than 100 cases of DDMS have been reported, but none of the cases to date have presented with oral manifestations (clinical and radiographic). The present case describes a 12-year-old male child with classical oral (clinical and radiographical) manifestations of DDMS that contributed to diagnosing the acquired type of DDMS.

II. Case Report

A 12-year-old boy reported to the department of oral medicine and radiology, with chief complaints of difficulty in mastication due to pain in the mandibular left premolar region that had persisted for one week. The patient's past history revealed an uneventful antenatal period and no complications in the post-natal period, but delayed developmental milestones were noted. There was no history of seizures or hemiparesis. The patient demonstrated slurred speech, delayed language learning, and mental retardation. Extremities were normal with both hands and feet of equal size. Facial asymmetry was noticeable, with an enlarged skull (pari-

Ritesh Kalaskar

Department of Pediatric & Preventive Dentistry, Government Dental College & Hospital, Medical College Premises, Medical Square, Nagpur 440003, India

TEL: +91-0712-2706899

E-mail: riteshpedo@gmail.com

ORCID: <https://orcid.org/0000-0001-7403-3364>

© This is an open-access article distributed under the terms of the Creative Commons Attribution Non-Commercial License (<http://creativecommons.org/licenses/by-nc/4.0/>), which permits unrestricted non-commercial use, distribution, and reproduction in any medium, provided the original work is properly cited.

Copyright © 2018 The Korean Association of Oral and Maxillofacial Surgeons. All rights reserved.

etal bone) on the left side. The facial skin on the same side showed generalized mild macular brownish diffuse pigmentation similar to café-au-late pigmentation.(Fig. 1. A, 1. B) Eye examination showed decreased vision in the left eye and a lipodermoid, for which surgical correction was advised.(Fig. 1. C) Intraoral examination revealed multiple over-retained right side maxillary and mandibular primary teeth that led to severe malocclusion with an open bite. The permanent maxillary anterior teeth were hypoplastic.(Fig. 2. A) The mandibular left second premolar was erupting buccally.(Fig. 2. B) The primary mandibular left second molar was mobile and slightly tender to vertical percussion. The patient underwent a panoramic radiograph and computed tomography (CT) scan.

The panoramic radiograph showed over-retained primary right-sided mandibular and maxillary canines and first and second molars. Delayed eruption of the permanent maxillary and mandibular canines, first and second premolars, and second molars on the right side was observed. The mandibular left second premolar was obliquely placed. Complete root and 3/4th crown resorption was present with the mandibular primary second molar, and the permanent maxillary and mandibular first molars showed taurodontism. The tooth buds of the permanent maxillary right third molar, bilateral permanent mandibular third molars, and maxillary left second premolar were missing.(Fig. 3) Overall, there was delayed eruption of the permanent teeth on the right side compared to the left, with the exception of the permanent first molars on the right side, which erupted normally in parallel with those on the left side.

The CT study of the brain (plain and contrast) showed mild

atrophy of the left cerebral hemisphere in the form of left lateral ventricle dilatation, sulcogyral space prominence, and cystic parenchymal changes. There was mild shifting of the midline structures toward the left.(Fig. 4) Other characteristic findings on the left side included frontal sinus enlargement,



Fig. 2. Intraoral photograph showing hypoplastic anterior teeth, severe malocclusion with an open bite (A), and a buccally erupting #35 (B).

Ritesh Kalaskar et al: Classical oral manifestations of Dyke–Davidoff–Masson syndrome: a case report with review of the literature. *J Korean Assoc Oral Maxillofac Surg* 2018



Fig. 1. Extraoral photograph showing facial asymmetry (A), café-au-late pigmentation (B), and lipodermoid of the left eye (C).

Ritesh Kalaskar et al: Classical oral manifestations of Dyke–Davidoff–Masson syndrome: a case report with review of the literature. *J Korean Assoc Oral Maxillofac Surg* 2018

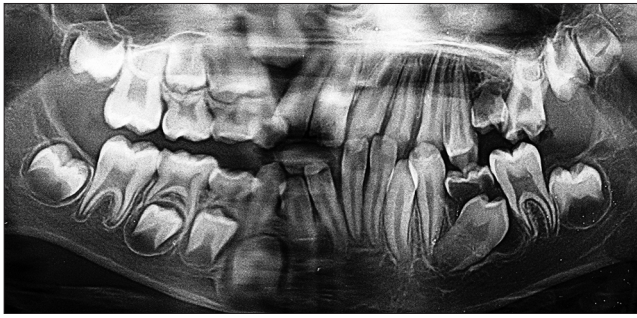


Fig. 3. Panoramic radiograph showing delayed eruption of the right side permanent teeth and taurodontism.

