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Pathology in Focus

Neural hearing loss in a child with poliomyelitis: a histopathological study

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Abstract

The temporal bones of a 26-month-old white female with a paralytic syndrome clinically and pathologically identical to poliomyelitis were examined. The aetiological agent was unknown although a non-poliomyelitis enterovirus infection seemed likely.

There was a complete absence of the cochlear neurons and substantially reduced peripheral and central axons with loss of some inner hair cells but preservation of outer hair cells. Scarpa's ganglion, and the geniculate ganglion were partially atrophied. The saccule and utricle were mildely dilated and Reissner's membrane of the apical turn was bulging.

In two previous audiological studies a 10–20 dB bilateral sensorineural hearing loss was found in poliomyelitis patients and a neuronal lesion was postulated which is now supported by our findings. This is a rare example of an almost pure neural hearing loss.

Key words: Poliomyelitis; Enterovirus Infections; Hearing Loss, Sensorineural; Spiral Ganglion; Histology

Introduction

Since the beginning of mankind poliomyelitis (polio) has been a scourge similar to the plague and leprosy. Polio is a clinical syndrome characterized by motor paralysis caused by degeneration of motor nuclei. Until the early 1950s, most cases of poliomyelitis were caused by the polio virus but after the development of poliovaccines by Salk and Sabin in 1953, the incidence of poliomyelitis caused by the polio virus has dramatically decreased.[1] Now there are sporadic cases of clinical poliomyelitis caused by other enterovirus infections such as coxsackie and echo viruses.[2] The immunologically suppressed[3] and newborns are particularly vulnerable.[4]

Weinstein *et al.* documented that the vagus and the glossopharyngeal were the nerves affected in 78 per cent of their patients and the eighth cranial nerve was the least involved (0.9 per cent).[5] Two studies from the same centre reported that patients with hearing losses from polio had flat thresholds of approximately 20 dB, postulated to be caused by a retrocochlear neural lesion.[6,7] Kelemen described the temporal bone of a premature neonate from a mother with acute poliomyelitis. Unfortunately no comment was made about the ganglion cell populations nor was the cause of death clarified.[8]

To the present time, apart from Kelemen's study, there have been no other temporal bone studies in polio. In this paper we present the histopathological findings in the temporal bones of a child with a polio-like ;syndrome.

Case report

A female infant was born after an uncomplicated full-term pregnancy to a 26-year-old mother. In her first eight months the infant's developmental milestones were normal. At six months she had her third diphtheria, polio and tetanus (DPT) inoculation and by eight months of age she could stand with support, responded to sound and could babble. Then she developed a high fever with an episode of hypoxia and left-sided tonic-clonic seizure. Immediately on admission she required mechanical ventilation because of diaphragmatic paralysis that was unresponsive to phrenic nerve stimulation. There was persistent mild elevation of cerebrospinal fluid protein. In her ninth month of hospitalization, however, her weakness progressed rapidly so that by the 10th month she had only eye movements and minimal finger motion. She had no facial expression and responded only to noxious stimuli. Audiometry was not performed. Computer tomography (CT), myelography, electroencephalography, electromyelography and nerve and muscle biopsies supported the clinical diagnosis of anterior horn cell disease. Her serum showed a 1:32 titre for polio virus consistent with vaccination. Virus isolation studies were not performed and limited study ruled out severe immune deficiency. She died from bronchopneumonia after 17 months of hospitalization.

At autopsy there was a degeneration of the grey matter and posterior columns of the cervical and lumbar spinal cord. The lumbar cord showed a subarachnoidal haemorrhage and intraparenchymal petechial haemorrhage, suggesting viral myelitis. There was a hydrocephalus ex vacuo.

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The meninges were unremarkable. The anterior and posterior horns of the spinal cord and all the motor nuclei of the brainstem demonstrated neuronal loss and gliosis consistent with healed polio.

The aetiological agent of the patient's disease remained unclear but was most likely to be an enterovirus.

Temporal bone study

The temporal bones were harvested 12 hours after death, fixed in 10 per cent formalin, decalcified in trichloroacetic acid and processed by the celloidin technique in the usual manner. The specimens were sectioned in the horizontal plane at a thickness of 20 μm, and every 10th section was stained with haematoxylin-eosin, and studied by light microscopy.

Left temporal one

The external auditory canal was normal. There was some thickened mucosa and inflammatory cells in the tympanum consistent with chronic otitis media. The ossicles were normal in size, shape and position. The facial nerve was severely atrophied in its entire course, with substantial reduction of cell bodies in the geniculate ganglion (Figure 1). The stapedius and tensor tympani muscles looked normal.

