



DYKE DAVIDOFF MASSON

SYNDROME WITH ABDOMINAL

EPILEPSY-RARE PRESENTATION

CASE REPORT, Vol-3 No.2

Asian Journal of Medical Science, Volume-3(2012)

<http://nepjol.info/index.php/AJMS>

Shitanshu Srivastava, Rajat Pratap Singh, K.L.Srivastava. Department of Pediatrics, Era's Lucknow Medical College, Lucknow.

ABSTRACT

CORRESPONDENCE:

Dr. Shitanshu Srivastava,
Associate Professor,
Dept of Pediatrics,
ELMC Lko
2/175 ,Vijay Khand Gomtinagar
Lucknow.226010
Mobile -09839512152,
Email ID:
dr_shitanshu@yahoo.co.in

Dyke Davidoff Masson Syndrome (DDMS) with abdominal epilepsy, is a rare clinical condition. It is characterized by severe abdominal pain with seizures, facial asymmetry, contralateral hemiparesis, and mental retardation. Diagnosis is made clinically coupled with radiological features which include cerebral hemiatrophy with homolateral hypertrophy of the skull and sinuses along with abnormal EEG and response to antiepileptic drugs. Here we report a case of DDMS with abdominal epilepsy in a 10 year old male child who presented with recurrent episodes of severe abdominal pain followed by seizures, hemiparesis and mental retardation. CT and MRI showed hemiatrophy of right cerebral hemisphere.

KEYWORDS: Dyke Davidoff Masson syndrome, abdominal epilepsy, cerebral hemiatrophy, mental retardation

*“Abdominal epilepsy is
an uncommon syndrome
in children and its
association with DDMS
is rare”*

INTRODUCTION

Dyke Davidoff Masson syndrome (DDMS) is characterized by focal cerebral hemiatrophy with homolateral hypertrophy of the skull and sinuses – resulting in facial asymmetry in association with contra lateral hemiplegia/hemiparesis, seizures, mental retardation, difficulty and impairment of speech development.¹

The disease is classified into congenital (infantile) or acquired variety.² Diagnosis is based on clinical features and radiological imaging (computed tomography and magnetic resonance imaging).^{3,4}

Abdominal epilepsy is an uncommon syndrome which consists of undiagnosed abdominal pain with seizures, abnormal EEG and dramatic response to antiepileptic drugs.^{5,6}

As to our knowledge DDMS presenting with abdominal epilepsy is a rare association, more so in paediatric age group.

CASE REPORT

A 10yr old male child presented to paediatric OPD of our hospital with history of recurrent abdominal pain since 6yrs of age. To begin with he had attacks of pain 1-2 episodes every month, which increased in frequency, intensity and duration. During the paroxysms he became pale, cold, lethargic, he sometimes use to fall down or shout aloud and went off to sleep after these attacks.

For these problems his parents consulted a local medical practitioner where some oral medications for abdominal pain were started. Over a period of 4 yrs, parents noticed weakness of left upper and lower limb along with progressively deteriorating school performance. His previous symptoms also persisted with no evident relief. He was referred to our hospital in view of his persisting and progressive complaints.

He was neurodevelopmentally normal till 6 yrs of age. After that parents noticed abnormal behaviour with deteriorating school performance along with

along with weakness in left upper limb and left sided limp. He was not able to perform his daily chores satisfactorily.

He has five siblings and there is a history of febrile seizures in the youngest sister who was admitted in our hospital at 6months of age and a diagnosis of febrile seizure was made. Her scans were normal. Rest all siblings have no history of any similar or other medical illness.

The natal, antenatal, post natal period was uneventful. Baby was delivered by normal vaginal route weighing 2.6kg and was apparently healthy at birth.

On physical examination his height is 151cm (>97 percentile), weight is 34kg (>50th percentile) and head circumference is 47cm (microcephaly <3rd centile for age), BMI is 14.91(50th percentile for age). There was no gross anomaly seen on general examination. He had left sided hypertonia with same sided extensor plantars and left sided power of 4/5 (evidence of unilateral pyramidal signs). All cranial nerves were intact. His sensory system was normal. He had a limping gait with restricted left arm swing. On abdominal examination no abnormality was detected. His I.Q. assessment score was between 55-70 which stated moderate mental retardation with mental age of 4-5yrs.

