CASE REPORT

Dyke Davidoff Masson Syndrome with Hypothyroidism Complicated with Respiratory and Urinary Tract Infections: A Case Report

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Abstract

The Dyke-Davidoff-Masson Syndrome (DDMS) is a rare neurological disorder characterized by unilateral cerebral atrophy. Clinical features may include seizures, hemi paresis, facial asymmetry, and intellectual disability. The causative factors include an insult to the brain in utero or early infancy *e.g.*, CNS infections, hypoxic-ischemic encephalopathy, intracranial haemorrhage, trauma, inherited vascular malformations, but in some cases, no risk factors are found. Here, we present the case of a known epileptic patient with a history of stroke who presented with fever, abdominal pain with distention, and facial swelling. The diagnosis was established after clinical history, examination, and MRI findings.

Keywords: *Dyke Davidoff Masson syndrome, epilepsy, stroke, hemi-atrophy, early onset seizures.*

INTRODUCTION

The Dyke-Davidoff-Masson syndrome (DDMS) is a rare neurological disorder which was presented for the first time in 1933 by researchers of the same name [1]. It is characterized by facial asymmetry, epilepsy, hemiparesis, cognitive challenges, and intellectual disabilities [2]. Diagnosis is made through imaging studies that are correlated with clinical symptoms. Specific radiological findings include cerebral hemiatrophy and enlarged ventricles. Additionally, there may be thickening of skull bones and hyperpneumatization of sinus [3, 4].

Case Presentation

A 26-year-old male with, a known case of epilepsy since childhood, controlled *via* medication (Epival), presented with high grade, intermittent fever for 7 days, and abdominal pain with distention for 3 days and facial swelling for 1 day. On further inquiry, the patient also complained of productive cough and dysuria with urgency and frequency. The patient also gave a history of constipation, joint pain, dryness of skin, cold intolerance, peri-orbital puffiness, pedal edema and oral ulcers. The patient had a significant history of delayed cry, cyanosis and jaundice at birth and a history of ischemic stroke 15 years back. On examination, the patient was of average height and build, with stable vitals. He was anemic, had bilateral pedal edema and raised JVP. There was no clubbing, jaundice, or lymphadenopathy and the thyroid was not palpable. Chest examination revealed crepitations audible on the mid-lower side of the left chest with dull percussion notes and decreased vocal resonance. On CNS examination, GCS was 15/15, and pupils were BERL, with left-sided facial nerve palsy of UML type. The rest of the cranial nerves were intact. On motor system examination, the left side of the body had reduced bulk. Powers were 3/5 and reflexes were +3, with clonus in both upper and lower limbs. Hoffman's sign was negative. The right-sided examination was normal. Abdominal examination revealed shifting dullness but no hepatosplenomegaly. On lab investigations, there was low haemoglobin, raised WBCs and inflammatory markers (**Table 1**).

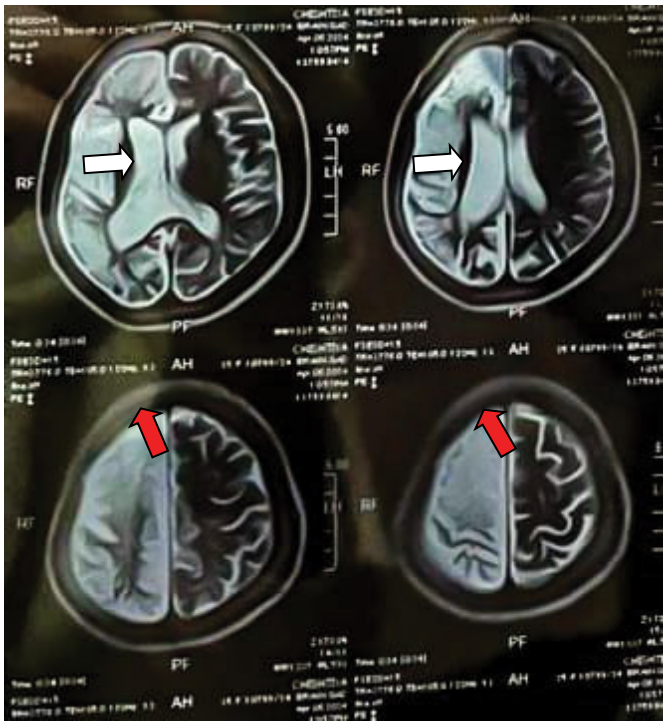

Fig. (1): Axial T2-weighted MRI of the brain showing hemiatrophy of the right cerebral hemisphere along with ex-vacuo dilatation of the ipsilateral lateral ventricle (white arrows) and hypertrophy of the right side of calvarium predominantly the frontal bone (red arrows).

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Received: February 09, 2025; Revised: April 13, 2025; Accepted: April 25, 2025
DOI: https://doi.org/10.37184/jlnh.2959-1805.3.21

  ISSN: 2960-2963 (Online) *Journal of Liaquat National Hospital 2025; 3(2): 168-170*

Table 1: Lab investigations.