Ritesh Kalaskar et al: Classical oral manifestations of Dyke-Davidoff-Masson syndrome: a case report with review of the literature. J Korean Assoc Oral Maxillofac Surg 2018

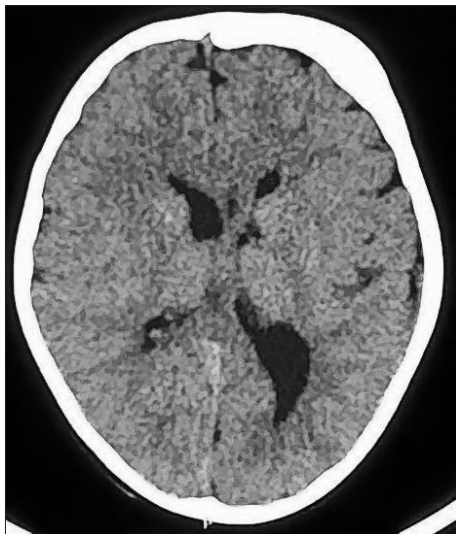


Fig. 4. Computed tomography image of the brain showing mild atrophy of the left cerebral hemisphere, dilatation of the left lateral ventricle, sulcogyral space prominence, and calvarial thickening of the frontal and parietal bones.

Ritesh Kalaskar et al: Classical oral manifestations of Dyke-Davidoff-Masson syndrome: a case report with review of the literature. J Korean Assoc Oral Maxillofac Surg 2018

calvarial thickening of the frontal bone and parietal bone, thickening of the squamous part of the temporal bone, and mild elevation of the left orbital roof.(Fig. 4, 5) A diagnosis of DDMS was made based on the characteristic imaging findings.

Considering the complexity of the case, only symptomatic treatment was planned by the pediatric dentist, which included extraction of the deciduous mandibular second molar and follow-up for the erupting mandibular second premolar. However, the patient was lost to follow-up and further treatment.

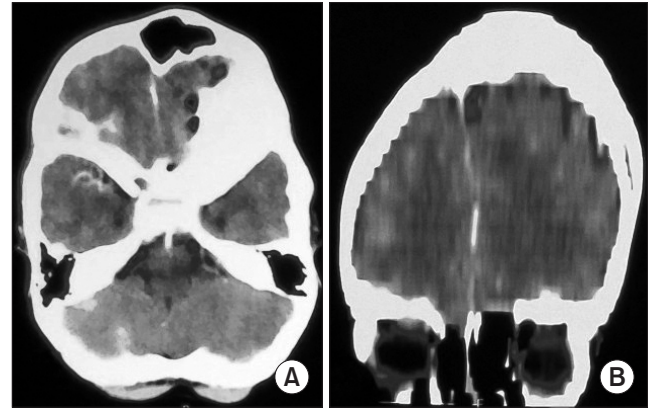


Fig. 5. Computed tomography image of the brain showing left frontal sinus enlargement, thickening of the squamous part of the temporal bone (A), and mild elevation of the left orbital roof (B).

Ritesh Kalaskar et al: Classical oral manifestations of Dyke-Davidoff-Masson syndrome: a case report with review of the literature. J Korean Assoc Oral Maxillofac Surg 2018

III. Discussion

Dyke-Davidoff-Masson syndrome was first described by Dyke, Davidoff, and Masson¹ in 1933; it is a rare syndrome with characteristic imaging findings. Two types of DDMS have been identified, congenital and acquired. The congenital type involves utero vascular occlusion of the middle cerebral artery, unilateral cerebral arterial circulation anomalies, coarctation of the mid-aortic arch, or infections, and the patient becomes symptomatic in infancy. The acquired type results from various causes such as birth asphyxia, prolonged febrile seizures, trauma, tumor, infection, ischemia, and hemorrhage^{4,5}. Thus, the age of presentation depends on the time of vascular occlusion of the middle cerebral vascular territory. This injury to the brain leads to cerebral hemiatrophy, which is associated with hypoperfusion and decreased metabolic activity of the affected cerebrum^{6,7}. The cerebral atrophy is followed by compensatory changes in the adjacent areas only if brain damage occurs before three years of age⁸. Thus, overlying structures in the brain grow inward, resulting in ipsilateral calvarial thickness, hyperneumatization of the ipsilateral paranasal sinuses, enlargement of mastoid cells, and an elevated orbital roof. Similar findings to this case have been reported, which suggests that the case could be a congenital variant of DDMS. These characteristics are very well documented and can be identified radiographically by CT imaging. Other characteristic changes, which could also be seen in these patients, are elevation of the petrous ridge and sphenoid wing, hypoplasia of the anterior or middle cranial fossa, and shifting of the midline structures toward the atrophic hemisphere^{9,10}. Such classical radiographic features have

