SYNDROME OF AVELLIS

By C. C. CORBETT (Waterford, Eire)

PARALYSIS of the larynx occurring together with one or more of the last four cranial nerves has been classified into groups associated with personal names. Some observers (Lovesey, 1944) disagree with this classification as impractical and would favour a simplified nomenclature, i.e. an Anatomical Classification.

George Avellis, a German, first described the condition that bears his name in a series of ten cases published in 1891. Since then there have been only about 30 cases reported in the available literature (Imperatori; 1925; Croizier *et al*, 1912; Dejerine *et al*, 1912; New, 1923; Collet, 1896; di Lauro, 1930; and Pinson, 1936). The last available report of such a case was that of Font in 1952 and the two cases reported were probably not classical examples of this syndrome.

The Syndrome of Avellis, as described by Musgrove (1952) consists of paralysis of the soft palate and larynx on the same side and partial paralysis of the constrictors of the pharynx. There may be a contralateral loss of pain and temperature sense of one half of the body.

It is not the purpose of this paper to discuss the related syndromes to the one under review, but for classification, the following classifications are presented from Lederer (1946):

(1) *Syndromes of Associated Paralysis of the Larynx*

	IX (Taste)	X (Vocal Chord and Palate)		XI (Shoulder)	XII (Tongue)
(*a*) Collet Sicard	1	1	1	1	1
(*b*) Hughlings Jackson	—	1	1	1	1
(*c*) Vernet	1	1	1	1	1
(*d*) Schmidt	—	1	1	1	—
(*e*) Avellis	—	1	1	—	—
(*f*) Tapia	—	1	1	—	1
(*g*) Villaret	1	1	1	—	1

(2) *Anatomic Sites for Lesions carrying Associated Vocal Cord Paralysis*

Syndrome	Paralysis involving Nerves	Vagus Lesion
(*a*) Bulbar	IX, X, XI, XII (V, VI, VII, VIII)	High Vagus Paralysis
(*b*) Jugular Foramen	IX, X, XI, XII, Sympathetic	High Vagus Paralysis
(*c*) Parapharyngeal Space	IX, X, XII (VII) Sympathetic	Deep Vagus Paralysis
(*d*) Arch of Aorta	X, Phrenic, Sympathetic	Recurrent Laryngeal of Deep Vagus

In order to understand this syndrome more fully, a knowledge of the relevant anatomy is necessary. There follows a brief review of the structures involved:

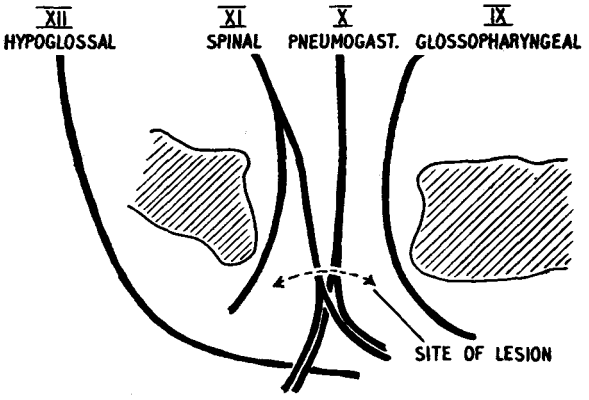
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Syndrome of Avellis

The vagus nerve on emerging from the jugular foramen is accompanied by and contained in the same sheath of dura-mater as the accessory nerve. They are separated by a fibrous septum from the glossopharyngeal nerve anteriorly. The accessory nerve on emerging from the jugular foramen separates into a cranial and a spinal portion. The vagus nerve, after its exit, is joined by the cranial part of the accessory nerve and enlarges into a ganglion (ganglion nodosum). The fibres of the cranial part of the accessory nerve pass from this ganglion uninterrupted. They are distributed to the pharyngeal and superior laryngeal branches of the vagus nerve. The branches of the pharyngeal portion of the vagus are joined by branches from the glossopharyngeal, sympathetic and external pharyngeal nerves to form the pharyngeal plexus. This plexus supplies the muscles of the pharynx and soft palate except the tensor palati which is supplied by a branch of the trigeminal nerve.

The external branch of the superior laryngeal branch of the vagus supplies the cricothyroid muscle and gives off branches to the pharyngeal plexus and the inferior constrictor of the pharynx (Gray, 1958).

