Unusual presentation of more common disease/injury

Acute onset quadriplegia

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Correspondence to Professor Chandrasekharan Rajasekharan, drcrajasekharan@yahoo.com**Summary**

A 50-year-old man, with history of chronic alcohol intake was brought in a stuporous state to the emergency services having been found in that condition in his home the same day. Examination revealed the patient in an akinetic mute state with apparently normal cranial nerves, hypotonia and quadriplegia with bilateral extensor plantar reflex. CT scan and MRI of the brain revealed bilateral infarct parasagittally with normal Magnetic resonance venogram suggestive of bilateral anterior cerebral artery infarct. Follow-up magnetic resonance angiogram revealed an azygous anterior cerebral artery thus proving an infarct of unpaired anterior cerebral artery infarct as the cause for quadriplegia in this patient.

BACKGROUND

Unpaired anterior cerebral artery (ACA) infarcts resulting in quadriplegia mostly exist only as a theoretical cause for quadriplegia. Infarction of the ACA territory itself is rare accounts for only 0.3% to 4.4% of cerebral infarctions reported.^{1 2} Bilateral ACA infarction is even rarer. Twenty-seven cases of ACA territory infarction were reported among 1490 cases of cerebral infarction in the Lausanne Stroke Registry, however, there were only two cases of bilateral ACA territory infarction.³ Several morphological variations of the circle of Willis exist. Maurer *et al* reported that an anaplastic A1 segment was found in 0.3% to 2.0% of individuals.⁴ Here we report a case of quadriplegia due to ACA territory infarct of unpaired or azygous ACA.

CASE PRESENTATION

A 50-year-old small scale business man, who was consuming approximately about 375 ml bootlegged country made liquor daily, non-smoker, with no significant history of medical illnesses was found in his room by his wife on the floor in his vomit, incontinent with no response to calls. No history of preceeding chest pain, palpitation, headache or fever was forthcoming. He was brought to our centre on the same day evening and was admitted.

On examination, he was having a pulse-90 per min regular, blood pressure 110/70 mm Hg, temperature 99°F, respirations 16–18 per min. He was drowsy, easily arousable, not responding to calls or commands. Spontaneous eye opening and eye movement was preserved with normally reacting pupils. Vertical and horizontal dolls eye movements were elicitable. Rest of the cranial nerves was apparently normal though a complete testing could not be done. There was hypotonia of all four limbs with no movement of any of the limbs to spontaneous or noxious stimuli with bilateral extensor plantar response. There were no signs of neck trauma and neck movements were normal. The rest of the system examination was non-contributory.

INVESTIGATIONS

His routine blood examination including a peripheral smear and urine examinations were within normal limits. Toxicological analysis of the gastric aspirate revealed no benzodiazepines or tricyclic antidepressants. Renal function tests and serum electrolytes were normal. Liver function tests including prothrombin time, international normalised ratio, arterial ammonia and ultrasonography abdomen revealed no abnormalities. Antinuclear antibody, dsDNA, c-antineutrophil cytoplasmic antibody (ANCA), pANCA were negative. Titres of antophospholipid antibodies both IgG and IgM, protein C and protein S assay were normal. Echocardiogram and transesophageal echocardiogram were normal. Holter monitoring did not reveal any rhythm disturbance. Doppler study of the aortic arch, carotid and vertebral arteries revealed no abnormalities. Cerebrospinal fluid analysis was normal. Electroencephalogram examination showed normal awake recording. CT scan head showed hypo densities in bilateral parasagittal fronto parietal area suggestive of bilateral ACA infarct. MRI T2 weighted images brain showed hyperintense signal in bilateral ACA territory suggestive of infarct (figure 1). Magnetic resonance venogram detected no abnormality. Magnetic resonance angiogram revealed azygous ACA on the right side and hypoplastic left A1 segment; anterior communicating artery A2, A3 arising from left ACA (figure 2).

DIFFERENTIAL DIAGNOSIS

- ▶ Central pontine myelinolysis
- ▶ Wernicke's encephalopathy
- ▶ Acute demyelinating encephalomyelitis
- ▶ Non-convulsive status epilepticus
- ▶ Top of the basilar artery stroke
- ▶ Atlantoaxial dislocation
- ▶ Cervical epidural haematoma.
- ▶ Fracture upper cervical vertebra
- ▶ Spinal arteriovenous malformations
- ▶ Acute cervical spondylotic myelopathy
- ▶ Acute transverse myelitis.

Figure 1 MRI T2 weighted images showing hyper intensities in bilateral ACA territory (white arrows).

TREATMENT

The patient was started on 20% mannitol 100 milligrams every 6th hourly intravenously and oral glycerine which was continued for 7 days. Thiamine 100 milligrams was given intravenously daily for 3 days and 100 mg was continued daily. Aspirin 300 mg tablet per orally daily was given. Atorvastatin 10 mg per orally daily was continued. He was fed by nasogastric tube and was on continuous bladder drainage using Foley's catheter.

OUTCOME AND FOLLOW-UP

He was on a nasogastric tube for feeding and indwelling catheter and supportive measures. He did not show any improvement in his higher mental functions or motor status. On follow-up after 3 months, his condition remains static and is highly dependant on his care givers.

DISCUSSION

ACA territory accounts for only 0.3% to 4.4% of cerebral infarctions reported, bilateral ACA infarct being even rarer.¹ Manifestations essentially consist of bifrontal disease which include bilateral hemiparesis, bulbar palsy, abulia or akinetic mutism, bland affect, release reflexes and sphincter incontinence.⁵ Cardio embolic or artery to artery embolism is the major cause of ACA occlusion, vasospasm due to subarachnoid haemorrhage being another. Anomalies of ACA are considerable but three distinct patterns are well recognised: accessory ACA, bihemispheric ACA and unpaired or azygous ACA. The true incidence of bilateral ACA infarction is unknown, with few cases reported in the literature. In 2004, Yamaguchi *et al* reported a similar case with a patient presenting with lower limb weakness and magnetic resonance angiography demonstrating bilateral anaplastic

Figure 2 Magnetic resonance angiogram anterior view showing azygous ACA (unpaired anterior cerebral artery) and hypoplastic left ACA with absent A1 segment (white arrows).

anterior cerebral arteries. In the study by Kumral *et al* of the spectrum of ACA territory infarction in 48 consecutive patients over a 6 year period only two patients had bilateral infarction and they presented with akinetic mutism, severe sphincter dysfunction, and dependent functional outcome.⁶ A recent report by Menezes *et al* in which bilateral ACA infarct due to unpaired ACA initially presented as space occupying lesion.⁵ Ferbert and Thron also reported two cases of bilateral ACA infarct with vertical gaze palsy and concluded that bilateral ACA territory infarction should be considered in the differential diagnosis of basilar artery occlusion, even if accompanied by vertical gaze palsy.⁷

Learning points

- ▶ Anterior circulation stroke can cause quadriplegia similar to infarcts of posterior circulation as in top of the basilar stroke.
- ▶ Bilateral ACA infarction remains a rare but important cause of quadriplegia.
- ▶ Bilateral ischaemia or infarction can result from unilateral occlusion in patients with anomalies of Circle of Willis.
- ▶ An occlusion of an unpaired ACA where by the blood supply to medial and parasagittal area of both frontal and parietal lobe can result in akinetic mute state, quadriplegia or bilateral hemiparesis, sphincter dysfunction and poor functional outcome.

Competing interests None.

Patient consent Obtained.

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