been reported in most DDMS cases^{11–13}. Further, magnetic resonance imaging helps to clarify the extent of cerebral parenchymal involvement¹⁴. In addition to these characteristic imaging findings, clinical findings of seizures, hemiparesis, and mental retardation are also characteristic of a DDMS diagnosis^{11,12}.

To date, reported DDMS cases have included patients as young as 18 months to as old as 42 years, and almost all of the cases have reported seizures and hemiparesis of varying intensity^{11–13,15}. Because the severity of clinical features depends on extent of brain injury, in this case, the cerebral hemiatrophy was mild, and mental retardation, learning disability, speech or language disorders, and facial asymmetry were the only presenting features. While hemiparesis, seizures, and psychiatric disorders were absent, it is possible that these characteristics could manifest later in adolescence. Seizures usually postdate hemiparesis by months or years⁴, and the prognosis of these patients is considered to be better if the onset of hemiparesis is after 2 years of age and in the absence of seizures¹⁶. Thus, the prognosis in this present case could be considered better than other reported cases.

Moreover, the present case manifested with other findings that have not been reported in other DDMS cases: café-au-lait pigmentations on the facial skin of the affected side and lipodermoid of the left eye. Café-au-lait pigmentations are typically seen in neurofibromatosis type I, Albright syndrome, and Silver-Russell syndrome as well as other conditions¹⁷, whereas ocular lipodermoids are choristomas that contain adipose tissue, are commonly associated with Goldenhar syndrome¹⁸, and rarely require surgery, only if the condition interferes with vision.

Another interesting finding in this patient was delayed eruption of both maxillary and mandibular permanent teeth on the right side compared to left side. Dutta et al.¹¹ and Sethi et al.¹⁹ reported shortening of the right hand and foot in patients with left cerebral hemiatrophy. This correlates with the dental findings of the present case, even though the extremities did not demonstrate any variations. In this case, occlusal imbalance and the mobile mandibular primary second molar lead to difficulty in mastication.

This is the first case report of the DDMS syndrome identified by dental findings. In most DDMS patients, seizures and hemiparesis are significant findings that require identification and treatment on a priority basis. Therefore, it is possible that the oral cavity is neglected or given lower priority. This could provide an explanation for the absence of DDMS cases identified via classical dental findings. In the present

case, because seizures and hemiparesis were absent, the patient's parent was more concerned about the oral issue. Early identification of the dental problems could prevent already compromised patients from further dental complications. In addition, the parents of these patients could be educated and advised of preventive measures, which include proper oral hygiene, pit and fissure sealants, fluoride application, and regular dental check-ups. As these patients are on long standing anticonvulsants, there is a possibility for gingival hyperplasia, which could lead to periodontitis and early exfoliation of teeth without adequate care. There is no specific treatment for delayed eruption of teeth, but proper counselling and assurance might help prevent further aggravation of psychiatric problems in these patients.

In regard to the delayed eruption of teeth, many conditions have been reported to date; however, unilateral delayed eruption has not been reported. In addition, taurodontism in all of the permanent first molars could be an isolated finding or a component of the syndrome. Because this has not been observed in other cases, the correlation may be difficult to diagnose. Taurodontism has been reported commonly in amelogenesis imperfecta, Down syndrome, Klinefelter syndrome, and Orofaciodigital syndrome²⁰. Of note in this case, in spite of multiple delayed eruptions of permanent teeth on the right side compared to the left, the permanent first molars on the right side erupted normally. In addition, the permanent incisors on the right side were mildly delayed in eruption compared to the left side.

