Vestibular neuronitis: a review of a common cause of vertigo in general practice

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SUMMARY. Vestibular neuronitis is an interesting condition characterized by the acute onset of vertigo, nausea and vomiting, in the absence of hearing loss or tinnitus. There is often evidence of a recent or concurrent upper respiratory tract infection. The disease follows a benign course of between two days and six weeks. It often occurs in epidemics. Following the acute attack, mild transitory episodes of dizziness may recur over a period of 12 to 18 months. Clinical and histopathological evidence suggests that it is caused by an isolated lesion of the vestibular nerve, although the exact aetiology remains obscure. Vestibular neuronitis is a relatively common condition in general practice, but has lacked clear definition, partly as a result of confusion over its nomenclature. Current knowledge of vestibular neuronitis is reviewed. Clinical diagnostic criteria are described, and the diagnosis and differential diagnosis of the syndrome in general practice are outlined. There remains a need to describe the occurrence of vestibular neuronitis in general practice in greater detail.

Keywords: vertigo; vestibular diseases; ear diseases; aetiology; diagnosis; management of disease.

Introduction

VESTIBULAR neuronitis is a relatively common condition in general practice manifested by a spectacular symptom: the sudden onset of vertigo in a previously well, young or middle-aged, adult. The vertigo is initially severe, then subsides gradually over a period of days or weeks. There is no hearing loss or other neurological abnormality — it is this absence of associated symptoms and signs which forms the basis for the recognition of the disease.

There has been some confusion over nomenclature which has left the impression that the syndrome lacks definition. This is not the case, and there is no doubt that vestibular neuronitis exists as a distinct condition. There are few general practitioners who have not seen epidemic clusters of patients with this syndrome presenting to their practice over a period of days or weeks in winter and spring.

There is a large body of medical literature describing the syndrome of vestibular neuronitis and the older research papers provide much of what is known. It is now recognized as a benign condition which seldom finds its way into specialist clinics. It has become, in effect, a condition managed by general practitioners.

The purpose of this article is to review what is known about the condition and to provide clear practical guidelines for its diagnosis and management in general practice.

Nomenclature

The term vestibular neuronitis first appeared in a classic paper by Dix and Hallpike which was published in 1952. The term was based on their clinical findings which suggested that the syndrome was caused by an isolated lesion of the vestibular nerve and its central connections. This view, and the term, remain widely accepted. When the syndrome occurs in epidemics it has been termed epidemic vertigo.

The terms acute labyrinthitis and epidemic labyrinthitis have occasionally been used to describe the syndrome but suffer from the disadvantage that there is no evidence that the disease involves the labyrinth. Strictly speaking, the term acute labyrinthitis describes a different and distinct syndrome which involves hearing loss and which usually occurs as a well established complication of specific viral and bacterial infections.

The term that will be used here is vestibular neuronitis. Hallpike used the term neuronitis to describe a lesion of the vestibular nerve and/or the vestibular nuclei and its second order neurones. If one could say the lesion involved the peripheral vestibular nerve alone, neuritis would be a more appropriate term, but one cannot say this with certainty. An alternative term, vestibular neuropathy, has been suggested as there is no firm evidence that the pathology involves inflammation, but this term lacks general acceptance.

Clinical features

Vestibular neuronitis is diagnosed using three clinical diagnostic criteria:

- vertigo: usually sudden onset;
- an absence of cochlear onset or symptoms (deafness and tinnitus);
- an absence of associated neurological symptoms and signs.

A non-essential criterion is a reduced or absent response to the caloric test.

The central feature is vertigo, usually occurring without warning. The onset is described as sudden in 73% of cases, but in a considerable minority of cases (27%) onset is described as gradual with a short prodrome (one or two days) of feeling ‘off balance’. Onset often occurs at night with symptoms present on waking. The vertigo varies in severity from a vague sensation of unsteadiness to it being severe and incapacitating, but is usually severe enough to cause nausea (94% of cases) and vomiting (54% of cases). It is commonly sufficiently severe, or sufficiently worrying, to cause presentation to a hospital accident and emergency department where it is the most common cause of vertigo.

In severe attacks the vertigo is described as constant but in milder attacks it is usually described as episodic. Attacks are precipitated or aggravated by movement of the head. There is a conspicuous absence of associated symptoms. In particular, there is no deafness or tinnitus, or symptoms suggesting brainstem involvement. In fact, the patient is usually an otherwise healthy young or middle-aged adult, there is no difference in prevalence between men and women.

On examination the only physical finding is nystagmus, which is present in 68% of cases; range 0–100%. The nystagmus is fine horizontal or rotary. It is always directed away from the side of the lesion, and this serves as an important localizing sign. The patient usually has an unsteady gait with a tendency to fall; falling is to the same side as that of the lesion. Nystagmus is only present during the acute phase of the disease and resolves within between seven and 25 days after onset.


