Unusual association of diseases/symptoms

Dyke-Davidoff-Masson syndrome-like picture in a case of Takayasu arteritis: an enigma

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Summary

Authors describe the case of a 16-year-old girl who presented with fever, tonic-clonic seizures, unequal arm blood pressures and pulselessness in the left upper limb. On examination, there was a systolic bruit over umbilical region, a pansystolic murmur of mitral regurgitation was found. Neurological examination was normal except for an asymmetry in brain hemicircumference one side compared with the other. She has borderline intelligence (IQ 70) according to Wechsler Adult Performance Intelligence Scale. Magnetic resonance imaging (MRI) of brain revealed atrophic of left cerebral hemisphere with mildly ventricular dilatation, prominent paranasal and mastoid air cells, suggestive of Dyke-Davidoff-Masson syndrome (DDMS). Conventional angiography showed narrowed left internal carotid artery. There was also stenosed brachial artery, absent left renal artery with narrowed infrarenal abdominal aorta. The patient was put on antihypertensive drugs. We hypothesise that Takayasu arteritis and related vascular occlusion is the cause of her acquired cerebral changes.

BACKGROUND

Hemiatrophy of cerebral hemisphere is not frequently encountered in clinical practice. Dyke-Davidoff-Masson syndrome (DDMS) is a rare disorder involving hemiatrophy of one cerebral hemisphere secondary to an insult in the developing brain. The following is a case presentation of a young lady who presented to the emergency department with complaints of seizures and subsequently diagnosed as having Takayasu arteritis (TA). 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 TA has been found in the literature.

CASE PRESENTATION

A 16-year-old lady presented with a 3-day history of low-grade fever without any localising symptoms. On the third day of fever, she had three episodes of generalised tonic-clonic seizures. She recovered well following admission. She had a normal birth and developmental history. She denied any history of seizures, persistent headache, visual disturbance or vomiting or joint pain. On examination, there was sinus tachycardia, brachial and femoral pulses were absent on the left side. All other pulses were well palpable. There was no radiofemoral delay. Blood pressure (BP; supine) in all four limbs was as follows: right upper limb 150/100 mm Hg, left upper limb—not recordable, right lower limb—180/120 mm Hg and left lower limb—180/110 mm Hg. On auscultation, a pansystolic murmur was heard over the mitral area with S₃ gallop. Higher functions were essentially normal. In Wechsler Adult Performance Intelligence scale IQ was 70, indicating borderline intelligence. Head circumference was 48 cm, with hemicircumference on the right being 25 cm and on the left being 23 cm, indicating asymmetry. Rest

of the neurological examinations was within the normal limit. A systolic bruit in the umbilical region was present.

INVESTIGATIONS

Investigations showed normal blood count and erythrocyte sedimentation rate (ESR). Biochemical profile revealed normal renal and liver functions, and routine urine analysis did not show any abnormalities. Electrocardiogram showed sinus tachycardia with features of left ventricular hypertrophy.

Chest radiograph was normal. Echocardiography showed mitral regurgitation. Doppler ultrasound detected diminished velocity in subclavian, axillary, brachial, radial and ulnar artery (velocity in left subclavian 57 cm/s compared to right subclavian 115 cm/s; left axillary velocity 24 cm/s, resistance index 6; compared to right axillary velocity 91 cm/s, resistance index 85; left brachial 15 cm/s, right brachial 88 cm/s) with no evidence of intraluminal obstruction or thickening, suggestive of proximal narrowing. Magnetic resonance imaging (MRI) of brain showed atrophic changes in the left cerebral hemisphere characterised by prominent cortical sulci and sylvian fissure with ipsilateral occipital horn dilation. In addition, ipsilateral frontal paranasal sinus appears enlarged (figure 1A). There was white matter signal abnormality in the right temporal and occipital lobes. The posterior right cerebral parenchyma also appears swollen (figure 1B). Gliotic changes (hyperintense in T2-weighted image) seen in both hemispheres predominantly more on the left side (figure 1C); the whole picture was suggestive of DDMS. CT angiography of brain was normal. Conventional angiography showed complete obstruction of left subclavian artery distal to the origin of vertebral artery (figure 2A) with development of several collaterals and patent distal two-third of brachial artery, stenosed left internal carotid artery (figure 2B) with normal left external carotid artery,

Figure 1 (A) Axial T1-weighted image: left cerebral hemiatrophy with ipsilateral occipital horn dilatation. In addition, ipsilateral pneumosinus dilatans (frontal) is seen; (B) axial T2-weighted fluid-attenuated inversion recovery MRI demonstrating signal changes in right temporal and occipital cortex with oedematous posterior right cerebral parenchyma; (C) axial T2-weighted image: hyperintensity in white matter represents gliotic changes (more on left side). MRI, magnetic resonance imaging.

