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Dyke-Davidoff-Masson Syndrome (DDMS) in Adult: A Rare Case in Rural Area

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ABSTRACT

Introduction: Dyke-Davidoff-Masson syndrome (DDMS) could be a rare neurological disorder characterized by cerebral hemiatrophy and an engine shortfall on the contralateral side of the body. Furthermore, facial asymmetry and seizures are common symptoms. Classic imaging findings in individuals with DDMS are ventriculomegaly, cerebral hypoplasia, paranasal sinus hyperpneumatization, and compensatory osseous broadening. The relationship between symptoms and imaging findings is used to determine DDMS. **Case:** A 46-year-old female patient arrived at the hospital with an inability to speak three days before admission, refused to eat or drink, and had a fever. She also had a slanted mouth and right-sided hemiparesis with a history of seizures. A firm boundary consolidation on the right pericardium was found on the chest X-ray, suggesting pulmonary TB. On a non-contrast head CT scan, thickening of the left hemicalvaria was found, accompanied by a widening of the left frontal-ethmoidal sinus, hyperpneumatization of the left mastoid air cell, and left cerebral hemiatrophy, all accompanied by left lateral ventricular dilatation. **Conclusion:** The proper diagnosis and early causative treatment significantly improve the outcome of the disease.

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INTRODUCTION

Dyke-Davidoff-Masson syndrome (DDMS), first described in 1933, is an extremely rare condition that makes it challenging for medical professionals to identify and distinguish it from other illnesses, resulting in underdiagnosis. Since 1963, less than 100 cases of DDMS have been reported, with 21 cases among adults.[1] The Dyke-Davidoff-Masson syndrome (DDMS) results from an insult to the growing brain in utero or early infancy, leading to the loss of neurons and compromising the growth of the brain. DDMS can be congenital or acquired due to an alteration in cerebral perfusion during the prenatal, perinatal, or early childhood periods.

In 1933, Dyke, Davidoff, and Masson documented nine patients with pneumatic encephalographic abnormalities on a skull radiograph who had hemiparesis, facial asymmetry, seizures, and mental retardation.[2] Cerebral hemiatrophy, ipsilateral ventriculomegaly, hyperpneumatization of the sinuses on the affected side, and compensatory calvarial thickening are all characteristics of this entity as shown on CT and MRI.[3,4] The treatments consist of seizures control with suitable anticonvulsant, physiotherapy, speech therapy and occupational treatment.[5]

We reported a 46-year-old female patient with clinical symptoms and radiological features that led to the diagnosis of Dyke-Davidoff-Masson syndrome. The symptoms of DDMS are often underdiagnosed. Therefore, proper history-taking, physical examination, and investigations are needed to assist clinicians in establishing this diagnosis.

CASE

A 46-year-old female patient presented to the RSUD Prof. Dr. WZ Johannes Kupang emergency department with the chief complaints of inability to speak three days before hospital admission, refusal to eat and drink, and fever three days prior. The patient had also obtained a slanted mouth three days before hospitalization. Since seven months ago, the patient has only been in bed and has started talking incoherently (unlike before). There was no history of seizures, nausea, vomiting, or headache.

A history of right-sided weakness and deformity was documented in the first year of life. In her first year of life, the patient also had seizures but did not seek treatment because she was far from healthcare facilities. In addition to seizures, the patient also had speech and motor delays. The patient did not attend school; she remained at home and is being cared for by his brother. From the age of 20, the patient began to eat, drink and bathe on her own; then, at the age of 30, she was taught by relatives to cook rice and clean the house, and she was very good at it.

This patient's seizures continued for 15 years without treatment. There was no family history of seizures. In the history of vaginal delivery, the baby cried immediately, at term, with a birth weight of 2.5 kg. During pregnancy, the patient's mother did not regularly check with the midwife or doctor; there was no known infectious disease and there was no history of trauma.

On physical examination, the patient's *Glasgow Coma Scale* (GCS) was E4VxM5, with global aphasia, dysphagia, central type VII right nerve paresis, and positive neck stiffness. On physical examination of the lungs, crackles were found at the base of the right lung field. On motor examination, there was an impression of right lateralization with spastic tone and the presence of contractures in the right limb. There was an increase in physiological reflexes in the right limb and positive Babinski sign on the right leg, but no autonomic system disturbance was found. Following that, blood tests, including liver and kidney function, were normal. Blood glucosea and blood electrolytes (Na, K, Cl, Ca) were within normal limits.

