

## CASE REPORT

# Recurrent facial hemiparesis due to dolichoectatic vertebrobasilar artery: an unusual and ignored cause

Vijay Nath Mishra,<sup>1</sup> Rameshwar Nath Chaurasia,<sup>1</sup> Shailesh Gupta,<sup>2</sup> Deepika Joshi<sup>1</sup>

<sup>1</sup>Department of Neurology, Institute of Medical Sciences, Banaras Hindu University, Varanasi, Uttar Pradesh, India

<sup>2</sup>Department of Physiology, Institute of Medical Sciences, Banaras Hindu University, Varanasi, Uttar Pradesh, India

**Correspondence to**  
Dr Rameshwar Nath Chaurasia,  
goformashwar@gmail.com

## SUMMARY

Dolichoectatic arteries are elongated tortuous aneurysms of intracranial arteries most commonly of vertebrobasilar tree presenting with ischaemic, haemorrhagic, thromboembolic lesions or with cranial nerve compression. The clinical presentation includes tic douloureux, neuralgia, tinnitus, vertigo, motor or sensory deficits, ataxia, dementia, Parkinsonism, hydrocephalus, headache, migraine, aneurysm, neoplasm, stroke/transient ischaemic attacks, leukoencephalopathy, central sleep apnoea and cerebellar dysfunctions. We present a case of recurrent facial nerve palsy secondary to vertebrobasilar dolichoectasia, an interesting and rare condition.

## BACKGROUND

The diagnostic criterion for vertebrobasilar dolichoectasia (VBD) is a basilar artery or vertebral artery diameter of  $>4.5$  mm or deviation of any portion of them higher than 10 mm from the shortest expected course, or basilar length  $>29.5$  mm or intracranial vertebral artery length  $>23.5$  mm.<sup>1</sup>

The pathophysiology in VBD ranges from vascular aneurysms, stenosis, occlusion, thrombo-embolism and turbulent blood flow to nerve palsies of trigeminal, abducent, auditory, ocular motor, trochlear or facial. Facial involvement especially with sporadic presentation has never been described previously.

## CASE PRESENTATION

We herein present the case of a 49-year-old man who presented with off and on clinical feature of facial hemiparesis in the neurology outpatient department of Institute of Medical Sciences, Banaras Hindu University. This patient had no history of diabetes or tobacco smoking but was a known chronic hypertensive. The patient complained of recurrent and alternating facial hemiparesis on either side since 1975 (table 1). The patient presented to us with facial heaviness of 6 days followed by deviation of angle of mouth to the right side along with drooling of saliva from left corner of mouth and sticking of food in the left side of the cheek for the next 3 days. Paresis was incomplete. There was no history of earache, ear discharge or any herpetic eruptions. There was no dysgeusia or auditory abnormality on history and detailed examination. The general physical and neurological examination yielded no other abnormality except left-sided lower motor neuron-type facial weakness.

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## INVESTIGATIONS

The patient was investigated and on MR angiography (MRA) a dolichoectatic vertebrobasilar artery as shown in MRI plates (figures 1 and 2) was established.

## TREATMENT

Patient was referred to the department of neurosurgery where he was operated for the condition. Clipping was done for anomaly.

## OUTCOME AND FOLLOW-UP

Patient is in follow-up and with improvement in weakness up to 95% in 5 month duration.

## DISCUSSION

Dolichoectasia is a rare entity and, its presentation is often mistaken for other neurological abnormalities. The incidence of dolichoectatic vascular events is very minuscule; around 0.06–5.8%.<sup>2–4</sup> Various clinical manifestations of a dolichoectatic artery as observed in the literature and past experience had involved cranial nerves and various related neural and non-neural structures. A very common occurrence has been cranial nerve compressive manifestations of trochlear,<sup>5</sup> trigeminal,<sup>6</sup> abducens nerves,<sup>7</sup> facial hemispasm<sup>8–9</sup> and the vestibulo-cochlear<sup>10</sup> nerves. Compression of the optic tract has been described with visual loss and nystagmus.<sup>11–12</sup> Vestibular hypo function has been reported along with the sensorineural hearing loss.<sup>13</sup>

Various other manifestations came in the form of tic douloureux, trigeminal neuralgia,<sup>6</sup> tinnitus and vertigo, motor or sensory deficits, ataxia, dementia and also parkinsonism. Obstructive hydrocephalus is also described<sup>14</sup> and so is dysarthria.<sup>15</sup>