The cochlea was normally shaped (Figure 2(a)). Some inner hair cells were identifiable and there were plump outer hair cells (Figure 2(b)). The stria vascularis had a loose pattern that represented post-mortem artifact. In Rosenthal's canal, only a few pyknotic cell bodies of degenerating neurons could be identified and peripheral and central axons were substantially reduced (Figure 2(c), (d)). The vestibular endorgans, cupula and maculae are normal in shape and size but there was reduction of afferent nerve fibres and loss of ganglion cells. The saccule and the utricle were mildly dilated.

Right temporal bone

In the middle ear and mastoid cells there was evidence of sero-mucinous otitis media with thick fluid and macrophages. The geniculate ganglion contained very few ganglion cells and the facial nerve was severely atrophied. In the organ of Corti, although autolysis made quantification difficult, some inner hair cells (Figure 3b, d) and many outer hair cells could be clearly seen. In Rosenthal's canal the neuron population was reduced to 10 per cent and the remaining cell bodies were surrounded by collections of mononucleated cells (Figure 3(c)). The saccule and utricle were mildly dilated, and the endolymphatic duct and cochlear aqueduct were patent and were free of inflammatory cells.

The internal auditory canal contained some blood and mononucleated cells. The vestibular nerves stained very faintly compared to the surrounding tissues and approximately only 20 per cent of the vestibular ganglion cells remained.

Discussion

This child had a paralytic syndrome that was probably caused by a non-polio enterovirus infection but had no