The patient was then examined for a chest X-ray and a CT scan of the head without contrast. On chest X-ray examination, it was found that there was a firm boundary consolidation on the right pericardium, leading to pulmonary TB-DD malignancy and the presence of right pleural effusion. On a non-contrast head CT scan (Figure 1), thickening of the left calvarial hemi was found, accompanied by widening of the left frontal-ethmoidal sinus, hyperpneumatization of the left mastoid air cell, and left cerebral hemiatrophy, all accompanied by left lateral ventricular dilatation. (Figure 2)

For the management, patients were given NaCl 0.9% infusion at 20 drops per minute, ceftriaxone injection 2x1 g, piracetam injection 3x3 g, omeprazole injection 2x1 ampoule, dexamethasone injection 4x1 ampoule, paracetamol infusion 3x1 g, a nasogatric tube was inserted, and folic acid was given 3x1 mg, vitamin B complex 2x1 tablet, OAT category 1, MLP 6x200cc. In non-pharmacological management, this patient was referred to the medical rehabilitation unit to undergo physiotherapy, dysphagia therapy, and other therapies.

DISCUSSION

Clinically, the most typical symptoms of tuberculous meningitis (TBM) include a fever that lasts longer than two to three weeks, neck stiffness, and/or unconsciousness. In the more advanced phases

of the illness, papilledema and cranial nerve palsy are more frequently observed. Additionally, patients may develop seizures, aphasia, visual loss, and hemiparesis. The Dyke-Davidoff-Masson syndrome is characterized by facial asymmetry, seizures, hemiplegia or contralateral hemiparesis, and mental retardation of variable severity. Seizures can be focal or generalized. In some cases, psychiatric disorders can be found.[6] This female patient was found with a fever 3 days prior, accompanied by decreased consciousness, history of recurrent seizures, contralateral hemiparesis, possible mental retardation and learning disorders in the absence of psychiatric disorders, and positive neck stiffness. Fever and positive neck stiffness are the uncommon symptoms in DDMS; they are more common in meningitis TB, which is then supported by firm boundary consolidation on the right pericardium finding in the patient's chest x-ray.

TB disease may cause the advancement of parenchymal (cortical) and meningeal tuberculomas. Clinical examination and microbiological tests are the most commonly used methods for diagnosing TBM. The detection of TBMs in cerebrospinal fluid (CSF) utilizing spread microscopy, culture, DNA enhancement testing, and MRI is required in cases of tuberculoma.[7,8,9]

Tuberculous meningitis can be assessed by getting cerebrospinal fluid (CSF) for examination. Ordinarily, the CSF shows low blood sugar, a rise in protein, and a slightly raised white blood cell tally with a lymphocytic prevalence. The CSF examination most closely resembles the CSF examination of a viral meningitis disease. The conclusion of TBM could be advanced and supported by neuroimaging. When it comes to assessing the brainstem and spine to discover TBM, MRI has demonstrated that it is more prevalent than computed tomography (CT). Imaging can evaluate meningeal enlargement, cerebral edema, and infarcts in the brain. CT imaging is the best method for excluding an urgent hydrocephalus problem associated with TBM that would necessitate prompt neurosurgery intervention. Basal exudates can also be seen on a CT scan.[10]

Dyke-Davidoff-Masson syndrome can be congenitally present or acquired. There are typically no clear etiological factors or symptoms associated with the congenital type at birth or soon after. A fetal intrauterine vascular occlusion most likely caused the brain injury. In the acquired type, symptoms are linked to a central nervous system injury that occurse during or shortly after the perinatal period. Trauma, infection, vascular malformations, hemorrhagic and ischemic circumstances, amniotic bands, an intraventricular and subependymal germinal matrix, and amniotic bands are a few of the etiological causes.[11,12] The cause of DDMS in this patient was still unknown due to a lack of information obtained from the family and a lack of routine examinations during pregnancy and childbirth. Furthermore, the patient did not have a lumbar puncture because the family refused, and there was no MRI to confirm the diagnosis of tuberculoma.

Large cerebral sulci, lateral ventricular enlargement, brain hemiatrophy, frontal sinus hyperpneumatization, and compensatory skull thickening are all visible on one side of the MRI, according to the DDMS imaging findings. These imaging results become increasingly obvious as the child ages. Differential diagnoses for cerebral hypoplasia include Rasmussen encephalitis, Sturge-Weber syndrome, Fisherman syndrome, and brain tumors. A total clinical history, multisystem appraisal and paraclinical examination regularly lead to precise determination.[12,13] In this patient, only a CT scan of the head without contrast was performed due to limited equipment and no MRI. The results showed thickening of the left hemicalvaria, widening of the left frontal-ethmoidal sinus, hyperpneumatization of the left mastoid air cell, and left cerebral hemiatrophy accompanied by left lateral ventricular dilatation. The shortage in this article is that no lumbar puncture, MRI, and IQ tests were performed to diagnose tuberculous meningitis, DDMS, and mental retardation in these patients.