Vascular changes might present in various forms—saccular fusiform aneurysms, dissecting aneurysm, giant serpentine aneurysm, lacunar infarcts,<sup>1</sup> subarachnoid haemorrhage,<sup>16</sup> haemorrhage in posterior circulation confirmed on imaging.<sup>1</sup>

Associated symptoms or conditions observed are—exertional headache,<sup>17</sup> basilar-type migraine,<sup>18</sup> aneurysm,<sup>19–20</sup> neoplasm, anterior circulation stroke/transient ischaemic attacks, hypertension, diabetes mellitus, hyperlipidaemia, coronary artery disease, gait imbalance and urinary incontinence.<sup>14</sup>

The distorted morphology encroaches upon the closed surrounding structures and the clinical manifestations are influenced by the involvement of particular blood vessel, related branches and the associated supply area; compression or fluid impact over the inflicted root or nerves or it could be due

**Table 1** Details of the facial weakness observed in the patient since 1975

Serial number	Location of facial paralysis	Year of paralysis	Preceding illness	Duration of paralysis (months)	Treatment received	Residual weakness (on total subjective perception of the patient) (%)	Neuroimaging
1	Right facial	1975	None	3–4	Galvanic stimulation, multivitamins	5	Not performed
2	Right facial	1978	None	2–3	Galvanic stimulation, multivitamins	10	Not performed
3	Left facial	1982	None	2	Steroids, physiotherapy, electrical stimulation	10	Not performed
4	Right facial	1984	None	2	Steroids, physiotherapy, electrical stimulation	10	Not done
5	Right facial	1989	Vascular headache	1.5	Steroids, multivitamins	15	Not performed
6	Right facial	1991	Rhinitis, urinary tract infection	6	Steroids, multivitamins, antibiotics	15	Head CT scan – normal
7	Right facial	2009	None	8	Steroids, multivitamins, acyclovir	20	Head CT scan – normal
8	Left facial	August 2012	Non-specific headache	Improved in 5 months	Steroids, physiotherapy and referred for surgery	95	Brain MRI with angiography

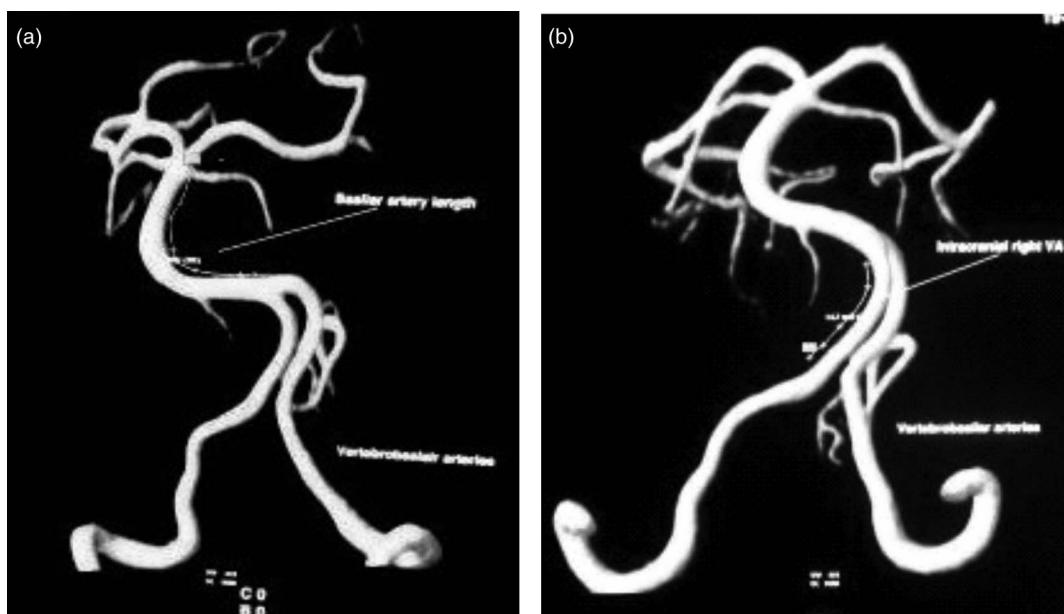
to the haemorrhage, thrombo-embolic or ischaemic pathology in the concerned region of the brain.

