Psychosis in a Patient with Davidoff-Dyke-Masson Syndrome

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Abstract

Objectives: To report the finding of psychosis in a patient with Davidoff-Dyke-Masson Syndrome. **Method:** Case report. **Conclusions:** Right-sided hemiatrophy may be an addition to the list of neuro-developmental and structural cerebral anomalies associated with psychotic disorders including schizophrenia.

Key Words: Davidoff-Dyke-Masson Syndrome, Psychosis, Schizophrenia

Introduction

In 1933, Davidoff, Dyke, and Masson described the plain skull radiographic and pneumato-encephalographic changes in a series of nine patients characterized clinically by hemiparesis, seizures, facial-asymmetry, and mental retardation (1). The radiologic findings included cerebral hemiatrophy, ventricle enlargement, shift to the affected side, dilation of sulci, and compensatory skull changes. Features of the skull were asymmetry, ipsilateral osseous hypertrophy of the calvarium and hyperpneumatization of the sinuses (2). For over two decades, neurodevelopmental alterations have been discussed as underlying risk factors for the development of psychotic disorders (3). As Davidoff-Dyke-Masson Syndrome (DDMS) is a pre- or perinatally acquired entity, it is of interest in this respect (4). Age of clinical presentation and appearance of the entire spectrum of characteristic clinical and radiological features depends upon the timing of the central nervous system insult (5). Psychiatric symptomatology has been only rarely reported; there have been reports of presentation as childhood-onset schizophrenia, adolescent-onset schizophrenia like psychosis and treatment-resistant schizoaffective disorder (6-8). In this case report we describe a patient with Davidoff-Dyke-Masson Syndrome presenting with psychosis.

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Case Report

A 25-year-old female born full term of non-consanguineous union presented with childhood-onset seizures, learning disability and psychotic symptoms for the last two years. Her antenatal and postnatal period was uneventful and she attained developmental milestones, age appropriately. She was apparently normal until three years of age when she developed generalized tonic clonic seizures. She was taken to faith healers and not given medical treatment for seizures. In the recent past (from five years) the seizure episodes were increasing in frequency to about one episode every three weeks.

She was brought to the hospital with complaints of paranoid thoughts about being harmed, spied upon and observed for the last two years. She reported hearing a male voice talking to her continuously. These auditory hallucinations were present in the absence of seizures. She reported of visual imagery in the form of a "black body." No Schneider

first rank symptoms were reported during the course of her illness. Family history was significant for mental retardation in a second-degree relative.

Neurological examination was within healthy limits except for bilateral brisk deep tendon reflexes. She had no neurocutaneous markers. Mental status examination revealed delusions of persecution, reference, second-person auditory hallucinations with visual imagery in the form of a "black body." Routine laboratory investigations were significant for the finding of hemoglobin (10.7 gm/Dl) and dimorphic anemia on peripheral smear. Her Intelligence Quotient (IQ) on Wechsler Adult Performance Intelligence Scale (WAPIS) was determined to be 57, suggestive of mild mental retardation. Electroencephalography (EEG) showed abnormal bursts of generalized slow wave discharges suggestive of bihemispheric dysfunction. Computerized Tomography (CT) scan of brain showed diffuse atrophic changes involving the right cerebral hemisphere with ipsilateral thickened calvarium, enlarged frontal sinus and mildly dilated third and right lateral ventricle (see Figures 1–4). It also showed an old lacunar infarct in left basal ganglia. Based on clinical and radiological findings, she was diagnosed with Davidoff-Dyke-Masson Syndrome. Her psychiatric presentation was differentially diagnosed: organic schizophrenia-like psychotic disorder versus psychosis NOS. She was started on sodium valproate (1,000 mg) for seizures and her psychotic symptoms remitted with olanzapine 10 mg/d. Anemia was treated with tablet ferrous sulphate (100 mg/d), cyanacobalamin

Figure 1 (Axial Non Contrast CT Scan-Head): Enlarged Third Ventricle on the Right Side

