THROMBOSIS OF THE POSTERIOR INFERIOR CEREBELLAR ARTERY OF LUETIC AETIOLOGY*

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The syndrome of occlusion of the posterior inferior cerebellar artery was first described by Wallenberg (1895). Since his first description, a number of reports and reviews on the subject have appeared in the literature and it appears that the condition is not uncommon (Goodhart and Davison, 1951; Levine, Cheskin, and Applebaum, 1949; Merritt and Finland, 1930; Ramsbottom and Stopford, 1924; Riley, 1930; Spiller, 1908; Thomas, 1907; Thompson, 1929; Wilson, 1909; Wilson and Winkelman, 1927).

Ramamurthi (1956) reported 22 cases of vascular occlusion of the posterior fossa, during a 5-year period (1950–1955). In this series the posterior inferior cerebellar artery was involved in twelve cases and appeared to suffer occlusion more frequently than any other blood vessel of the hind brain. But in comparison with the vascular accidents of the cerebrum, those of the hind brain may be said to be infrequent.

The posterior inferior cerebellar artery is the largest branch of the vertebral artery, arising like the medullary branches, nearly at right-angles to the main vessel. This anatomical arrangement may interfere with the free flow of blood and is probably a factor in the development of thrombosis in an already diseased artery. The posterior inferior cerebellar artery supplies the lateral portion of the medulla and the adjacent portion of the cerebellum. The area supplied by the artery includes the restiform body, the direct spino-cerebellar tract, the descending root of the trigeminal nerve, the motor nucleus of the vagus nerve (the nucleus ambiguus), the lateral spino-thalamic tract, the sympathetic centre, a portion of the hypoglossal nucleus and a portion of the cerebellum, Deiter’s nucleus, and occasionally the nucleus facialis.

These important centres and tracts in the lateral portion of the medulla become devoid of blood supply in thrombosis or occlusion of the posterior inferior cerebellar artery. The classic picture of the syndrome is unmistakable, but, depending upon the variations in the anatomic distribution of the vessel and the nature of the collateral circulation, some symptoms may be absent while others may be present.

Aetiology.—The lesion is stated to affect persons of the sixth and later decades, and the commonest aetiological factor is arteriosclerosis and hypertension (Riley, 1930; Merritt and Finland, 1930). Between the ages of 30 and 50 years, syphilis as an aetiological factor needs to be kept in mind. Only a few sporadic cases in which syphilis was the causative factor have been recorded. The earliest cited report of syphilitic thrombosis of the posterior inferior cerebellar artery which the authors could find was that of Salmon (1913). Kozevnikov (1927) and Wilson and Winkelman (1927) each reported one case of syphilitic thrombosis of the artery. Freidovitch (1929) reported the syndrome in a patient with syphilitic aortitis, who recovered with antisyphilitic therapy, so that it was presumed that the syphilis had caused the occlusion. Bianchi, Iribarren, and Querol (1942) cited by Levine, Cheskin, and Applebaum, 1949), reported a case in which autopsy examination revealed syphilitic arteritis of the left vertebral artery with thrombosis of the posterior inferior cerebellar branch.

Merritt, Adams, and Solomon (1946) recorded three cases of thrombosis of the posterior inferior cerebellar artery among the series of 42 cerebral vascular accidents of luetic aetiology observed at the Boston City Hospital. Among the 22 cases of vascular occlusion of the hind brain reported by Ramamurthi (1956), are listed five cases (22.7 per
cent.) as of luetic aetiology, but no details are given. Other rarer causes include trauma and tuberculosis (Ramamurthi, 1956), thromboangiitis obliterans (Hausner and Allen, 1938), metastatic neoplasm (Davison and Spiegel, 1945), and embolism from a rheumatic heart (Merritt and Finland, 1930).

Whatever the aetiology, the clinical picture of the syndrome is typical. The onset is abrupt with dizziness, vomiting, and falling towards one side but no loss of consciousness. Further signs and symptoms in accordance with the areas involved are listed in the Table.

<table>
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<tr>
<th>Areas of Involvement</th>
<th>Signs and Symptoms</th>
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<tr>
<td>Nucleus ambiguus</td>
<td>Difficulty in deglutition, contra-lateral deviation of uvula and palate, homo-lateral laryngeal paralysis.</td>
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<tr>
<td>Descending root of trigeminal nerve</td>
<td>Homo-lateral facial analgesia, thermo-anaesthesia</td>
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<tr>
<td>Lateral medullary sympathetic centre</td>
<td>Horner’s syndrome (miosis, enophthalmos, ptosis of the upper lid, and diminution of perspiration on face on side of lesion)</td>
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<td>Lateral spinothalamic tract</td>
<td>Contra-lateral anaesthesia of trunk and limbs, to pain and temperature</td>
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<tr>
<td>Direct spino-cerebellar tract or restiform body</td>
<td>Homo-lateral ataxia</td>
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<tr>
<td>Deiter’s nucleus</td>
<td>Nystagmus, dizziness, and falling towards side of lesion</td>
</tr>
<tr>
<td>Nucleus facialis</td>
<td>Homo-lateral facial paralysis</td>
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Diagnosis.—The symptom-complex is almost pathognomonic. The sudden onset of dizziness without loss of consciousness and inability to swallow indicate a vascular lesion of the medulla. Other conditions which are likely to complicate diagnosis are tumours of the medulla, syringobulbia, and the jugular foramen syndrome. The mode of onset, Horner’s syndrome, and involvement of the lateral spinothalamic tract and the descending root of the trigeminal nerve are sufficient to exclude other conditions.