The pathophysiology has not been established completely. The dilatation, convolution and displacement of these highly stressed vasculature likely stems from the specific vascular structure pathology such as disrupted internal elastic lamina that may lead to activation of metalloproteinases and turbulent blood flow. A recent study describes infiltration of IgG4-containing plasma cells in the adventitia and media of the vertebra-basilar system leading into multiple fibrous nodules forming pseudotumours which become evident on the outer surface of the affected arteries.<sup>21</sup> The lesions might be an outcome of traditional cardiovascular diseases<sup>20</sup> or newer disease course much of which needs to be elucidated. Other associated

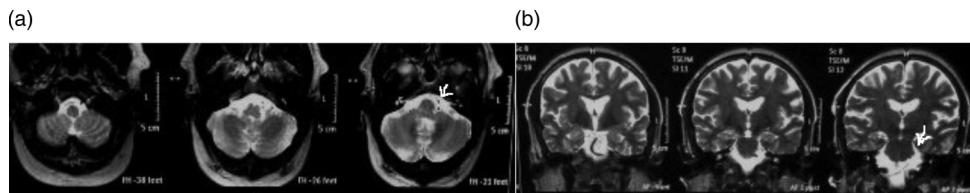
neurophysiological conditions or patient factors like smoking or work status; or any other pathological entity that creates this distortion needs to be explored further.

Traditionally, the diagnosis of VBD has been based on findings at catheter angiography. With the advent of CT scan and MRI it can be diagnosed non-invasively.<sup>1 22</sup> MRI has the ability to display multiplanar vascular anatomy and its relation to posterior fossa structures, to delineate mural thrombi and to show dissections.<sup>5–7</sup>

MRI and MRA are the optimal imaging techniques for demonstrating the compression of neural structures. The radiological evaluation with MRI,<sup>1</sup> MRA or CT exclude gross parenchymal involvement while histopathology has established the abnormality lying in the related vessels; which includes lesions like



**Figure 1** (A and B) Basilar artery is elongated and measures 30.4 mm in entire length. The artery appears to be deviated from the midline to the left side, approximately 11 mm from the midline. It, however, shows normal calibre and measures 38 mm in maximum diameter. Both vertebral arteries are normal in course, contour and calibre. The right vertebral artery is larger than the left artery and measures approximately 15 mm in intracranial course.



**Figure 2** T2-weighted axial (A) and coronal (B) images showing compression of left 7th and 8th cranial nerves by the deviated course of basilar artery.

atheromatous plaques, disruption of the internal elastic membrane and fibrosis of the muscular wall, etc. The resultant is a diffuse deficiency of the muscular wall and the internal elastic membrane.<sup>17</sup> A study in two brothers implied a genetic predisposition involving defects in arterial wall extracellular matrix components, as collagen, elastin, proteoglycans, laminin, etc, thus making these vessels susceptible to dilation.<sup>15</sup> The various pathophysiological outcomes could present clinically what could be divided into—ischaemic, cranial nerve compression and the signs from mass effect.<sup>17</sup>

Some of the attributable factors are male sex, advanced age, smoking, cardiovascular pathology, endovascular disease, diabetes and hypertension.

**Complications due to VBD:** The various defects in association with dolichoectasia are arterial dissection, VBD-induced aneurysmal formation and rupture, brainstem compression due to vessel enlargement and tortuosity, recurrent thromboses due to turbulent blood flow, vessel stenosis or occlusion. Thrombus with local or distal microembolisation, transient hypotension in a maximally compensated and autoregulated posterior circulation with abnormal flow has been hypothesised. VBD promotes brain ischaemia by occluding small perforating vessels, reduction of anterograde flow in the dilated artery, distortion and stretching of the branches of the basilar artery (Duret's haemorrhages).<sup>19</sup>

Haemorrhage in brain stem is common<sup>19</sup> with dolichoectasia in middle-aged patients while progressive dementia resulting from multiple cerebral infarctions, hypertensive leukoencephalopathy and anterior cerebral artery are usually seen in teenagers. The condition is also associated with cerebellar dysfunctions as well as central sleep apnoea.<sup>1</sup> Many manifestations could be attributed to cervicomedullary junction compression.<sup>1</sup>

### Learning points

- The dolichoectasia is the cause of many neurovascular manifestations with very discrete and dramatic clinical presentation in the form of neuralgia, headache, visual and vestibular deficit, facial hemispasms/palsy and other cranial nerve palsies.
- Vertebrabasilar dolichoectasia possess propensities towards stroke, nerves and brain tissue compression; much of the debilitating clinical conditions could be prevented if this condition is anticipated in advance.
- MRI or magnetic resonance angiography is useful in clinching the diagnosis if not digital subtraction angiography.

**Competing interests** None.

**Patient consent** Obtained.

**Provenance and peer review** Not commissioned; externally peer reviewed.

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