The Journal of Laryngology and Otology
August 1980. Vol. 94. pp. 877–883.

Vestibular neuronitis

Electron microscopy of Scarpa's Ganglion

By I. FRIEDMANN and W. HOUSE

THE idiopathic or viral vestibular neuronitis syndrome was first described by Dix and Hallpike in 1952 but its pathology and pathogenesis have remained obscure. The non-specificity of the lesion has been emphasized by Goodhill (1979) and other authors. A viral aetiology has been assumed but not proven (Paparella, 1973; Palva *et al.*, 1978).

Morgenstein and Seung (1971) have described some degenerative changes in Scarpa's ganglion and the superior division of the vestibular nerve in vestibular neuronitis. Sando, Hemenway, Miller and Black (1974), in a histopathological study of the cause of vertigo in otosclerotic patients, have noted some degeneration of the vestibular nerve and some moderate vacuolar degeneration of some of the neurones of Scarpa's ganglion.

The ultrastructure of the healthy vestibular ganglion has been studied by Ballantyne and Engström (1969), and Colman, Friedmann and Wright (1975) described the ultrastructural changes of the vestibular ganglion of a woman with a characteristic history of Menière's disease treated with ultrasound. The neurones contained lipoid inclusions and multicystic cytosomes and lipofuscin granules.

Clinical history

Mrs. N.T., a 47-year-old lady was first examined on 15 October 1975, with a history of two months' dizziness which was rather sudden in onset and was accompanied by severe nausea at the time of onset. She had no feeling of pressure in either ear, no tinnitus and no hearing loss, but she did have a feeling of a weight on the top of her head, and any motion caused her to feel unsteady.

Physical examination revealed normal tympanic membranes and she had normal hearing by pure tone audiometry and 100 per cent speech discrimination in both ears. X-rays of the internal auditory canals were normal, and blood studies, FTA, glucose tolerance and thyroid studies were also normal. Electronystagmography did show a very slight reaction to bithermal caloric stimulation in her right ear, but when the spontaneous nystagmus was corrected she had a 100 per cent decreased vestibular response on that side. This was unchanged about a year later when the ENG was repeated.

The clinical diagnosis was one of vestibular neuronitis and Mrs. N. T.was treated with a vasodilator regimen over a period of about a year, but there was no marked change in her symptoms. Because she remained so constantly unsteady, on 15 January 1975 a middle fossa section of the right vestibular nerves was

From Department of Electron Microscopy, Northwick Park Hospital and Clinical Research Centre, Harrow and the Ear Research Institute, Los Angeles.

https://doi.org/10.1017/S0022215100089635 Published online by Cambridge University Press

carried out. It was possible to do complete sections on the nerves and to remove Scarpa's ganglion.

Immediately after surgery she continued to feel unsteady, but she did lose the sensation of nausea which had been such a difficult problem for her. Six months later, she improved markedly and felt considerably better, 'though she still felt slightly unsteady when she moved suddenly.

About a year and a half after surgery, the hearing loss in the operated ear had dropped from an average of 10 dB to an average of 37 dB, and discrimination had dropped to 72 per cent. However, the loss is not progressive.

Materials and Methods

The surgical specimens were fixed in 3 per cent glutaraldehyde, transferred into a cacodylate buffer solution, post-osmicated and embedded in Epon. Semi-thin sections were stained with 1 per cent Toluidin-blue for light microscopy, and thin sections contrasted with uranyl-acetate and lead citrate were examined under a Phillips 300 electron microscope.