Figure 2 (A) Conventional angiography showed complete obstruction of left subclavian artery distal to the origin of vertebral artery with development of several collaterals; (B) stenosis of left internal carotid artery (arrow) with normal left external carotid artery; (C) absent left renal artery and with narrowing of infrarenal abdominal aorta.

absent left renal artery and with almost 50% narrowing of infrarenal abdominal aorta (figure 2C) of 7 cm in length.

DIFFERENTIAL DIAGNOSIS

The differential diagnosis for a cerebral hemiatrophy, the following entities to be kept in mind:

- ▶ TA induced chronic brain damage
- ▶ Acquired ischaemic disease
- ▶ Rasmussen's encephalitis
- ▶ Sturge-Weber syndrome
- ▶ Silver's syndrome

Acquired cerebral changes are more in favour of a vascular insult or viral infection like in Rasmussen's encephalitis.

TREATMENT

The patient was put on antihypertensive drug amlodipine and is now asymptomatic. Anticonvulsant was continued to prevent the recurrence of seizure.

OUTCOME AND FOLLOW-UP

Acute phase reactants (ESR and C reactive protein) were normal. No worsening vascular or constitutional symptoms and no newly developing vascular lesion was observed on follow-up angiography after 1 month. The patient was likely in the 'burnt-out' phase of the disease. The patient was put on antihypertensive therapy and is

now asymptomatic. No further episode of seizure occurred till now.

DISCUSSION

TA is a chronic, idiopathic, inflammatory disease that primarily affects large vessels, such as the aorta and its main branches.¹ The most common sites of lesion in TA are the aorta (65%) and the left subclavian arteries (93%). Carotid renal and vertebral arteries are also commonly affected.¹ The majority of the patients (53%) have lesions above and below the diaphragm.² In our case left subclavian, left internal carotid, left renal, and infrarenal aortic stenosis was found, so lesions were in both sides of the diaphragm. The patient presented with seizure, hypertension and unequal arm BP.

The common neurological manifestations include headache, visual disturbances, syncope, transient ischaemic attack, cerebral infarction, cerebral haemorrhage, hypertensive encephalopathy, seizures and paraplegia.^{3 4} Seizures as presenting symptoms are rare in patients with TA as in the present case. Borderline mental retardation and facial asymmetry were also present in our case.

As the natural history of TA is quite variable, clinical diagnosis of TA is always a challenge to the clinicians. The present case fulfilled five out of six criteria proposed by American College of Rheumatology for diagnosis of

TA. The presence of three or more criteria is 91% sensitive and 98% specific for the diagnosis of TA.⁵ Neurological manifestations of TA result either from decreased blood flow due to a steno-occlusive lesion and/or shunting of blood flow, thromboembolism or hypertension.

DDMS is characterised clinically by variable degrees of facial asymmetry, seizures, contralateral hemiparesis, mental retardation and learning, originally described on the basis of radiographic and pneumatoencephalographic changes in a series of nine patients in 1933.⁶ DDMS refers to the condition where cerebral hemiatrophy occurs due to an insult to the developing brain in the fetal or early childhood period. The classical clinical presentation includes seizures, facial asymmetry, contralateral hemiplegia or hemiparesis and mental retardation.⁷ 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 latter 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.⁸⁻⁹ In our case, cerebral hemiatrophy on the left side with prominence of the cortical sulci. The atrophied hemisphere will have prominent sulcal spaces if the vascular insult occurs after birth.¹⁰ Bilateral gliotic changes predominantly on the left were probably reflecting chronic ischaemic brain damage. The white matter signal abnormality in the right temporal and occipital lobes is likely to be caused by a hypertension-induced posterior reversible encephalopathy. This is also the most plausible aetiology of seizure in this case.

Postnatal hemiatrophy can develop secondary to cerebral trauma, tumours, infections and ischaemia. A possible aetiological relation of cerebral hemiatrophy with febrile seizures has been reported by Garg *et al.*¹¹ In another case report authors have related cerebral malaria and related vascular occlusion as the causes of this pathological changes.¹²

To conclude, the imaging findings are in favour of an acquired insult and TA-related occlusion left internal carotid artery probably lead to the cerebral changes.

Learning points

- ▶ Takayasu arteritis (TA) may cause vasculitic narrowing of major extracranial artery, gradually leading to hemiatrophy of brain and compensatory changes. The scenario may mimic a pattern of Dyke–Davidoff–Masson syndrome (DDMS).
- ▶ Chronic ischaemic insult to brain should be considered as a causative factor of cerebral hemiatrophy and related features. MR findings will help to differentiate adult-type DDMS from its counterpart.
- ▶ Accelerated hypertension as a complication of TA may lead to reversible encephalopathy, of which seizures are a common manifestation.
- ▶ Childhood onset TA may present with facial asymmetry and mental retardation and per se may cause seizure.

Competing interests None.

Patient consent Obtained.

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