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findings and hearing are always normal.\footnote{Clustering is usually unilateral\textsuperscript{29} but may be concurrently or sequentilla bilaterally.\textsuperscript{3,36} When unilateral, the affected sides are equally distributed among cases.\textsuperscript{31}}

Vestibular neuritis is a benign condition. The severe initial phase of the disease usually lasts between two and three days\textsuperscript{18} but may last a week or longer. The recovery phase is characterized by mild and gradually resolving vertigo or unsteadiness and usually lasts days or weeks.\textsuperscript{19} The course of the whole illness averages six weeks\textsuperscript{17,19} but may last nine weeks or longer.\textsuperscript{18}

**Association with infectious illness**

An interesting feature of vestibular neuritis is its association with preceding or concurrent infectious illness; this occurs in between 43\%\textsuperscript{13} and 46\% of cases.\textsuperscript{1} Most of these infections have been found to be non-specific upper respiratory tract infections,\textsuperscript{1,2,21,25-28,32-34} influenza,\textsuperscript{6,14,16,17,35,36} or focal sepsis of the upper respiratory tract, such as tonsillitis,\textsuperscript{1} and dental sepsis.\textsuperscript{1,6} A particularly high risk appears to be associated with sinusitis, which is present in up to 50\% of patients.\textsuperscript{26} A small minority of cases have been associated with a variety of apparently unrelated illnesses including gastroenteritis,\textsuperscript{2} hepatitis and brucellosis.\textsuperscript{37} Interestingly, there is not any clear association with acute or chronic otitis media.

**Epidemic vestibular neuritis**

Although usually sporadic,\textsuperscript{6,8,10,15,30,38} vestibular neuritis has frequently been noted to occur in epidemics.\textsuperscript{7-10,16,35,39-44} One of the earliest accounts of the disease described an epidemic among farmers and their labourers in a canton in Geneva in 1888\textsuperscript{41} and Walford described an interesting epidemic which occurred in an artists' colony in Chelsea, London and which became known locally as 'the stagers'.\textsuperscript{41}

A number of studies have noted the occurrence of vestibular neuritis among disease contacts,\textsuperscript{7,16,45} particularly family members,\textsuperscript{7,8,16,35,41} and neighbours.\textsuperscript{15} In an individual general practice epidemiology are frequently followed by a clustering of patients presenting over a period of days or weeks.\textsuperscript{8,16,35,39,42,46} This epidemic clustering can at times serve as a helpful clue in diagnosis.\textsuperscript{10} Epidemics are more common in winter and spring.\textsuperscript{1,7,17,41}

**Associated clinical syndromes**

In some patients, recurrent attacks of vertigo lasting days or weeks recur over a period of many months or years. This multiple attack variety of vestibular neuritis occurs in up to 43\% of patients, and is more common in younger patients and in patients who have suffered a more severe initial attack of vertigo.\textsuperscript{12} It is less common in patients who give a clear history of antecedent respiratory tract infection.\textsuperscript{2} Subsequent attacks are transitory (lasting less than 24 hours), and each attack is shorter than the one preceding it.\textsuperscript{8}

The place of this multiple attack variety of vestibular neuritis in the scheme of things has been difficult to resolve. Many authors believe it to be a severe variant of vestibular neuritis,\textsuperscript{1,5} while many others believe it is best considered a separate condition.\textsuperscript{2,21,26,47,48} Shuknecht and Kitamura have suggested, on the basis of clinical and histopathological findings, that the two syndromes should be considered variants of the same condition, and that the differences in severity is probably related to the degree of cell death in the vestibular neurones.\textsuperscript{5}

Equally perplexing is the occasional report of 'epidemic vertigo' to which are added symptoms and signs of brainstem involvement, in particular oculomotor dysfunction.\textsuperscript{7,8,10,11,49} This syndrome appears only to occur in epidemics.\textsuperscript{7,8}

The wider clinical spectrum of this condition falls outside the definition of vestibular neuritis as described by Dix and Hallpike\textsuperscript{1} and the view that it should be considered a separate condition has been convincingly argued on the basis of the presence of brainstem symptoms and signs, normal response to caloric testing and the presence of abnormal leukocytes in the peripheral blood.\textsuperscript{6,19} Although this syndrome has gained a place in the medical literature, it is much less common than vestibular neuritis. Hallpike commented that he had never seen a case, and speculated that the condition was rare.\textsuperscript{12}

The extent to which it should be considered a variant of vestibular neuritis remains unclear. In clinical practice, when overt symptoms or signs of brainstem involvement are also present with vestibular neuritis, further investigation to rule out cerebrovascular or neoplastic diseases is essential. When the syndrome occurs in an epidemic it is probably best considered to be viral brainstem encephalitis, and should be managed as such.\textsuperscript{6,8,10,49}