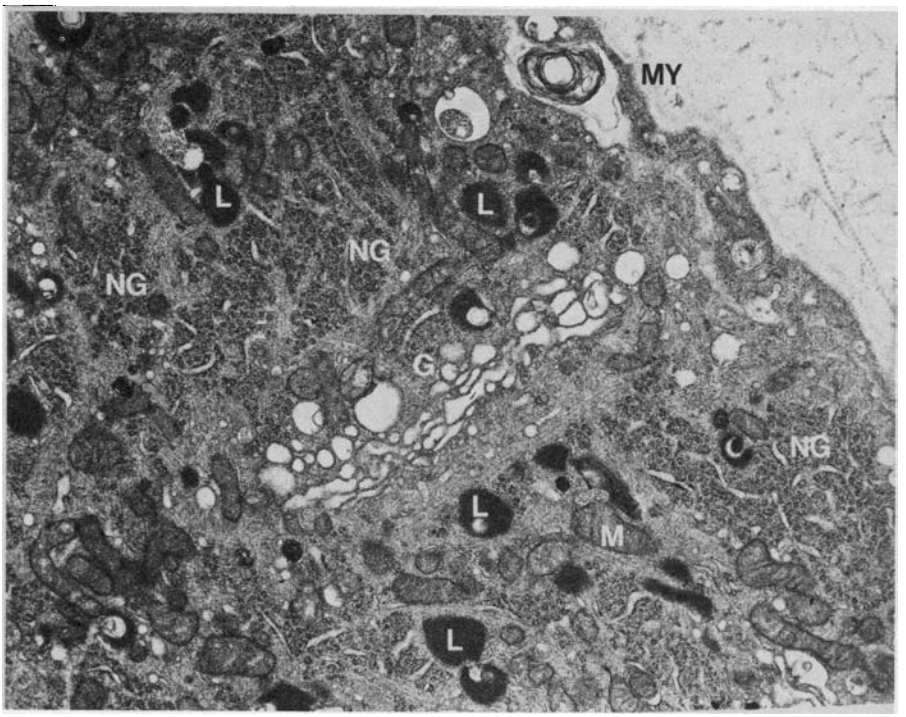


FIG. 1

Part of a neurone from Scarpa's ganglion surgically removed. Note prominent Golgi apparatus (G), multiple Nissl granules (NG) with abundant ribosomes and numerous mitochondria (M). There are many lipoid droplets (L) in the perikaryon and occasional myelin figures (MY) near the surface. ×30180.

https://doi.org/10.1017/S0022215100089635 Published online by Cambridge University Press

Light microscopy

Semi-thin sections showed a network of myelinated fibres of the vestibular nerve surrounding large numbers of neurones of Scarpa's ganglion. Some of the neurones were pale staining, others were more darkly stained.

Electron microscopy

The neurones of the vestibular ganglion measured 15 μm to 50 μm and contained large nuclei with prominent nucleoli. The darkly stained cells were richly endowed with rough endoplasmic cisternae forming the granules of Nissl (Fig. 1). The paler ones showed marked dilation of the endoplasmic cisternae (Fig. 2). The Golgi apparatus was prominent (Fig. 1) and the cytoplasm contained numerous glycogen granules. The basal lamina surrounding the cells was occasionally split by an amorphous material.

The significant finding in most of the neurones was the presence of cytoplasmic inclusions with multivesicular membrane-bound cytosomes or lipofuscin-like granules scattered in the cytoplasm and frequently wholly occupying the neurone (Figs. 3, 4 and 5).