The initiation of calcification of the first permanent molar begins at birth, and initiation of the permanent incisors occurs at approximately six months to one year of age, with the other teeth developing later. This indicates that, in the present case, the time of injury to the brain on the left side could possibly be between the ages of six months to one year because the permanent first molars were relatively unaffected. Observation from a panoramic radiograph adds information to the CT observations that the brain injury, rather than being congenital, is actually acquired and occurred before the age of one year. The oral (clinical and radiographical) findings thus serve as important adjuvant observations that can guide interpretation of the possible time of injury to the brain and, hence, the type of DDMS.

A differential diagnosis of DDMS includes Sturge-Weber syndrome, Rasmussen encephalitis, hemiconvulsion-hemiplegia-epilepsy (HHE), Silver-Russell syndrome, basal ganglia germinoma, Fishman syndrome, and linear nevus syndrome^{2,9,10}. The Sturge-Weber syndrome classically in-

cludes port-wine facial nevus, leptomeningeal angioma, and intracranial tram track calcification in addition to cerebral atrophy. Rasmussen encephalitis is an immune-mediated brain disorder that affects children between 6 and 8 years of age, leading to unilateral hemispheric atrophy, intractable focal epilepsy, and progressive neurological dysfunction. This condition can be very well differentiated on the basis of imaging features, which include unihemispheric focal cortical atrophy and ipsilateral head of the caudate nucleus, without any calvarial changes²¹. HHE is characterized by unilateral prolonged clonic seizures followed by the development of hemiplegia in younger children²². Patients with Silver-Russell syndrome have a normal head circumference and normal intelligence but poor growth, clinodactyly, hemi-hypertrophy, and a characteristic facial phenotype (triangular face, small pointed chin, broad forehead, and a thin wide mouth)²³. Basal ganglia germinoma is a rare tumor of the brain, associated with progressive hemiparesis and cerebral hemiatrophy. However, imaging reveals cystic areas, focal hemorrhages, and mild surrounding edema along with calvarial changes²⁴. Fishman syndrome or encephalocraniocutaneous lipomatosis is a rare neurocutaneous syndrome that includes unilateral cranial lipoma with lipodermoid of the eye, which typically presents with seizures. Neuroimaging, however, shows a calcified cortex and hemiatrophy²⁵. Linear nevus syndrome could be differentiated by typical facial nevus and unilateral ventricular dilatation that resembles cerebral hemiatrophy, in addition to recurrent seizures and mental retardation²⁶. Treatment options in symptomatic patients include anticonvulsion drugs for control of seizures, physiotherapy for hemiparesis, speech therapy for speech defects, and psychiatric counseling and medications, if required. For intractable and uncontrolled seizures and hemiplegia, hemispherectomy is the treatment of choice and has a reported 85% success rate¹⁶.

In conclusion, this study reports a possible mild case of DDMS with absence of hemiparesis and seizures. Classical oral manifestations are reported and emphasize the need for dental referrals for such patients to provide preventive oral care, in addition to early detection of malocclusion and other oral problems to provide prompt treatment and avoid further oral complications in an already compromised patient. These oral observations can also be useful in diagnosing this type of DDMS.

ORCID

Ritesh Kalaskar, <https://orcid.org/0000-0001-7403-3364>

Ashita Ritesh Kalaskar, <https://orcid.org/0000-0002-1415-5033>

Authors' Contributions

R.K. wrote the manuscript, photographs of patients. A.R.K. helped in drafting and reading/approving of manuscript.

Consent for Publishing Photographs

Written informed consent was obtained from the guardian of patient for publication of this article and accompanying images.

Conflict of Interest

No potential conflict of interest relevant to this article was reported.