The antituberculosis regimen, consisting of rifampin, isoniazid, pyrazinamide, and ethambutol for two months in the intensive phase and rifampin and isoniazid for 7 to 10 months in the continuation phase (2RHZE/4RH), was used to treat extrapulmonary tuberculosis. Experts recommend antituberculosis pharmacological therapy for tuberculous meningitis for at least 9 to 12 months. In addition to OAT, corticosteroid administration in treating tuberculous meningitis has been reported to be useful in several studies. Administration of intravenous corticosteroids reduced the mortality rate.[14] Combining dexamethasone with an antituberculosis regimen can improve the adequacy of TB treatment and diminish the rate of an antagonistic response.[15]

No spesific protocol exist for the treatment of DDMS; instead, anticonvulsant medications, which are also used to treat convulsions, are typically used to manage symptoms. Children with hemiplegia and frequent, severe seizures may be candidates for hemispherectomy, which has an 85 percent success rate in carefully selected patients. Physiotherapy, occupational therapy, and linguistic therapy are the cornerstones of long-term patient management.[16]

For the treatment of tuberculous meningitis, this patient received a category 1 OAT regimen that will be continued for at least 9 months. She also received a 4x1 ampoule injection of dexamethasone. This patient had no seizures since the age of 15 years, so

anti-epileptic drugs were not given. Due to a lack of equipment, this patient did not undergo a hemispherectomy, but she was referred to the medical rehabilitation unit for physiotherapy, dysphagia therapy, and other therapies aimed at allowing the patient to perform minimal activities such as before entering the hospital.

The prognosis will be better if the patient responds well to OAT treatment and dexamethasone. The prognosis of the syndrome is favorable if the motor impairment appears after two years and is accompanied by manageable seizures.[17] In this case, the prognosis is good because, after five days of treatment, the patient's consciousness and general condition improved. There were no recurrent seizures even though hemiparesis was found at less than two years old.

CONCLUSION

Dyke-Davidoff-Masson syndrome is a rare neurological condition with several characteristics, including recurrent seizures, hemiparesis, and intellectual disability with hemiatrophy, skull hypertrophy, and sinus hyperpneumatization. It is often underdiagnosed by physicians. In this case, suspected DDMS was present alongside suspected meningitis TB. It is critical to evaluate DDMS as a possible differential diagnosis for all patients presenting with facial asymmetry, recurrent seizures, intellectual disability, and hemiparesis so that the patients can be adequately managed.

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Author contributions
IP contributed to the conceptualization, resources, writing, review, and editing.

Conflict of Interest
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TABLES AND FIGURES

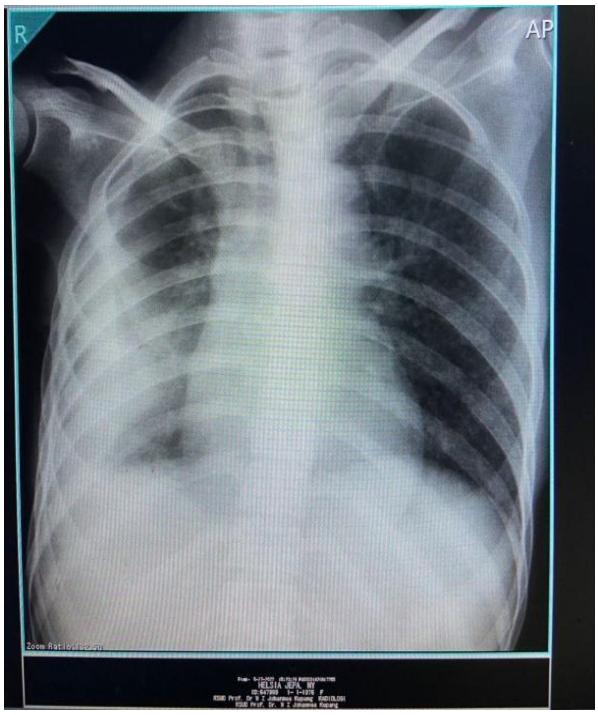

Figure 1. Thorax Rontgen

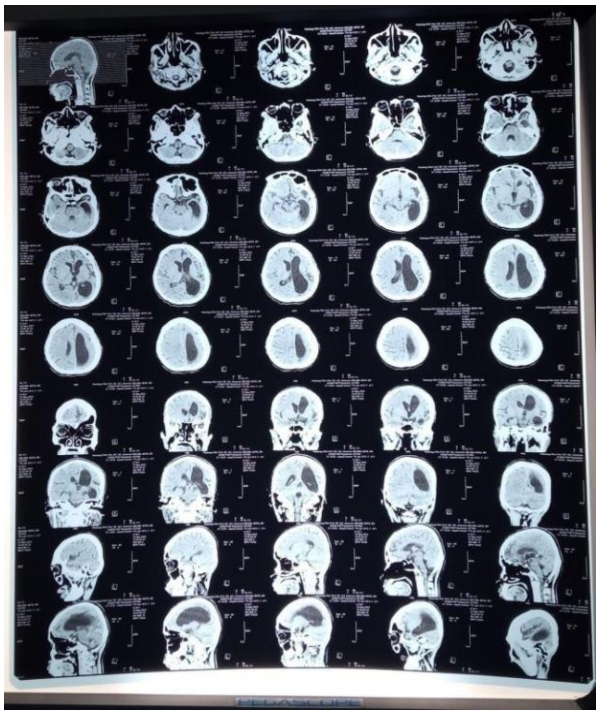

Figure 2. Head CT Scan Without Contrast