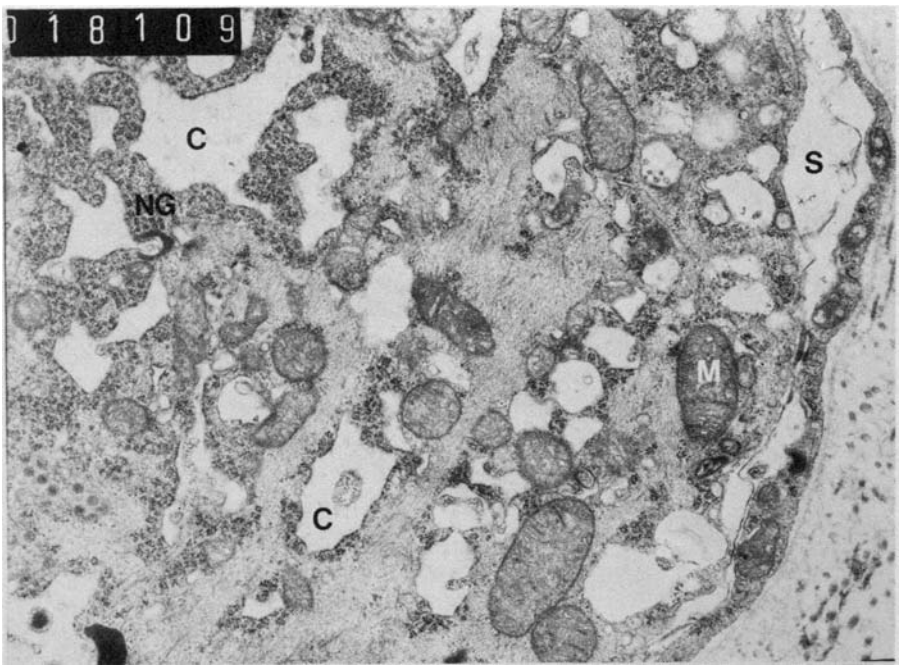


FIG. 2

Part of a different neurone from Scarpa's ganglion showing greatly distended endoplasmic cisternae (C) of the Nissl substance. Note the fibrillar cytoplasm and dilated (?oedematous) subcapsular space (S). ×29400.

https://doi.org/10.1017/S0022215100089635 Published online by Cambridge University Press

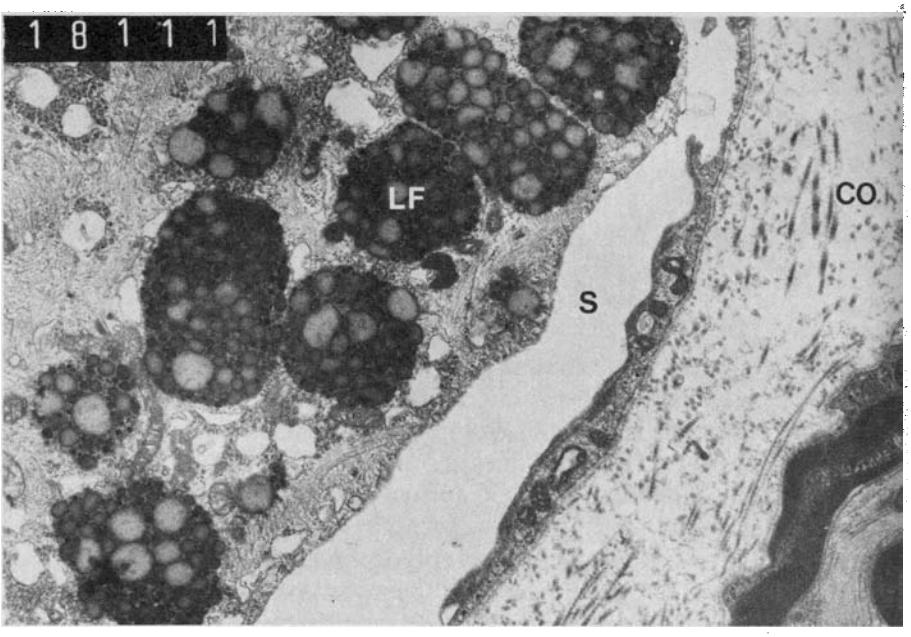


FIG. 3

Part of another neurone from Scarpa's ganglion containing multi-cystic cytosomes or lipofuscin-like granules (LF) of grape-like structure composed of round vesicles of uniform shape but varying lipid content. Collagen fibres (CO). ×22500.

