Vagal-Accessory-Hypoglossal Syndrome: Schmidt's or Jackson's?

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Key Words

Vagus nerve · Accessory nerve · Hypoglossal nerve · Traumatic nerve injuries · Medullary syndrome

Abstract

Paralysis of the vagal-accessory-hypoglossal nerves has many eponyms. Many cases were caused by injuries in the First World War. Confusion arose because of the varied anatomical sites and varied extent of tissues damaged. Hughlings Jackson's name is justified by historical precedence, but descriptive terms of axial and extra-axial lesions are preferred.

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A unilateral paralysis of the soft palate, vocal cord, sternomastoid and trapezius muscles has been named Schmidt's syndrome (one vocal cord and sternomastoid paralysis – 'vago-spinal') [1]. It is caused by lesions affecting the vagus, spinal accessory, and hypoglossal nerves or nuclei. Some arise from extra-axial, some from intramedullary diseases. Several minor variants with many alternative eponyms include: Collet's syndrome – from the shrapnell ball behind the right mastoid (glosso-laryngo-scapulopharyngeal paralysis) [2], Villaret's retroparotid syndrome ('hemiatrophy of the tongue, dysarthria, enophthalmos and miosis, paralysis of one vocal cord, hoarseness, paralysis of the soft palate and dysphagia') [3], Sicard's syndrome – revolver ball behind the left mastoid (the condylo-posterior lacerated foramen) [4], Anto-

nio Garcia Tapia's syndrome (extra-axial paralysis of the larynx and tongue with or without paralysis of the sternocleidomastoid and trapezius, sparing the soft palate) [5], and Avellis's syndrome (ipsilateral palato-laryngeal paresis and contralateral hemiparesis and/or hemihypoaesthesia) [6].

Hughlings Jackson (1835–1911) had much earlier reported the syndrome [7] in 1864, and described a further case caused by medullary haemorrhage in 1872 [8] with hypoglossal palsy and incomplete vagal paresis:

'The patient was a gentleman, fifty-one years of age [first seen in 1864]. Six years before when abroad had a "sunstroke". He got well of this in about 10 months; but at the end of this time had an attack of hemiplegia. His tongue was turned to one side, but was put out when required... Got well again and resumed his practice. But shortly after doing so, one day, i.e., in June 1861... he lost the power of articulation... very suddenly, and from that time to this (1864) he has not said a single word. He was, after he lost his articulation, insensible for a few hours.'

'1864. – He is universally weak, and staggers when he walks; but he has no local paralysis of the arm or leg. He has total paralysis and atrophy of the tongue on both sides. He cannot cough, and he has very great difficulty in swallowing. Dr. Morell Mackenzie's aid: "there did not appear to be any paralysis of the vocal cords. They were seen to flap together in an unsteady manner, and seemed rather disposed to remain apart than to become closely approximated... could produce a simple expiratory sound, such as ah, eh or oh. On inspiration, the cords re-

mained equally distant from the median line. There was a quantity of slimy mucus in the larynx.”

‘...After this, he gradually grew worse, became unable to walk and took to his bed. By September 1864 he had lost the use of both arms and legs... He died on September 26th.’

Dr. Lockhart Clarke’s examination gave the following results.

‘Upper surface of brain there was great venous congestion... Basilar artery almost choked by atheroma, consistency of a cord. Superior cerebellar artery was not much affected. Posterior cerebral was loaded with atheroma throughout its whole course... Internal carotid on the right before ophthalmic branch was loaded with the same kind of deposit. That on the left was not much affected. Both middle cerebral arteries throughout their course... extensive atheromatous degeneration... multiple cerebral softenings; small lesion of left corpus striatum; larger chocolate coloured mass, size of a brazil nut, and right corpus striatum and optic thalamus would account for left hemiplegia. Whole cerebellum was softer than natural... superior vermiform process and parts adjacent reduced almost to a pulp. Under surface of left lateral lobe a roundish space about half an inch in diameter was reduced to complete pulp and somewhat sunken. Left cerebellar haemorrhage... Corpus dentatum almost wholly obliterated and partially replaced by a small, round, and sharply defined cyst, size of a pea, containing thick, opaque milk-like fluid... Haemorrhagic lesions of the medulla oblongata, left olivary body... old apoplectic clot.’

‘The lesions, which give the case its greatest value, are those in the medulla oblongata. Why the tongue was paralysed on both sides from a lesion on but one side of the medulla oblongata, I do not understand... the sudden onset of paralysis of the tongue, and the significant posi-

tion of the haemorrhage, ... justify the conclusion that on the day the man lost his articulation haemorrhage occurred in the olivary body.’

Jackson reported a further example involving the vagus and hypoglossal nerve in 1886 [9]. The otorhinolaryngologist, Sir Stephen MacKenzie described two further instances on February 12, 1886; he acknowledged Jackson’s similar report of 1864. The first was a 30-year-old man with syphilis with paralysis of the left tongue, soft palate, vocal cord, sternomastoid and trapezius and Horner’s syndrome. The second patient, a nephritic, had partial paralysis of the left side of the tongue, palate and vocal cord and trapezius [10].

Other causes have been described, some extramedullary, some intramedullary lesions. These are secondary to: shotgun injuries, atlas and occipital condylar fractures, cranial polyneuropathy, basilar invagination, tumours of the skull base, multiple myeloma, vasculitis, carotid fibromuscular dysplasia, coiling and dissections of the internal carotid artery. Vascular lesions are based on a blood supply via paramedian bulbar branches supplied by the vertebral artery and the anterior spinal artery, and lateral bulbar branches supplied by the intracranial vertebral artery or the posterior inferior cerebellar artery. Because the dura over the intrajugular foraminal septum is perforated by the glossopharyngeal meatus for the glossopharyngeal nerve and by the vagal meatus for vagal and accessory nerves, jugular foramen lesions can produce a comparable picture.

The many eponyms arose mainly because of a variety of wartime injuries in the First World War. They cause confusion because of the varied anatomical sites and extent of tissues damaged. Jackson’s name is justified by historical precedence, but descriptive terms – such as vagal-accessory-hypoglossal palsy – are preferred.

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