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Dyke-Davidoff-Masson Syndrome: A Case Report.

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Abstract
Background*: Dyke-Davidoff-Masson syndrome is a rare condition of unknown frequency resulting from brain injury due to a multitude of causes; especially in early life. Characteristics include cerebral hemiatrophy/ hypoplasia, contralateral hemiparesis, seizures, and compensatory osseous hypertrophy. Due to Its rarity , it may be misdiagnosed or under-reported by majority of clinicians.[1-3].*
Case presentation: *We present a case of 8 yr old girl who presented to us with breakthrough seizures*
Conclusions: *Due to its rarity, Dyke-Davidoff-Masson syndrome may easily be missed by the majority of treating clinicians. Knowledge of its features on imaging enables timely and accurate diagnosis – allowing appropriate management.*
Keywords: *Dyke-Davidoff-Masson syndrome, Computed tomography (CT)*

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I. Case Report

We present a case of a 8 years old girl who presented to us with breakthrough seizures. Patient had a history of seizures since 5 years of age which were focal in origin and limited to left side of body lasting for about 5 secs. Mother gave a history that baby was unresponsive at the time of birth and was kept in neonatal intensive care unit for 4 days , probably due to birth hypoxia which baby suffered at the time of birth. At 2 months of age parents started noticing abnormal enlargement of head and complains of 2 episodes of vomiting. CT brain was done which revealed dilated bilateral lateral and third ventricle due to aqueductal stenosis (obstructive hydrocephalus), for which patient underwent ventriculo-peritoneal shunting. As she grew her mother noticed that she could not use her left upper limb for carrying out any activity and holds her left shoulder internally rotated and adducted with elbow flexed and pronated. She drags her left foot while walking occasionally. On examination higher mental functions appear to be normal, Power is 4/5 on left upper and lower limb and 5/5 in right upper and lower limb and there is spasticity in left upper limb and lower limb. Deep Tendon Reflexes are exaggerated on left side and normal on right side and plantars extensor on left foot, all features pointing towards cerebral palsy with left hemiparesis. At the age of 5 she was initially started on Tablet Oxcarbazepine and Tablet Clobazam but still has breakthrough conulsions so gradually she was shifted on Tablet Levetiracetam 250 mg BD and Tablet Clobazam 5mg BD. Seizures have been controlled on these medications since 2 months and patient is clinically and symptomatically better.

Investigations

All basic blood reports were normal with hemoglobin 13.2 gm/dl, TLC – 6,600/cumm; platelet count – 2.8 x 10^6; serum creatinine 1.2 mg/dl; Na- 144mEq/L; K-4.1mEq/L; all liver enzymes were normal. Fundus

examination showed no significant findings; ECG and Chest x-ray were normal; patient was negative for HIV, Hepatitis B, Hepatitis C. Urine examination also showed no significant findings.

CT Brain Findings:

Chronic cystic gliotic changes along periventricular regions of right lateral ventricle extending into fronto-parieto-temporal and occipital lobes with significant white matter volume loss and thinning of overlying cortices. Few areas of calcifications are seen. Gliotic changes are also seen involving right temporal lobe cortex subcortical white matter. Right hemicerebral atropy is seen with mild compensatory left cerebral hypertrophy. Ventriculo-peritoneal shunt is seen reaching till right temporal region and generalized severe hypoplasia of corpus callosum noted. There is right lateral ventriculomegaly. Scalloping of inner table of calverium. Widening of diploic space on right side. Changes indicating Dyke Davidoff Masson Syndrome.