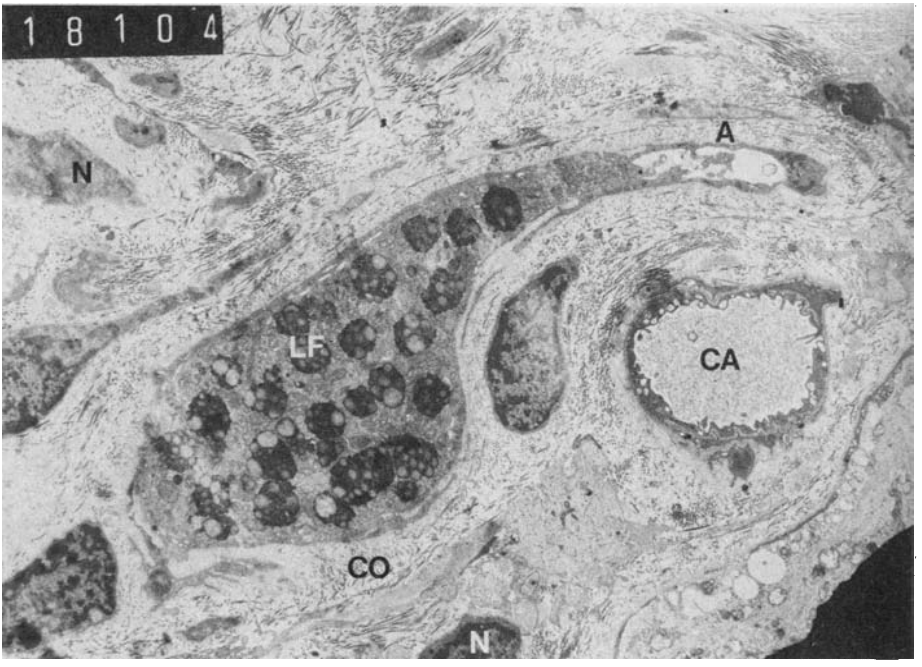


FIG. 4

A small neurone from Scarpa's ganglion surrounded by collagenous tissue. Note multicystic cytosomes or lipofuscin-like granules (LF) in the cell and vacuolated axon (A). There are several nuclei (N) of Schwann cells in the surrounding tissue which contains a small capillary (CA). ×6600.

https://doi.org/10.1017/S0022215100089635 Published online by Cambridge University Press

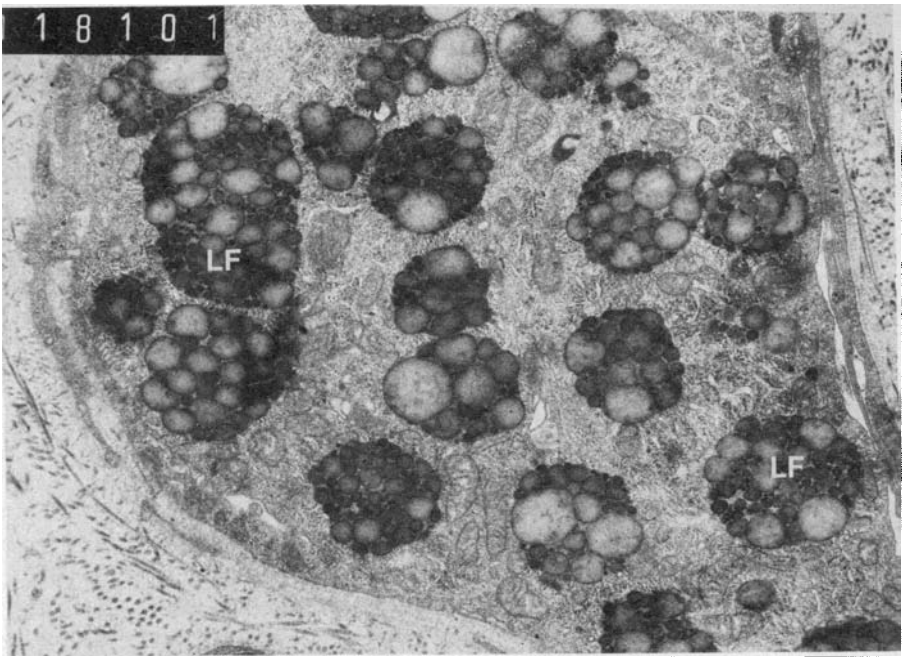


FIG. 5
Detail of Fig. 4. ×22500.