Prognosis.—This is considered good whatever the aetiology. In syphilitic arteritis, dramatic recovery follows specific therapy. Most of the symptoms and signs disappear with time, but the sensory changes generally persist.

Case Report

A married Muslim male aged 35 years was admitted to the E.N.T. unit of the Government General Hospital complaining of inability to swallow of 3 weeks’ duration. During investigation, an attempt was made to feed him through a Ryle’s tube, but he was unable to retain the tube and was uncooperative. As he was becoming emaciated, a gastrostomy was performed by a surgical colleague and the patient was fed through the gastrostomy tube. The blood serological reaction being reported positive to syphilis, the patient was transferred to the Institute of Venereology on August 27, 1957.

Past History.—The patient admitted having had a penile sore and urethral discharge 12 years before and had had no specific treatment.

Present Illness.—Until about 6 weeks before, the patient had been in normal health. One night, when he got up to pass urine, he felt extremely dizzy and was not able to walk, with a tendency to fall towards the left. He was helped to bed and was given some water to drink, but he could not swallow and the attempt brought on a choking sensation with violent bouts of coughing and regurgitation through the nose. He was perfectly conscious, and complained of numbness of the left side of the face and difficulty in phonation. He was taken to a private medical practitioner who was alleged to have given him two intramuscular injections. As there was no improvement, he was admitted to hospital.

Physical Examination.—The patient was a poorly-nourished individual of asthenic habitus. The gait was rather unsteady because of a feeling of insecurity on the left side, but he was able to walk with a stick. The voice was husky. There was a scar on the anterior aspect of the glans penis. The regional lymph nodes were slightly palpable and discrete. A rubber tube was protruding through a gastrostomy wound in the epigastrium.

Neurological Examination:

Cranial Nerves:

1. Horner’s syndrome on the left side (narrowing of the palpebral aperture, enophthalmos, partial ptosis of the upper eye lid, miosis of the pupil, absence of perspiration on the left side of the face).
2. Loss of sensation to pain and temperature in the distribution of the left trigeminal nerve with impairment of the corneal reflex.
3. Voice husky, with paralysis of the left vocal cord, marked dysphagia, and deviation of the uvula to the right on phonation.
4. Pupils reacted to light and accommodation although the left pupil was miotic.
5. Horizontal nystagmus on lateral gaze to the left.
6. Slight deviation of the tongue to the right.

Motor System: Normal.

Sensory System: Impairment of sensation to pain and temperature of right arm, leg, and right of trunk. Light touch, vibratory, and position sense normal.

Reflexes: Superficial and deep reflexes and plantar response normal.

Laboratory Investigations:

Blood pressure: 108/78.
Total red blood count: 4.4 millions per cmm.
White blood count: 8.200 per cmm.
Differential count: polymorphs 70 per cent.; lymphocytes 26 per cent.; eosinophils 3 per cent.; monocytes 1 per cent.
Erythrocyte sedimentation rate: 8 mm. one hour.
Urine analysis: no abnormality.
Skagram of skull normal.
Cerebrospinal fluid: Clear and not under pressure; cells, 9 per cmm. all lymphocytes; total protein, 60 mg.; V.D.R.L. test positive 4 dilutions; Wassermann reaction positive.
Ophthalmoscopic examination normal.

Course.—The patient remained in hospital for 3 weeks and was given 8.8 mega units PAM at the rate of 600,000 units daily. After the first four injections of PAM he was able to swallow both liquids and solids, the gastrostomy tube was removed, and the wound healed rapidly. Normal voice was restored and he was able to walk without the aid of a stick. He insisted on going home though at the time of discharge the Horner’s syndrome and sensory disturbance were still present.

Diagnosis.—Wallenberg’s syndrome of luetic aetiology was diagnosed on the basis of the neurological and laboratory findings.

Summary
A case of syphilitic thrombosis of the posterior inferior cerebellar artery in a man aged 35 is reported, with a short discussion of the syndrome in general. Although the patient could not be kept under observation long enough to observe the final outcome, the initial good response to treatment during the period of hospitalization suggested that he would obtain complete recovery.

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REFERENCES