



## Dyke-Davidoff-Masson syndrome with Coarctation of Aorta: Case Report of A Novel Association

Authors

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### Introduction

Dyke-Davidoff-Masson syndrome (DDMS) refers to atrophy or hypoplasia of one cerebral hemisphere (hemiatrophy), which is usually due to an insult to the developing brain in fetal or early childhood period<sup>1</sup>. DDMS is described as skull radiographic and pneumatoencephalographic changes in their series of nine patients whose clinical characteristics included hemiparesis, seizures, facial asymmetry, and mental retardation which was proposed in 1933 by Dyke et al<sup>2</sup>. The clinical findings may be of variable degree according to the extent of the brain injury. The following is a case presentation of a young male who presented with complaints of recurrent seizures and subsequently diagnosed as having coarctation of aorta (CoA). Neuroimaging findings were suggestive of DDMS. The present case is still remaining an enigma to us and no previous documentation of hemicerebral atrophy in the context CoA has been found in the literature.

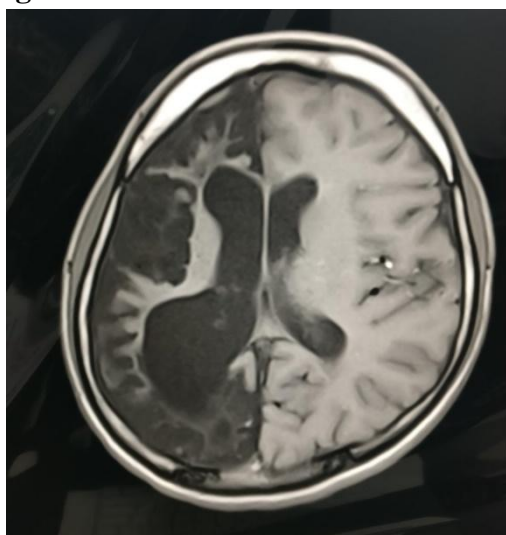
### Case Presentation

A 16-year-old boy was referred for management of recurrent seizures and left hemiparesis. He was born full-term, after a normal delivery in the hospital. At 2 years of age, he developed paralysis of left upper and lower limbs. He subsequently had delayed motor and speech milestones and fared poorly in scholastic activities. He had history of persistent headache and calf pain. On examination, there was hypertension in upper limbs (190/100 mm Hg) and lower limb blood pressure were not recordable. His lower limb pulses were also not felt. On examination, he had microcephaly, relative atrophy of the left half of the body, high arched palate, left-sided torticollis and thoracic scoliosis with convexity toward right (Figure 1). Neurological examination revealed spastic weakness of left upper and lower limbs with a spastic deformity of the left upper limb and a spastic left hemiplegic gait. His magnetic resonance imaging (MRI) showed right cerebral hemisphere atrophy with multiple subcortical cystic dilatation and ipsilateral exvacuolization of

ventricles, ipsilateral basal ganglia atrophy and midline shift towards right side(Figure 2). There was hyperpneumatization of ipsilateral frontal sinus and calvarial vault thickening. His magnetic resonance angiography was suggestive of coarctation of aorta (Figure 3). Patient was managed conservatively with control of seizures and hypertension.



**Figure 1** Torticollis and thoracic scoliosis



**Figure 2** Right cerebral hemisphere atrophy and right temporal sclerosis



**Figure 3.**Coarctation of aorta

Our patient, therefore, had hemiplegia, mental retardation and well-controlled generalized tonic clonic seizures and MRI findings characteristic of the rare syndrome known as DDMS with CoA not described in the previously published literature.

### Discussion

In 1933, Dyke, Davidoff and Masson first described the syndrome in plain radiographic and pneumoencephalographic changes in a series of nine patients<sup>2</sup>. It is characterized by asymmetrical growth of cerebral hemispheric with atrophy of one side and midline shift, ipsilateral osseous hypertrophy with hyper pneumatization of sinuses mainly frontal and mastoid air cells with contralateral paresis<sup>3</sup>. Clinical presentations include variable degree of facial asymmetry, seizures (focal ,complex partial, generalized), contralateral hemiparesis, mental retardation, learning disabilities, impaired speech, etc<sup>4</sup>.Both sexes and any of the hemisphere may be affected, but male gender and left side involvement are more common<sup>5</sup>.Two forms of the disease exist: a congenital form manifesting in the perinatal or infancy period, likely due to vascular occlusion in the gestational or neonatal phase and an acquired form, likely due to a later trauma, infection, ischaemia, or haemorrhage. The acquired form is likely to have a variable time period of presentation, appearing up to and into adolescence. Congenital type of DDMS, in contrast to acquired DDMS, shows shift of midline structures towards the side of the disease, enlargement of calvarium, diploic spaces and paranasal sinuses. The compensatory cranial changes occur to take up the relative vacuum created by the hypoplastic cerebrum<sup>6</sup>, as was found in our case.

Important differential diagnosis includes– Sturge–Weber Syndrome, basal cell germinoma, Fishman syndrome, Silver–Russell syndrome, linear nevus syndrome, and Rasmussen encephalitis. Rasmussen encephalitis does not show calvarial changes, and Sturge–Weber syndrome

additionally shows enhancing pialangiomas and cortical calcifications<sup>7</sup>.

Coarctation of the aorta also called aortic narrowing, is a congenital condition whereby the aorta is narrow, usually in the area where the ductus arteriosus inserts. The word "coarctation" means narrowing. Coarctations are most common in the aortic arch. Coarctation may be preductal, ductal or post ductal. Coarctation is about twice as common in boys as it is in girls. Symptoms may be absent with mild narrowings (coarctation). When present, they include breathing difficulties, poor appetite or trouble feeding, and failure to thrive. They may experience dizziness or shortness of breath, fainting episodes, chest pain, abnormal tiredness or fatigue, headaches, or nosebleeds. They have cold legs and feet or have pain in their legs with exercise. The classical finding in CoA is arterial hypertension in the arms with low blood pressure in the lower extremities and weak pulses in the lower extremities arteries<sup>8</sup>, as was found in our patient. Transthoracic echocardiography is the primary imaging modality for suspected CoA. However, cardiac magnetic resonance imaging is the preferred advanced imaging modality for non-invasive diagnosis and follow-up of CoA. Surgical resection is the treatment of choice in neonates, infants and young children. In older children (> 25 kg) and adults, transcatheter treatment is the treatment of choice<sup>9</sup>. To conclude, our patient has congenital form of DDMS with coarctation of aorta as a novel association.

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