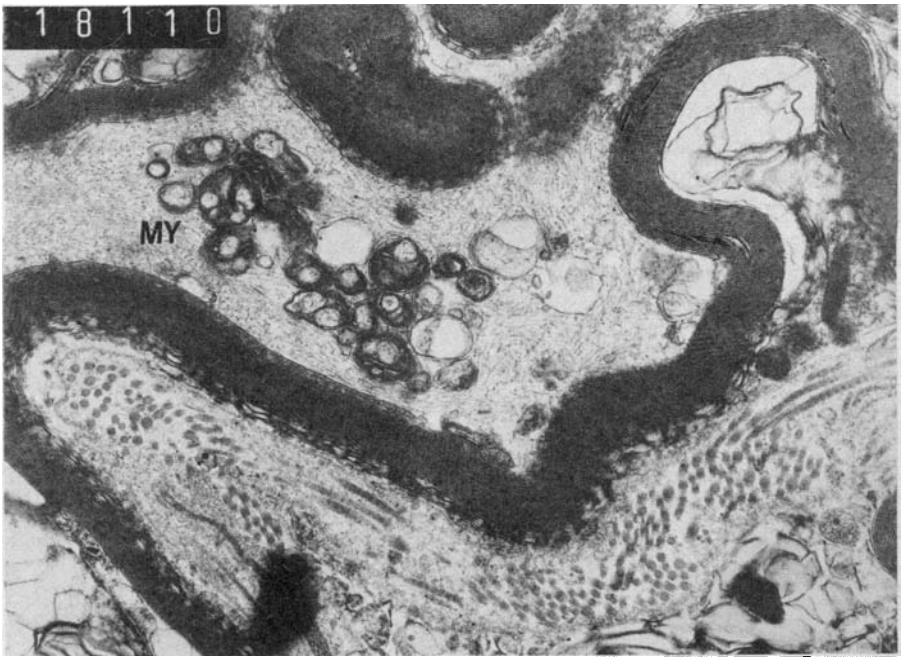


FIG. 6
Myelinated nerve fibre from superior vestibular nerve with many myelin figures (MY) in its axon. ×39000.

https://doi.org/10.1017/S0022215100089635 Published online by Cambridge University Press

The myelinated nerve fibres were on the whole well preserved and their myelin layers clearly defined. In contrast several axons were vacuolated or contained myelin figures of degeneration (Fig. 6).

Discussion

The neuronal lipofuscin-like inclusions observed in this case resembled those described in the vestibular ganglion of patients treated for Menière's disease (Friedmann, Cawthorne and Bird, 1965; Colman, Friedmann and Wright, 1975). These structures should not be ignored or considered to be of no pathological interest and/or significance since they participate in the everchanging cellular processes affecting the contents of the neurone and consequently its proper functioning.

The true nature of the inclusions of lipofuscin type has not been clearly established but they are considered to be true lysosomes. Brunk and Ericsson (1972) have identified acid phosphatases in the lipofuscin granules of rat brain ganglia forming the residual body type of lysosomes resulting from cellular autophagy. Accumulation of the residual bodies in the cells due to failing exocytosis (DeDuve, 1969) may have a profound effect on the cell. It may be assumed that such cells were more prone to undergo degenerative changes under pathological conditions than were 'young' cells free of lipofuscin (Brunk *et al.*, 1972).

Ballantyne and Engström (1969) observed no such inclusions in the neurones of normal vestibular ganglia. Lipofuscin-like granules occur, however, in ageing cells especially in non-dividing cells such as neurones. Engström *et al.* (1977) described them in the vestibular epithelia of elderly men and monkeys, and similar structures have been observed in cerebral lipidoses (Dekaban and Herman, 1974).

It may be suggested that neurones undergoing physiological 'wear and tear' through ageing or when affected by e.g. viral disease (following an upper respiratory tract infection) would suffer some fundamental degenerative changes responsible not only for the morphological phenomena described but also, as in our cases, for the clinical symptoms and signs of vestibular disease.