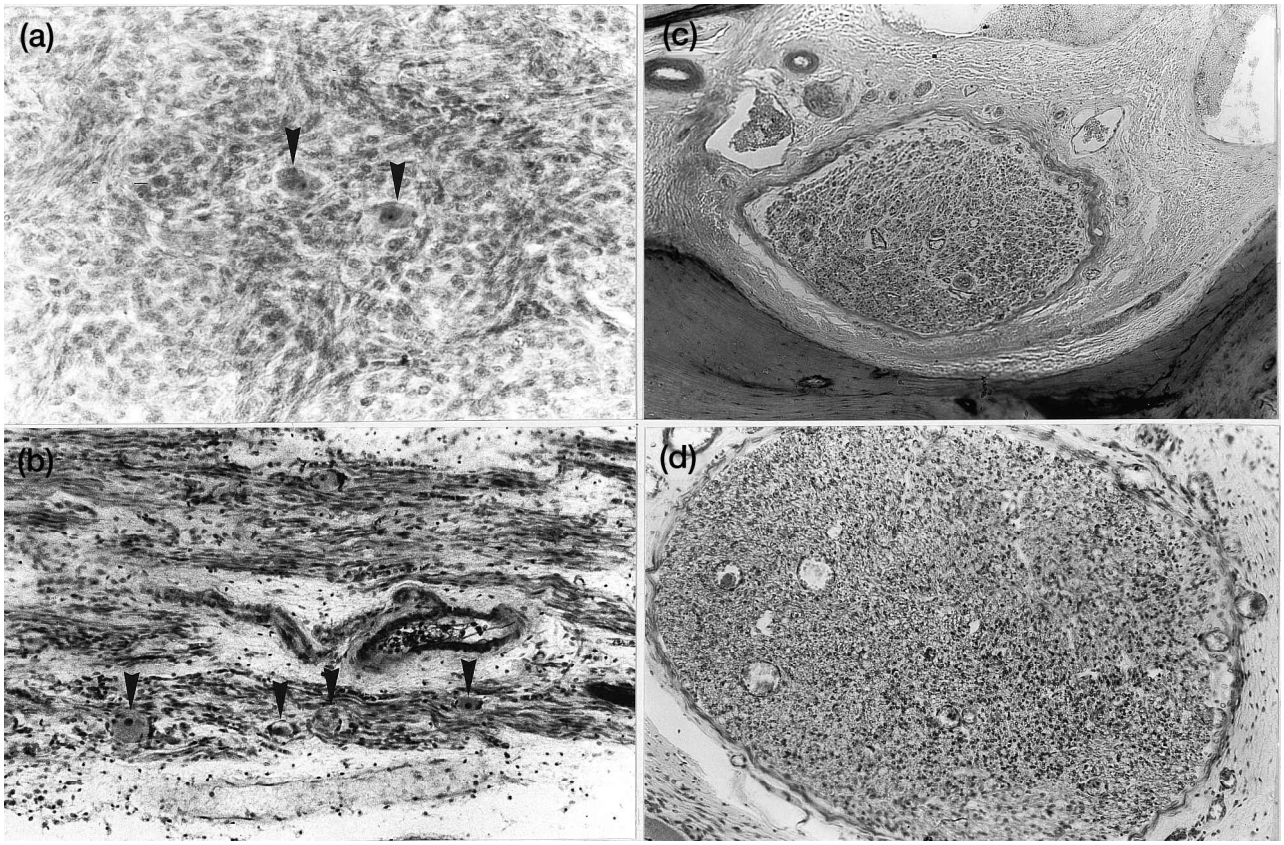

FIG. 1

(a) Left geniculate ganglion with a substantially reduced ganglion cell population (arrowheads) of the geniculate ganglion. (H & E; ×200). (b) Left side Scarpa's ganglion. Normal cell bodies are sparse, and the remaining ganglion cells are degenerating. (H & E; ×200). (c) Vertical segment of the left facial nerve with few nerve fibres (H & E; ×200). (d) For comparison, a normal facial nerve in a five-day old newborn (H & E; ×200).

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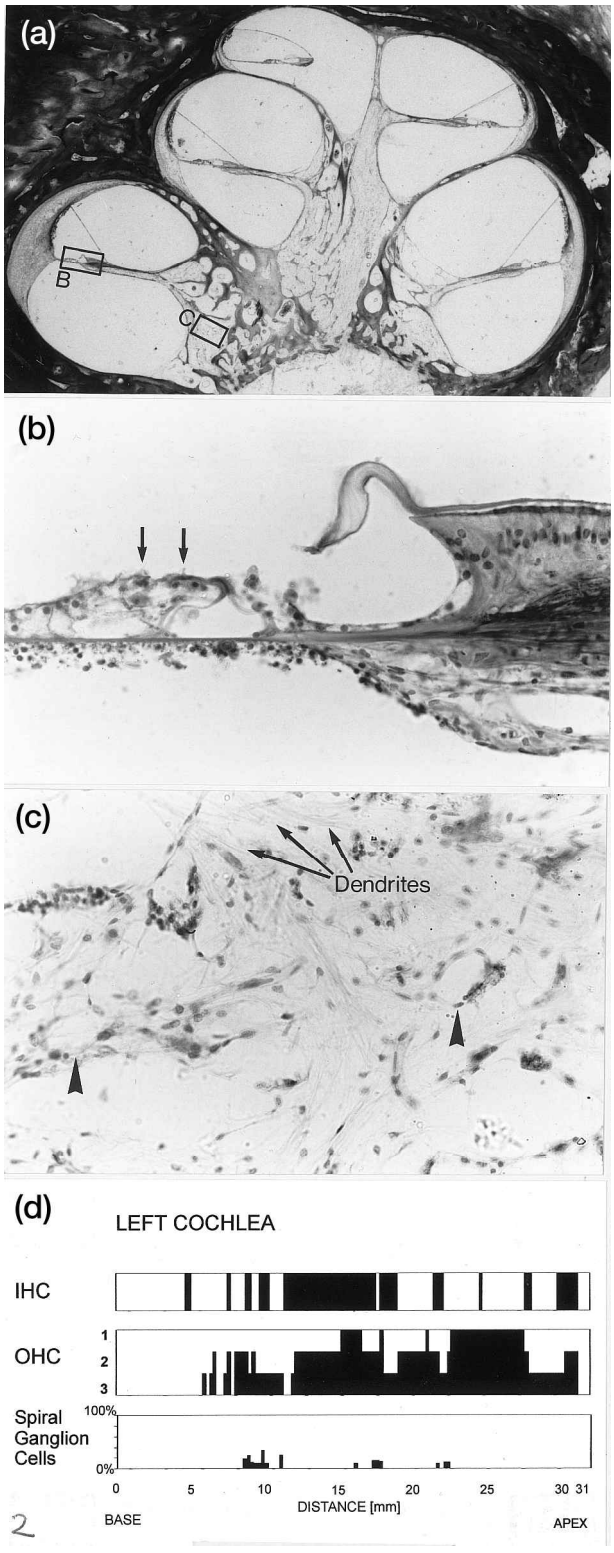

FIG. 2

(a) Overview of the left cochlea and the internal auditory canal (IAC). No neuronal structures are seen in the modiolus and the IAC (H & E; ×20). (b) Magnified view of the organ of Corti of the basal turn. There is a degenerated inner hair cell and plump outer hair cells with clumped sterocilaie (arrows) (H & E; ×100). (c) Higher magnification of Rosenthal's canal. There are few ganglion cells but normal satellite cells remain (arrow heads) (H & E; ×100). (d) Bar diagrams of present hair cell and spiral ganglion populations (black areas).