**Epidemiology**

Vestibular neuritis occurs most commonly in young and middle-aged adults aged between 20 and 60 years.\textsuperscript{1,2,6,21,25-27,28,32-34,50,51} The mean age of onset is 39 years; the oldest recorded case was a patient aged 73 years and the youngest case a patient aged 15 years.\textsuperscript{19} However, the syndrome described as 'epidemic vertigo of childhood' is indistinguishable from vestibular neuritis and may well be the same condition.\textsuperscript{38}

If this is the case, vestibular neuritis may be much more common in childhood than was previously thought. Vestibular neuritis occurs worldwide with no particular regional or geographical preference. It has been reported in the United Kingdom,\textsuperscript{1} France,\textsuperscript{11} Germany,\textsuperscript{5,25} Denmark,\textsuperscript{9} Sweden,\textsuperscript{7} the United States of America,\textsuperscript{5,45} Kenya,\textsuperscript{13} Malaysia,\textsuperscript{39} Australia,\textsuperscript{38} and Japan.\textsuperscript{56}

There has been no true epidemiological survey to determine the incidence rate of vestibular neuritis in a population at risk, although in one general practice in the UK there were 14 cases of vestibular neuritis in a practice population of 8200 in one year giving an incidence rate of 1.71 cases per 1000 of the population per year.\textsuperscript{17}

**Aetiology**

It is a well established axiom of otoaryngology that when the inner ear is involved in disease, both cochlear and vestibular elements are compromised, leading to both hearing loss and vertigo. It is on this basis that Dix and Hallpike suggested that in vestibular neuritis, in which vertigo occurs in the absence of hearing loss, the inner ear is not involved and that the lesion lies in the vestibular neurones central to the labyrinth.\textsuperscript{1} This view is now generally accepted.

The absence of overt features of wider brainstem involvement suggests that the lesion is limited to the vestibular nerve and its connections. The lesion may well be limited to the peripheral vestibular nerve itself, but no such assumption can be made.\textsuperscript{12} This view is supported by autopsy histopathological studies of patients who had died some time after the onset of symptoms of unrelated clinical problems. These studies have demonstrated isolated degeneration of the superior vestibular nerve and vestibular ganglion\textsuperscript{6,45,57} with a deficiency in the population of the nerve fibres,\textsuperscript{2} and light and electron microscopic findings of myelin degeneration.\textsuperscript{54} One study demonstrated associated thrombosis of a large vessel in the internal auditory meatus,\textsuperscript{57} although another study could find no evidence of vascular occlusion in four specimens studied.\textsuperscript{2} More recently, magnetic resonance imaging has demonstrated distinct decreased visualization of the vestibular aqueduct during the acute episode of the illness.\textsuperscript{58}

Although by no means certain, it is probably most helpful to think of vestibular neuritis as being a mononeuropathy or
Although by no means certain, it is probably most helpful to think of vestibular neuritis as being a mononeuropathy or polyneuropathy affecting the vestibular nerve and its connections.

An infective aetiology of vestibular neuritis has long been hypothesized, on the basis of its association with respiratory tract, and other infections, and on its frequent epidemic occurrence. A number of studies have demonstrated serological evidence of recent viral upper respiratory tract infections, particularly those caused by influenza virus A, influenza virus B and adenoviruses, but also herpes simplex virus, cytomegalovirus, Epstein-Barr virus, rubella virus, and parainfluenza virus.7,59,60 No serological association could be demonstrated with other encephalitides: eastern equine encephalitis, western equine encephalitis, St Louis encephalitis, lymphocytic choriomeningitis or poliovirus.14

Of importance is that, despite clear serological evidence of recent viral infection no virus has ever been isolated from the blood, respiratory tract or cerebrospinal fluid despite repeated attempts.17,60

Whether the disease is caused by direct infection, localized thrombosis, compression as a result of oedema, or an autoimmune reaction, remains unclear. Hallpike has hypothesized that it may be a result of a highly selective vestibulotoxin, similar to streptomycin.12

The characteristic interval which separates the onset of the respiratory tract infection and the onset of vertigo, and the fact that the virus has never been recovered from the respiratory tract or cerebrospinal fluid, may suggest that the disease is caused by an immune mediated complication of infection rather than direct viral infection of the nerve. Immune mediated neurological disease is a well recognized sequela of infectious fevers, and one interesting parallel of localized immune mediated peripheral neuropathy is vaccine-induced brachial neuropathy which occasionally complicates deltoid immunization. The view that vestibular neuritis is immune mediated is largely speculative but is supported by a study which demonstrated an increased risk of vestibular neuritis associated with a specific human leucocyte antigen genotype, and an elevation in the ratio of T-helper (CD4) to T-suppressor (CD8) cells in patients during the acute episode.61