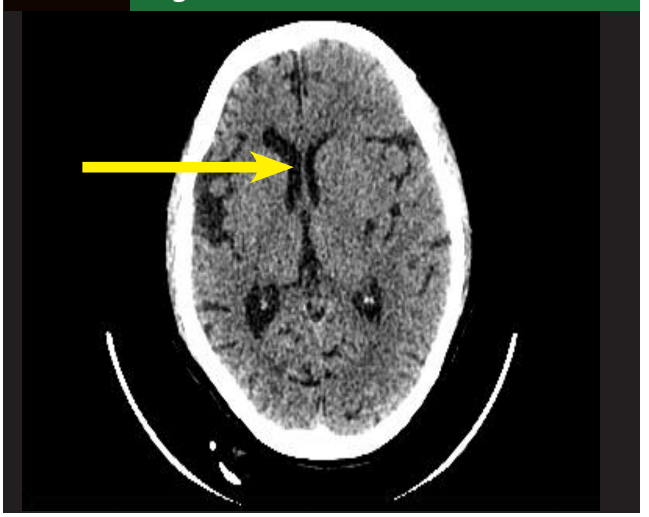


Figure 2 (Axial Non Contrast CT Scan-Head): Diffuse Right-Sided Cerebral Atrophy

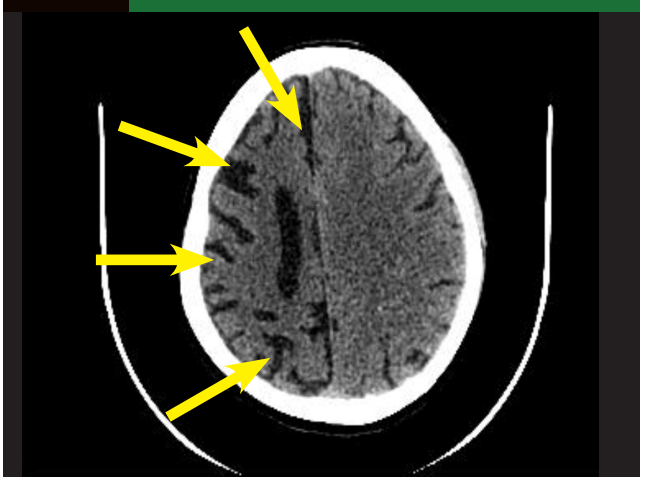


Figure 3 (Coronal Non Contrast CT Scan-Head): Dilated Frontal Horn of the Lateral Ventricle and Prominent Cerebral Sulci on the Right Side

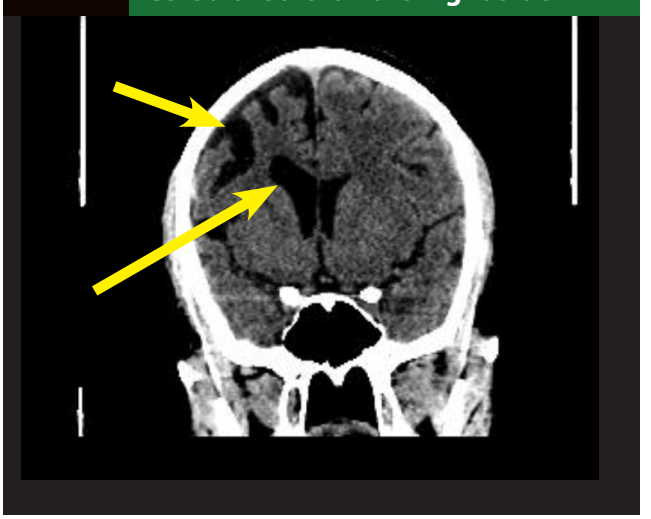
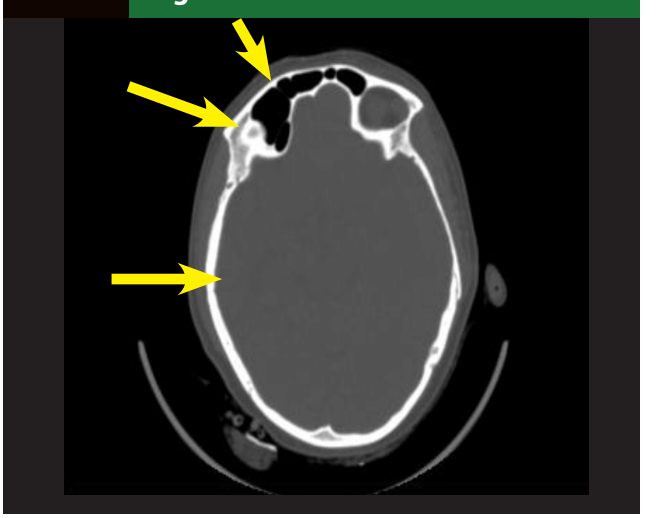


Figure 4 (Axial Non Contrast CT Scan-Head): Thickening of the Calverium and Dilated Frontal Sinus on the Right Side



(1,500 mcg/d) and folic acid (5 mg) for six months. During the course of follow-up (over one year) she has been doing well, with remission of psychotic symptoms and her seizures well controlled. She is working as a daily wage laborer and earning her livelihood for the last six months.

Discussion

Etiopathogenesis of Davidoff-Dyke-Masson Syndrome has been described as either due to perinatal insult or due to any brain damage sustained in the first three years of life (9). Brain damage can be due to trauma, tumor, infection, ischemia, hemorrhage or even prolonged febrile seizures. Age of presentation depends on time of insult and characteristic changes may be seen only in adolescence or adulthood (9). Our patient had generalized tonic clonic seizures at the age of three years and psychotic symptoms in adulthood.

Psychotic disorders—particularly schizophrenia—have been associated with brain volume loss, ventricular enlargement, striatal abnormalities and cerebral asymmetry (3). There is an argument that right hemisphere language functions are necessary for successful social communication and the core deficit in psychosis is a failure of segregation of right from left hemisphere functions (10). There is also a significant association with perinatal and neonatal hypoxia, adding credence to a neurodevelopmental model vis-à-vis a neurodegenerative hypothesis (11). The psychotic symptoms in our patient can be explained empirically on the basis of similar findings clinically and radiologically. Common etiological factors affecting neurodevelopment and structural abnormalities may underlie the presentation of the two syndromes: schizophrenia and Davidoff-Dyke-Masson Syndrome. The other possible cause of the psychotic symptoms could be due to recurrent seizures as epilepsy has a significant association with psychosis.

The case is the first description of a patient with right-sided hemiatrophy and psychosis; the other cases described in literature had left-sided hemiatrophy (4-7). Cerebral asymmetry has been variably associated with schizophrenia, with both left-sided asymmetry and reversal of normal asymmetry described in literature.

Conclusions

Right-sided hemiatrophy may be an addition to the list of neurodevelopmental and structural cerebral anomalies associated with psychotic disorders including schizophrenia.

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