From this description it can be appreciated that a lesion of the vagus nerve outside the jugular foramen can give rise to a syndrome, such as that described by Avellis. One symptom that may cause some confusion in the mind of the diagnostician is the dysphagia, as this may be the presenting symptom complained of by the patient and, as in the case of the author, may lead the examiner to suspect œsophageal neoplasm. This dysphagia is due to the partial paralysis of the pharyngeal constrictors and indicates a lesion of the vagus trunk above the origin of the pharyngeal branches of that nerve. The following sketch shows the probable site of the lesion.



(After Maurice Vennet)

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Ætiology

In view of the scarcity of reports and the resulting shortage of post-mortem confirmation, it is difficult to state the precise ætiology of this condition. The following conditions are those usually accepted:

(1) *Vascular*, e.g. *Aneurysm*
Rupture of a vessel
Thrombosis of bulb

Lovesey (1944) considered that his case was one of vascular origin.

(2) *Inflammatory*, e.g. *Mastoiditis*
Cervical adenitis
Cellulitis

Fox and West (1947) reported a case complicating acute mastoiditis.

(3) *Neoplastic*, e.g. *Primary* or *metastatic tumours*

(4) *Toxic*

(5) *Herpetic*

Font (1952) alleges that many of these cases are caused by unobserved herpetic phenomena.

Discussion

It is obvious from the literature that the causes of associated paralysis of the larynx are obscure. This is made more obvious by the lack of precise ætiological data in the available text books. However, it is now presumed that the Syndrome of Avellis is due to any lesion involving the nucleus or lower motor neurones.

Clinical Picture

The patient may present complaining of difficulty in swallowing, hoarseness or change in speech. On examination, the following are the usual findings: (*a*) Paralysis of the soft palate accompanied by deviation of the uvula to the unaffected side. Poli (1906) states that quite frequently the onset of soft palate paralysis occurs after that of the pharynx and larynx. This paralysis is accompanied by ipsilateral paralysis of the larynx accompanied by anæsthesia of the corresponding part of the palate and pyriform fossa. This is accompanied by partial paralysis of the constrictors of the pharynx. Lovesey (1944) draws attention to the fact that the X-ray findings are suggestive.

Prognosis

As regards life, this depends on the causative condition and as a rule the laryngeal paralysis appears to be permanent.

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Case Report

Miss X, an unmarried female, aged 53 years. First seen in May, 1958, complaining of difficulty in swallowing solids, and indistinct speech accompanied by feeling of "something" going down the back of her throat for two weeks. The patient's previous medical history was good and she was feeling quite well except for the fact that she could not eat normally.

On examination, her ears, nose, pharynx and post-nasal space were normal, but there was a paralysis of the right vocal cord which lay in the para-median position. A barium swallow was advised and the report was that a neoplasm of the upper part of the œsophagus was suggested. Wassermann reaction and chest X-ray were negative, blood pressure was 135/80 and the blood picture was normal.

Œsophagoscopy was carried out 10 days after the patient was first seen and no neoplasm was seen. However, in view of the radiological report, the patient was referred to the Radiotherapy Department in Dublin and there investigations were also negative. The patient was then sent home and was again seen by me. By this time, she was able to swallow much more comfortably though her speech was more indistinct than at the first examination. Examination on this occasion showed right vocal cord still in the same position, but also at this time it was seen that the right half of the soft palate was paralysed with deviation of the uvula to the left side on phonation. There was also diminished sensibility of the paralysed part of the soft palate and the corresponding pyriform fossa.

The patient has since been seen by me on four occasions and though she is now eating quite normally, her speech is still very difficult to understand and the clinical picture has not changed at all. On the last occasion on which she was seen, i.e. 6th, October 1958, though she was feeling quite well and apparently taking plenty of nourishing food, she had obviously lost weight since the time of the first examination. In view of this, I again admitted her to hospital and repeated œsophagoscopy which again revealed a perfectly normal œsophagus. One point which was mentioned to me by the staff nurse in charge of the ward, and which I verified myself, was that the patient's speech was much more distinct when she was lying down than it was in the erect position. Unfortunately, due to the non-co-operation of the patient, it has been impossible to have a full neurological examination. However, this case is presented as a typical Syndrome of Avellis.

Summary

(1) A review of the Syndrome of Avellis with a special reference to the ætiology.

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(2) Case presented of a middle-aged female complaining of sudden dysphagia and alteration of voice.

(3) Attention is drawn to the onset of the soft palate paralysis after that of the pharynx and larynx.

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11 Catherine Street,
Waterford, Eire

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