He was investigated for abdominal pain, his X-ray (abdomen erect PA view), USG abdomen, endoscopy, barium study and blood tests (S. amylase, S. lipase, LFT, RFT, electrolytes) were within normal limits. His CT scan head revealed small volume of right cerebral hemisphere with dilated cortical sulci and right lateral ventricle and Colpocephaly was noted in right frontal and occipital horn. Right calvarium was slightly thickened as compared to left. The features were suggestive of DDMS. MRI Brain showed right hemiatrophy with large area of gliosis in temporal

parietal occipital lobes with sign of volume loss.



Fig 1: CT scan head which revealed small volume of right cerebral hemisphere with dilated cortical Sulci and right lateral ventricle sign of volume loss



Fig 2: MRI Brain (T2 axial flair sequence) showed right hemi atrophy with large area of gliosis in temporo parietal occipital lobes with sign of volume loss.

EEG showed background rhythm with mixed 7Hz and fast beta activity. There was a discharge of poly-

-spikes and waves. The child was put on carbamazepine@10mg/kg/day. Paroxysms of abdominal pain decreased in frequency and intensity but still persisted. In view of persisting symptoms, dose of carbamazepine was increased upto 30mg/kg/day. It resulted in symptomatic relief of patient on follow up. Hence based on clinical features (mental retardation, hemiparesis), radiological scans, no other pathology for abdominal pain, abnormal EEG and response to Antiepileptic drugs a diagnosis of Dyke Davidoff Masson Syndrome as a cause of abdominal epilepsy was made.

DISCUSSION

Dyke, Davidoff, and Masson described this syndrome with plain skull radiographic and pneumatoencephalographic changes in a series of nine patients [1]. It is characterized by varying degree of facial asymmetry, seizures, contralateral hemiparesis, mental retardation and learning disabilities with behavioural abnormalities. [7] These findings vary depending on the extent of brain damage. Mental retardation is not always present, Seizures may appear months or years after the onset of hemiparesis [8].

Abdominal epilepsy is characterized by 1. Unexplained paroxysmal gastrointestinal complaints.

2. Symptoms of central nervous system disturbances.

3. An abnormal electroencephalogram with findings specific for seizure disorder.

4. Improvement with anticonvulsant medication. [6]

Abdominal epileptic pain is perceived as a severe and sharp sensation (—like a knife, mostly in a periumbilical localization, and was of variable duration [9]. During an episode of pain, disorientation followed by exhaustion and sleep is suggestive of abdominal epilepsy. Treatment typic-

-ally begins with anticonvulsant medication and resolution of symptoms with therapy helps to confirm the diagnosis

Hence in our patient, abdominal pain was stabbing in nature and followed by period of discomfort, There was progressive left sided weakness and mental subnormality in our patient. Abnormal EEG and response to antiepileptic drugs along with triad of hemiparesis, mental retardation and seizures clinically and CT/MRI findings radiologically helped us to reach to a diagnosis of Davidoff dyke Masson syndrome in association with abdominal epilepsy.

REFERENCES

1. Dyke C G, Davidoff L M and Masson C B, Cerebral Hemiatrophy and homolateral hypertrophy of the skull and sinuses. *Surg Gynecol Obstet* 1933; 57: 588-600.
2. Pendse NA, Bapna P, Menghani V, Diwan A .Dyke-Davidoff-Masson Syndrome. *Indian JPaed.*2004; 71:943.
3. Kumar S, Lakshmaiah V, Karthik Naidu KC. A rare cause for seizures and mental retardation – Dyke Davidoff Masson Syndrome, *Int J Biol Med Res.* 2011; 2(4): 1186 -1188.
4. Sener RN, JinkinsJR.MR of craniocerebral hemiatrophy.*Clin Imaging.*2002; 26(1):13-17.
5. NoahTZinkin, MarkPeppercor.Abdominal epilepsy .*Clinical gastroenterology*2005; 19:263-274.
6. Renata C. Franzon, Camila F. Lopes, Kátia M.R. Schmutzler, Maria Isabel, R. Morais, Marilisa M. Guerreiro. Recurrent abdominal pain-When an epileptic seizure should be suspected? *Arq Neuropsiquiatr* 2002; 60(3-A):628-630.
7. Narain NP, Kumar R, Narain B.Dyke-Davidoff- Masson Syndrome .*Indian Pediatrics.*2008;45:927-928.
8. OnoK, Komai k, IkedaT.Dyke-Davidoff-Masson syndrome manifested by seizure in late childhood: a case report.*JClin Neurosci.*2003; 10:367-71.
9. Daniel Eschle, Adrian Siegel, Heinz-Gregor Wieser.Epilepsy with severe abdominal Pain.*Mayo C Lin Prac* 2002;77; 1358-1360