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. Aguiar PH, Liu CW, Leitão H, Issa F, Lepski G, Figueiredo EG, et al. MR and CT imaging in the Dyke-Davidoff-Masson syndrome. Report of three cases and contribution to pathogenesis and differential diagnosis. *Arq Neuropsiquiatr* 1998;56:803-7.
3. Singh P, Saggat K, Ahluwalia A. Dyke-Davidoff-Masson syndrome: classical imaging findings. *J Pediatr Neurosci* 2010;5:124-5.
4. Sener RN, Jenkins JR. MR of craniocerebral hemiatrophy. *Clin Imaging* 1992;16:93-7.
5. Stred SE, Byrum CJ, Bove EL, Oliphant M. Coarctation of mid-aortic arch presenting with monoplegia. *Ann Thorac Surg* 1986;42:210-2.
6. Demir Y, Sürücü E, Çilingir V, Bulut MD, Tombul T. Dyke-Davidoff-Masson syndrome with cerebral hypometabolism and unique crossed cerebellar diaschisis in 18F-FDG PET/CT. *Clin Nucl Med* 2015;40:757-8.
7. Shrestha B. Acquired cerebral hemiatrophy: Dyke-Davidoff-Masson syndrome--a case report. *Turk Neurosurg* 2013;23:117-21.
8. Solomon GE, Hilal SK, Gold AP, Carter S. Natural history of acute hemiplegia of childhood. *Brain* 1970;93:107-20.
9. Uduma FU, Emejulu JK, Motah M, Okere PC, Ongolo PC, Muna W. Differential diagnoses of cerebral hemiatrophy in childhood: a review of literature with illustrative report of two cases. *Glob J Health Sci* 2013;5:195-207.
10. Arora R, Rani JY. Dyke-Davidoff-Masson syndrome: imaging features with illustration of two cases. *Quant Imaging Med Surg* 2015;5:469-71.
11. Dutta A, Bose S, Sen K, Pandit N, Sharma S. Refractory seizure in childhood: Dyke-Davidoff-Masson syndrome revisited. *Oman Med J* 2016;31:304-8.
12. Kumar NV, Gugapriya TS, Guru AT, Kumari SN. Dyke-Davidoff-Masson syndrome. *Int J Appl Basic Med Res* 2016;6:57-9.
13. Jain D, Aggarwal HK, Goyal S, Mittal A. Dyke-Davidoff-Masson syndrome: a rare case report. *Iran J Neurol* 2014;13:255-6.

14. Shen WC, Chen CC, Lee SK, Ho YJ, Lee KR. Magnetic resonance imaging of cerebral hemiatrophy. *J Formos Med Assoc* 1993;92:995-1000.
15. Roy U, Panwar A, Mukherjee A, Biswas D. Adult presentation of Dyke-Davidoff-Masson syndrome: a case report. *Case Rep Neurol* 2016;8:20-6.
16. Behera MR, Patnaik S, Mohanty AK. Dyke-Davidoff-Masson syndrome. *J Neurosci Rural Pract* 2012;3:411-3.
17. Kumar LSV. Café au lait spot: case report. *J Adv Clin Res Insights* 2014;1:106-7.
18. Bhallil S, Benatiya I, El Abdouni O, Mahjoubi B, Hicham T. Goldenhar syndrome: ocular features. *Bull Soc Belge Ophtalmol* 2010;17-9.
19. Sethi NK, Sethi PK, Torgovnick J, Arsura E. Acquired Dyke-Davidoff-Masson syndrome: a clinicoradiographic correlation. *Eastern J Med* 2011;16:269-71.
20. Witkop CJ Jr, Keenan KM, Cervenka J, Jaspers MT. Taurodontism: an anomaly of teeth reflecting disruptive developmental homeostasis. *Am J Med Genet Suppl* 1988;4:85-97.
21. Varghese B, Aneesh M, Singh N, Gilwaz P. A case of rasmussen encephalitis: the differential diagnoses and role of diagnostic imaging. *Oman Med J* 2014;29:67-70.
22. Auvin S, Bellavoine V, Merdarius D, Delanoë C, Elmaleh- Bergés M, Gressens P, et al. Hemiconvulsion-hemiplegia-epilepsy syndrome: current understandings. *Eur J Paediatr Neurol* 2012;16:413-21.
23. Qiu BP, Shi CH. Silver-Russel syndrome: a case report. *World J Pediatr* 2007;3:68-70.
24. Moon WK, Chang KH, Kim IO, Han MH, Choi CG, Suh DC, et al. Germinomas of the basal ganglia and thalamus: MR findings and a comparison between MR and CT. *AJR Am J Roentgenol* 1994;162:1413-7.
25. Amor DJ, Kornberg AJ, Smith LJ. Encephalocraniocutaneous lipomatosis (Fishman syndrome): a rare neurocutaneous syndrome. *J Paediatr Child Health* 2000;36:603-5.
26. Jacoby CG, Go RT, Hahn FJ. Computed tomography in cerebral hemiatrophy. *AJR Am J Roentgenol* 1977;129:5-9.