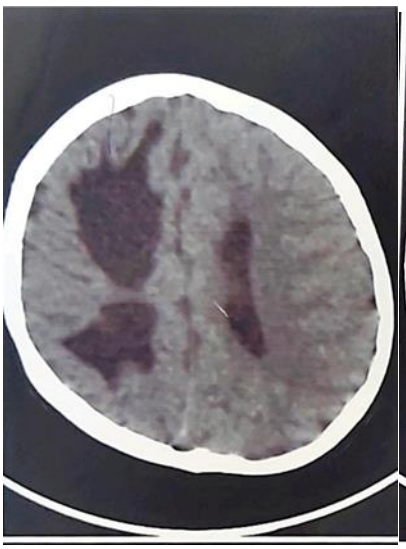

Figure 1

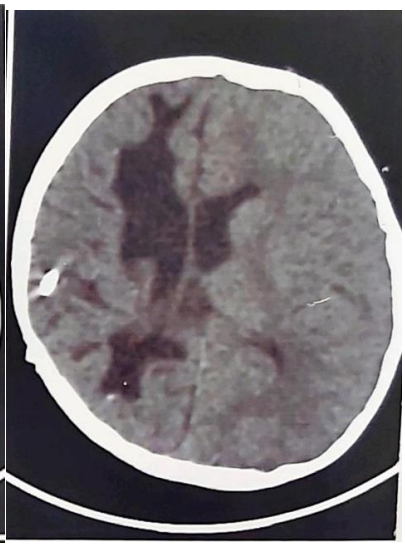

Figure 1

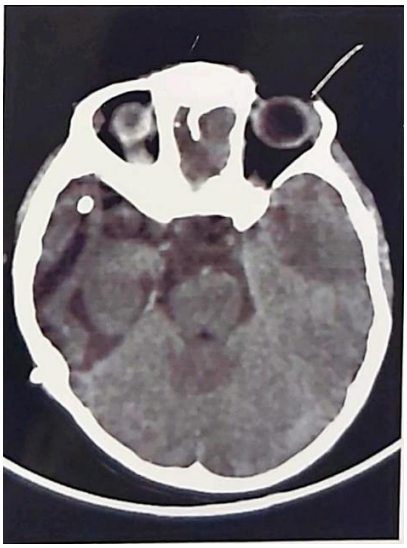

Figure 2

Figure 1 :CT Scan Axial Section shows Right Frontoparietal Hemiatrophy ; Figure 2: Axial Section at Basal Ganglia level- Shows V-P Shunt in situ with Right Cerebral Hemiatrophy. ; Figure 3- Axial Section at level of Pons- shows right temporal lobe atrophy.

II. Discussion And Conclusion.

In 1933, Dyke, Davidoff, and Masson described 9 patients with clinical characteristics of hemiparesis, facial asymmetry, seizures, and mental retardation noted to have pneumatoencephalographic changes on skull radiograph [4]. CT and MRI features of this entity include cerebral hemiatrophy, ipsilateral ventriculomegaly, hyperpneumatization of the sinuses on the affected side, and compensatory calvarial thickening [1, 3, 5]. Affected

patients are largely from the paediatric population; however, occurrence in adult patients have been reported [3]. Common causes include congenital anomalies, perinatal hypoxia, intracranial hemorrhage, and infections [1].

Clinically, patients may have seizures- focal or generalised often uncontrolled, mental retardation, contralateral hemiparesis, hemiatrophy, sensory disturbances, facial asymmetry , learning and speech impairment.[6-9] Some reports have also described presence of complex partial seizures with secondary generalisation. Th disease has been found to be more common in males and left hemisphere appears to be more frequently involved. Our patient a 8 year old girl child presented with seizures and left cerebral palsy and only after CT Brain imaging she was diagnosed. Understandably, the rarity of this condition makes accurate diagnosis a challenge – as evident in our experience.