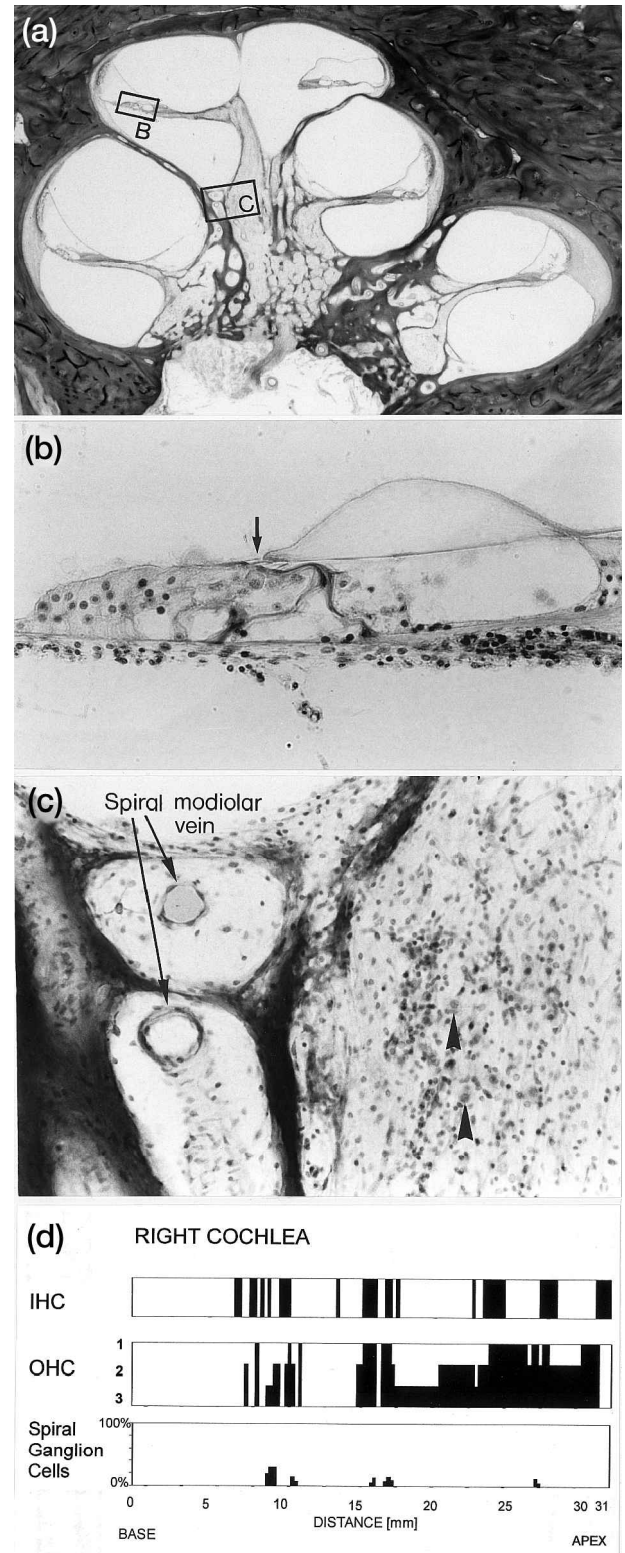

FIG. 3

(a) Overview of the right cochlea. The modiolus seems to be devoid of neural structures and there is only a thin remnant of the cochlear nerve (H & E; ×20). (b) Higher power view of the organ of Corti of the apical turn. An inner hair cell and outer hair cells (arrow) with stereocilia are clearly identifiable. The tectorial membrane is normal (H & E; 200). (c) The spiral modiolar vein has no sign of inflammation. Rosenthal's canal is infiltrated with small round cells surrounding few spiral gangion cells (arrow heads) (H & E; ×100). (d) Bar diagrams of present hair cell and spiral ganglion populations (black areas).

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clinical evidence of an immune deficiency although tests for human immunodeficiency virus (HIV) and detailed immunological studies were not performed.

The polio virus is known to be neurotrophic with an affinity for motor neurons especially the gray matter of the anterior horn of the spinal cord and the motor nuclei of the pons and medulla.[9] Nevertheless previous audiological studies with polio[6,7] had suggested involvement of the cochlear nerve which is a sensory nerve. This theory that is supported histologically by our findings of gross degeneration, and infiltrates of inflammatory cells around the spiral ganglion cells, which has been described in other tissues in polio-like infections.[4,9,10] Harris *et al.* reported accumulation of inflammatory cells around the spiral modiolar vein after viral antigen stimulation of the inner ear in guinea pigs[11] and Karmody found round cell infiltration of the spiral ganglion in early viral labyrinthitis.[12] In our case, the inflammatory reaction was comparatively less probably because of the protracted clinical course of 17 months.