Diagnosis and differential diagnosis

The diagnosis of vestibular neuritis is essentially one of exclusion. Diagnosis rests first on confirming the presence of vertigo (as opposed to dizziness), secondly, on confirming the absence of associated symptoms and signs, and finally, on excluding other less common conditions which may rarely cause isolated vertigo. True vertigo can usually be clearly differentiated from dizziness by its spinning character, the presence of concurrent nausea and vomiting, and the presence of nystagmus. The important associated symptoms and signs which require exclusion are: hearing loss, tinnitus, and evidence of brainstem involvement (in particular diplopia and dysarthria).

When vestibular neuritis recurs over a period of months each subsequent attack is less severe than the one preceding it. If this is not the case a diagnosis of vestibular neuritis cannot be made and other causes should be considered.

Potentially serious conditions which may be confused with vestibular neuritis are much more common in elderly patients. Here mistakes may be made, and a full general and neurological examination is of particular importance. Five conditions should be considered in the differential diagnosis, particularly in elderly patients.

Vertebrobasilar insufficiency or brainstem infarction or haemorrhage should be considered. Vertebrabasilar insufficiency is a relatively common condition among elderly patients, especially among those who are hypertensive. Sudden vertigo is the most common symptom of vertebrobasilar insufficiency but is usually associated with other symptoms and signs of brainstem involvement, such as diplopia, dysarthria and hemiparesis. Vertigo may be the only symptom (in 25% cases), although other symptoms of brainstem involvement usually reveal themselves in time.18 Where doubt exists referral for a scan using computerized tomography may be justified.

Ménière's disease can nearly always be excluded on the basis of the absence of hearing loss.19 Hearing loss may rarely be absent early in the course of Ménière's disease, but where doubt exists the character of the vertigo may help to differentiate the two conditions. The vertigo of Ménière's disease is characteristically episodic, lasting a few hours, whereas the vertigo of vestibular neuritis is usually constant, lasting several days.5

Disequilibrium of ageing is caused by degenerative changes in the peripheral labyrinth and causes unsteadiness following sudden movement of the head.63 Symptoms are usually milder than vestibular neuritis, and the onset more gradual.

Acoustic neuroma usually causes hearing loss and/or tinnitus.5 Ototoxic agents, such as streptomycin or phenytoin may cause vertigo as the main feature and are easily excluded by a careful drug history.

In younger adults one final condition to consider is multiple sclerosis. Multiple sclerosis does very rarely present as isolated vertigo, and when it does it may be impossible to distinguish from vestibular neuritis until further evidence of central nervous system involvement emerges. It should be stressed that this presentation is rare, whereas vestibular neuritis is common.

In children, vestibular neuritis is almost certainly synonymous with the condition known as epidemic vertigo of childhood.64 In any event the two conditions have the same prognosis and in clinical practice should be managed in the same way. Posterior fossa tumours in children may present with ataxia but this is usually accompanied by signs and symptoms of increased intracranial pressure or multiple cranial nerve palsies. A scan of the brain using computerized tomography should be considered where doubt exists.

Investigations are seldom justified in patients presenting in general practice with typical features of vestibular neuritis. There is no specific investigation which confirms the diagnosis. Caloric tests are invariably abnormal, but although this response was one of the disease criteria suggested by Dix and Hallpike,1 it lacks specificity and is probably not justified as an investigation in general practice.

Management

The management of patients with vestibular neuritis includes reassurance, explanation, advice, and specific treatments. Vestibular neuritis may be a frightening condition for patients who are often aware that vertigo can be caused by serious neurological disease, such as a stroke or a brain tumour. Patients should be constantly reassured that the condition is quite common and not at all serious. Specific anxieties should be gently explored and answered with specific reassurances.

The nature of the condition should be explained in detail including information that the vertigo is caused by inflammation in the inner ear and is due to a virus (although this may not be strictly true, it is easy to understand, and close enough to the truth). It is useful to give patients some idea of the prognosis, that is, that the vertigo will get better by itself in a few days, although a feeling of unsteadiness may persist a little longer. It is probably a good idea to be a little more circumspect and limit explanation in patients in whom the diagnosis is in doubt, such as in elderly patients, until the situation can be clarified.
Vestibular neuronitis is a common cause of vertigo in general practice. It is a benign condition which often follows an infectious illness, but its cause remains elusive. Management comprises reassurance and symptomatic treatment.

References

64. Basler L. Benign paroxysmal vertigo of childhood (a variety of Vestibular Neuritis). Brain 1964; 87: 141-152.

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