Imaging via CT and MRI proves to be of great significance; enabling correct diagnosis and institution of appropriate management. These two imaging modalities are valuable in that they provide cross sectional images, with thin slices enabling visualisation of minute details. Pertinent imaging features for DDMS include cerebral hemiatrophy/ hypoplasia, hyperpneumatization of the paranasal sinuses, and compensatory osseous hypertrophy. The disease presentations are a result of cerebral insult during gestation or early in life, and hence can be divided into 2 types, the congenital (infantile) and acquired type. In the congenital type, structural abnormalities of the cerebral vasculature seems to be the cause of this leading to cerebral injury during fetal life.[2,6,8.]. It could also be due to some genetic defects[10]. The acquired type of disease arises due to CNS injury during the perinatal period or later secondary to causes such as infections, injuries, tumors, vascular anomalies,prolonged febrile seizures, ischemia, hypoxia, and various kinds of intracranial bleeds. If the disease develops within the first 2 years of life, compensatory changes, such as homolateral skull and sinus hypertrophy can result may occupy the vacuum left by the cerebral hypoplasia. The concerned side has a wide sulci replaced by gliotic cerebral tissue. In the congenital type, there is a midline shift towards the diseased side, and the sulcal prominence replacing the gliotic tissue is not present. This is what differentiates the 2 types from each other[10,11]. In essence, due to the rarity of this syndrome, it may be easily misdiagnosed by the untrained eye. CT and MRI are powerful imaging modalities to diagnose the pertinent imaging features associated with this syndrome. Due to the refractory nature of the seizures, the treatment revolves around the use of one or more appropriate antiseizure Medications. In the longer term, the patient needs to be managed with physiotherapy, speech therapy, and occupational therapy in addition to the medical treatment[9]. For intractable seizures and hemiplegia, the management of choice is hemispherectomy[9]. However, due to unavailability of hemispherectomy in many settings, the optimal way of managing the disease includes early diagnosis by appropriate imaging study, molecular genetics, medical management of seizures, and physiotherapy[12].Knowledge of the clinical presentation, risk factors, and imaging features is therefore indispensable for appropriate patient management.

References

[1] Verma R, Sahu R. Dyke-Davidoff-Masson Syndrome. Bmj Case Reports. 2012. Https://Doi.Org/10.1136/Bcr-2012-006729.

[2] Piro E, Piccione M, Marrone G, Giuffre M, Corsello G. Dyke-Davidoff-Masson Syndrome: Case Report Of Fetal Unilateral Ventriculomegaly And Hypoplastic Left Middle Cerebral Artery. Ital J Pediatr. 2013;39:32.

[3] 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.

[4] Dyke Cg, Davidoff Lm, Masson Cb. Cerebral Hemiatrophy And Homolateral Hypertrophy Of The Skull And Sinuses. Surg Gynecol Obstet. 1933;57:588–600.

[5] Gokce E, Beyhan M, Sade R. Radiological Imaging Findings Of Dyke-Davidoff Masson Syndrome. Acta Neurol Belg. 2017;117(4):885–93.

[6] Shrestha B. Acquired Cerebral Hemiatrophy: Dyke-Davidoffmasson Syndrome - A Case Report. Turk Neurosurg 2013; 23: 117-121.

[7] Zilkha A. Ct Of Cerebral Hemiatrophy. Ajr Am J Roentgenol 1980; 135: 259-262.

[8] Shetty Ds, Lakhkar Bn, John Jr. Dyke-Davidoff-Masson Syndrome. Neurol India 2003; 51: 136. .

[9] Singh P, Saggar K, Ahluwalia A. Dyke-Davidoff-Masson Syndrome: Classical Imaging Findings. J Pediatr Neurosci 2010; 5: 124-125.

[10] Atalar Mh, Icagasioglu D, Tas F. Cerebral Hemiatrophy (Dykedavidoff-Masson Syndrome) In Childhood: Clinicoradiological Analysis Of 19 Cases. Pediatr Int 2007; 49: 70-75.

[11] Behera Mr, Patnaik S, Mohanty Ak. Dyke-Davidoff-Masson Syndrome. J Neurosci Rural Pract 2012; 3: 411-413

[12] Sultan T, Malik Ma, Khan Mm. Chromosomal Analysis In Epilepsy. J Coll Physicians Surg Pak 2007; 17: 309.