Galic and Helms (1977) investigated the vestibular nerve and ganglion from six patients with Menière's disease. There were large amounts of lipofusein present especially in some of the dark neuroides. Degeneration secondary to an underlying capillary lesion has been implicated in the neuronal changes. Furthermore the authors suggest that the pathologically altered neurones may play a role in the causation of transient vertiginous attacks.

Abstract

Scarpa's ganglion and the superior vestibular nerve of a patient with vestibular neuronitis examined under the electron microscope displayed

https://doi.org/10.1017/S0022215100089635 Published online by Cambridge University Press

degenerative neuronal changes and axonal lesions considered to be the cause of the clinical features of vestibular neuronitis.

REFERENCES

BALLANTYNE, J., and ENGSTRÖM, H. (1969) Morphology of the vestibular ganglion cells. *Journal of Laryngology and Otology*, **83**, 1.

BRUNK, U., and ERICSSON, J. L. E. (1972) Electron microscopical studies on rat brain neurons. Localization of acid phosphatase and mode of formation of lipofuscin bodies. *Ultrastructure Research*, **38**, 1–15.

BRUNK, U., ERICSON, J. L., PONTEN *et al.* (1972) Residual bodies and 'aging' in cultured human glial cells. *Experimental Eye Research*, **79**, 1.

COLMAN, B. H., FRIEDMANN, I., and WRIGHT, J. L. W. (1975) Menière's disease. Electron microscopy of the vestibular ganglion and end organs after ultrasound. *Acta Otolaryngologica*, **79**, 189–196.

DEDUVE, C. (1969) *Lysosomes in biology and pathology*, (ed. J. T. Dingley and H. B. Fell). Wiley, New York.

DEKABAN, A. S., and HERMAN, M. (1974) Childhood, juvenile and adult cerebral lipidoses. *Archives of Pathology*, **97**, 65–72.

DIX, M. R., and HALLPIKE, C. S. (1952) The pathology, symptomatology and diagnosis of certain common disorders of the vestibular system. *Annals of Otology, Rhinology and Laryngology*, **61**, 987.

ENGSTRÖM, H., ADES, H. W., ENGSTRÖM, B., GILCHRIST, D., and BOURNE, G. (1977) Structural changes in the vestibular epithelia in elderly monkeys and humans. *Advances in Oto-Rhino-Laryngology*, **22**, 93–110 (Karger, Basel).

FRIEDMANN, I., CAWTHORNE, T., and BIRD, E. S. (1965) The laminated cytoplasmic inclusions in the sensory epithelium of the human macula. *Journal of Ultrastructural Research*, **12**, 92.

GOODHILL, V. (1979) *Ear Diseases, Deafness and Dizziness*, p. 532. Harper and Row, Hagerstown, Maryland.

GALIC, M., and HELMS, J. (1977) Electron microscopic studies of lipofusein in the vestibular ganglion in Menière's disease (in German). *Inner Ear Biology*, pp. 243–261. Eds. Portmann, M., and Aran, J.-M. INSERM, Paris.

MORGENSTEIN, K. M., and SEUNG, H. I. (1971) Vestibular neuronitis. *Laryngoscope*, **81**, 131–139.

PALVA, T., HORTLING, L., YLIKOSKI, J., and COLLAN, Y. (1978) Viral culture and electron microscopy of ganglion cells in Menière's disease and Bell's palsy. *Acta Otolaryngologica*, **86**, 269–275.

PAPARELLA, M. M. (1973) Otological manifestations of viral disease. *Advances in Otorhinolaryngology*, **20**, 144–154.

SANDO, I., HEMENWAY, W. G., MILLER, D. R., and BLACK, F. O. (1974) Vestibular pathology in otosclerosis. Temporal bone histopathological report. *Laryngoscope*, **84**, 593–605.

https://doi.org/10.1017/S0022215100089635 Published online by Cambridge University Press

https://doi.org/10.1017/S0022215100089635 Published online by Cambridge University Press