Purely neuronal hearing loss, is extremely rare and has been documented only a few times.[13–15] Ishii and Toriyama,[13] Schuknecht and Donovan[14] and Yoon *et al.*[15] found a severe loss of cochlear neurons with well preserved organs of Corti in patients with sudden hearing loss. All these authors claimed viral infection as the aetiology of the isolated cochlear nerve degeneration. Additionally experimental infection of the inner ear in animals with the measles virus caused a loss of cochlear ganglion cells.[16] The pathological findings in our case, therefore seems to be similar to those described in previous reports of supposed viral infections and support the audiological hypothesis of a neural hearing loss in polio.[6,7] In our patient the vestibular end organs were also preserved but Scarpa's ganglions were affected to the same extent as the spiral ganglion. Isolated neural degeneration of the vestibular nerve is rare, and has been only documented by Nadol and Schuknecht.[17] The aetiology in their 92-year-old patient, however, was not known. In summary we have presented a temporal bone study in polio of probable enteroviral origin with severe degeneration of cochlear, vestibular and facial ganglia and preservation of the cochlear and vestibular end organs which, clinically, would have presented as a rare, primarily neural, hearing loss.

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References

1 Modlin JF. Poliovirus. In: Mandell R, Douglas G, Bennett JE, eds. *Principles and Practice of Infectious Diseases.* New York: Churchill Livingstone, 1990;1359–67
2 Melnick JL. Enterovirus type 71 infections: a varied clinical pattern sometimes mimicking paralytic poliomyelitis. *Rev Inf Dis Suppl* 1984;**6**:S387–90
3 McKinney RE Jr, Katz SL, Wilfert CM. Chronic enteroviral meningocephalitis in agammaglobulinemic patients. *Rev Infect Dis* 1987;**9**:334–56
4 Cherry JD. Enteroviruses: Polioviruses (poliomyelitis), coxsackieviruses, echovirues, and enteroviruses. In: Feigin RD, Cherry JD, eds. *Textbook of Pediatric Infectious Diseases.* Philadelphia: W.B. Saunders, 1987;1729–90
5 Weinstein L, Shelokov A, Seltser LR, Winchell GD. A comparison of the clinical features of poliomyelitis in adults and in children. *N Engl J Med* 1952;**246**:296–302
6 Batson R, McConnel F. A study of the hearing level following severe poliomyelitis. *Am J Dis Child* 1958;**95**:139–45
7 McConnel F, Batson R. Audiologic findings in 73 severe poliomyelitis patients. *Arch Otolarygol* 1962;**76**:32–7
8 Keleman G. Acute poliomyelitis of the mother with aural lesions in the premature infants. *Arch Otolaryngol* 1955;**62**:602–10
9 Bodian D. Histopathologic basis of clinical findings in poliomyelitis. *Am J Med* 1949;**6**:563–78
10 Hashimoto I, Hagiwara A, Komatsu T. Ultrastructural studies on the pathogenesis of poliomyelitis in monkeys infected with poliovirus. *Acta Neuropathol (Berl)* 1984;**64**:53–60
11 Harris JP, Fukuda S, Keithley EM. Spiral modiolar vein: Its importance in inner ear inflammation. *Acta Otolaryngol* (Stockh) 1990;**110**:357–65
12 Karmody CS. Viral labyrinthitis: Early pathology in the human. *Laryngoscope* 1983;**93**:1527–33
13 Ishii T, Toriyama M. Sudden deafness with severe loss of cochlear neurons. *Ann Otol Rhinol Laryngol* 1977;**86**:541–8
14 Schuknecht HF, Donovan ED. The pathology of idiopathic sudden sensorineural hearing loss. *Arch Otorhinolaryngol* 1986;**243**:1–15
15 Yoon TH, Paparella MM, Schachern PA, Alleva M. Histopathology of sudden hearing loss. *Laryngoscope* 1990;**100**:707–15
16 Davis LE, Johnson RT. Experimental viral infections of the inner ear. I. Acute infections of the newborn hamster labyrinth. *Lab Invest* 1976;**34**:349–56
17 Nadol JB Jr, Schuknecht HF. Pathology of peripheral vestibular disorders in the elderly. *Am J Otolaryngol* 1990;**11**:213–27

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