Investigations	On presentation	After treatment	Additional Investigations on Presentation	
Hb (mg/dl)	5.1	8.01	ESR	120 mm/hr
MCV (fl)	89.5	83.2	TSH	12.7 mIU/L
Ret (%)	1.39	-	FT3	1.5 pmol/L
TLC (/µL)	27.6	8.7	FT4	0.9 pmol/L
Lympho (%)	11	38	Phos	3.7 mg/dl
Neutro (%)	75	46	Mg	1.8 mg/dl
Plat (/µL)	23	284	Amy	56 U/L
CRP (mg/dl)	266.3	20.5	HbA1c	5.7%
Na (mmol/L)	135	138	LDH	129 U/L
K (mmol/L)	3.6	3.7	UA	4.5 mg/dl
Cl (mmol/L)	99	102	Cho	118 mg/dl
Ca (mg/dl)	8.3	8.3	TGs	132 mg/dl
T.Bili (mg/dl)	0.6	0.5	HDL	11.9 mg/dl
SGPT (U/L)	11	18	LDL	80 mg/dl
Alk Phos (U/L)	147	262	MP	Negative
Albumin (g/dl)	2.8	2.4	Dengue	Negative
PT (sec)	13.8	11.1	HbsAg	Negative
INR	1.32	21.4	Anti-HCV	Reactive
APTT (sec)	26.6	1.06	Anti-thyroid Peroxidase Antibodies	Negative
BUN (mg/dl)	14	6	-	-
Creat (mg/dl)	0.4	0.5	-	-
UCS	Growth of Candida			
AsDR	Bloodstained, protein=1.5, albumin=0.8, WBCs=437, Polymorphs=35%, Lymphocytes=65%			
UDR	pH=8.5, protein 2+, ketones +, leucocytes 3+, RBCs 10-15, pus cells 30-35, epith cells 2+			

He was found to be Hepatitis C positive. Liver, renal and coagulation profiles were normal. Thyroid profile showed raised TSH, low FT3 and FT4. Ascitic fluid D/R was transudative. Urine D/R showed pus cells and leucocytes, with leucocyte esterase and nitrates positive. The pan cultures and TB workup were negative. Ultrasound abdomen showed coarse echotexture of the liver, thick-walled gallbladder and moderate ascites. Echocardiography was normal. CT CAP showed pleuro-peritoneal infection with sub-centimetre-sized mediastinal, and bilateral hilar lymph nodes, mild to moderate free fluid in the abdominopelvic cavity with mild mesenteric and peritoneal fat stranding. Multiple sub-centimetre-sized mesenteric and para-aortic lymph nodes were also appreciated. These findings likely represented an infective/ inflammatory etiology. MRI brain with contrast revealed hemiatrophy of the right cerebral hemisphere, along with ex-vacuo dilatation of the ipsilateral lateral ventricle. There was hypertrophy of the right side of the calvarium, predominantly the frontal bone, raising the possibility of Dyke-Davidson-Masson syndrome (**Fig. 1**).

Keeping all the findings in consideration, the patient was kept on broad spectrum antibiotics and thyroxin 25µg. A significant improvement was noted and patient got discharged with advice to follow-up.

DISCUSSION

Dyke, Davidson and Masson were the first researchers to describe this extremely rare syndrome, with less than 100 cases reported worldwide [1, 5]. Its primary symptoms include early-onset seizures, reduced cognitive function, hemiparesis, and facial asymmetry [2]. CT and MRI are the most commonly used imaging techniques for diagnosing this syndrome, with key findings such as cerebral hemiatrophy accompanied by compensatory enlargement of the ventricles [2, 3]. Plain X-ray films can also show bony changes like thickening of calvarial bones, sphenoidal and frontal sinus dilation, elevation of the greater wing of the sphenoid and petrous ridge, and slanting of planum-sphenoidal [6].

This syndrome is categorized into two subtypes based on the age at which it appears: congenital, which occurs in infancy, and acquired, which appears in early childhood. The congenital form is believed to result from brain hypoxia during fetal development, whereas the acquired form is linked to risk factors such as central nervous system infections, trauma, ischemic incidents, and cerebrovascular events [4]. There have also been

a few reported cases of DDMS in adults, which are notably idiopathic [7]. Our patient has a history of at-home, post-term birth at 42 weeks with delayed cry and cyanosis which points to the possibility of a hypoxemic CNS event during prenatal or perinatal period.

Keeping in mind the presenting complaints of our patient, the initial differential diagnoses that were considered included peritoneal/disseminated Tuberculosis, hypothyroidism, Amyloidosis, MELAS (Mitochondrial encephalomyopathy with lactic acidosis and stroke-like episodes), and HIV with complications. The distinguishing features of MRI Brain with subsequent clinical correlation ruled out these conditions in our patient.

Our patient presented with hypothyroidism, which can be attributed to a side effect of Sodium valproate [8], as the autoimmune workup of this patient was negative. TSH levels may be restored to normal after the withdrawal of valproate, as demonstrated in a study by Attilakos *et al.* [9].

The treatment of DDMS involves long-term antiepileptic drug therapy; however, many patients with this syndrome experience drug-resistant epilepsy [8]. In addition to antiepileptic medications, the treatment also includes physical, speech, language, and occupational therapy [3]. For children with severe, intractable seizures and hemiplegia, hemispherectomy may be considered, which has an 85% success rate. The prognosis is generally better for patients who develop hemiparesis after the age of 2 and who do not have prolonged or recurrent seizures [10].

CONCLUSION

Because Dyke Davidoff Masson syndrome is so rare, it can easily be overlooked by even seasoned practitioners. Accurate diagnosis relies on a comprehensive history, a detailed physical examination, and specific radiological findings.

CONSENT FOR PUBLICATION

Written informed consent was taken from the patient.

CONFLICT OF INTEREST

The authors have no conflict of interest to declare.

ACKNOWLEDGEMENTS

Declared none.

AUTHORS' CONTRIBUTION

All authors reviewed and approved the final manuscript. MG prepared the case and wrote the first draft. AF and SMHZ finalized the draft and prepared the images. SMH supervised the case and the article.

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