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View from Beneath: Pathology in Focus

Acoustic schwannoma of traumatic origin? A temporal bone study

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Abstract
A tumour of the singular nerve was found on examination of the temporal bones of a child who died 13 months after meningitis. The tumour consisted of a main mass with the appearance of an acoustic neuroma but close by and not connected were some nests of tumour cells inside the vestibule. This very unusual finding raises questions of the aetiology of this tumour which may have a bearing on the aetiology of other tumours of the VIIIth. nerve.

Introduction

On examination of a temporal bone from a two-year-old child with total deafness after labyrinthitis a tumour of the singular nerve was found. This was associated with a number of nests of tumour cells in the adjacent labyrinth. The case is described and the literature searched for similar cases.

Case reports

In August, 1980, a male child was born at term. He had no family history of genetic disorders or neurofibromatosis. In December, 1980 he was admitted to hospital for investigation of failure to thrive and a diagnosis of tyrosinosis Type 1 was made. At that time he responded normally to voices and

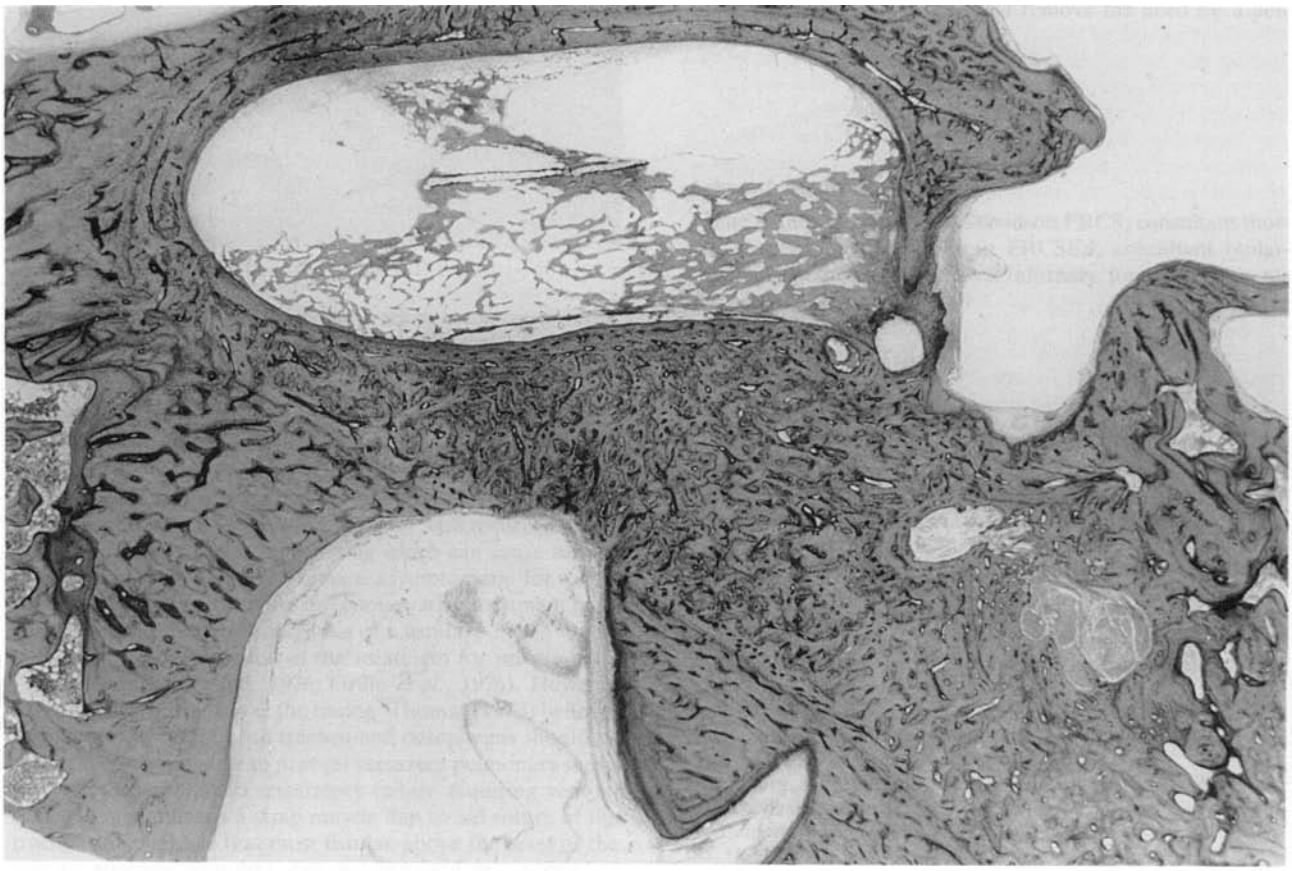

Fig. 1
The right temporal bone in horizontal section at the level of the round window. The basal turn of the cochlea shows labyrinthitis ossificans and the tumour can be seen in the residual lumen of the posterior semicircular canal. (Original magnification ×4)

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sound. In January 1981, a laparoscopy and liver biopsy were performed to exclude a hepatoma. In July 1981, whilst on the special care baby unit, he contracted *E. coli* meningitis, without evidence of middle ear infection. Following treatment of the infection with ampicillin, he recovered but remained hypotonic and bedridden. His condition gradually improved although he was thought to be deaf. This was confirmed by electrocochleography in September 1981, with no evoked response on either side to stimuli up to 100 dB. Otoscopy and tympanometry were normal. There was no further otological changes until death in August 1982 from hepatic failure.

Temporal bone preparation

The temporal bones were removed two hours post mortem. They were fixed in four per cent formaldehyde, decalcified and embedded in celloidin. The blocks were cut in the horizontal plane in serial sections of 20 microns thickness. Every tenth section was stained with haematoxylin and eosin and some selected sections stained for myelin with the Wolckes stain, and some for reticulin.

The temporal bones had a normal anatomical appearance. The middle ears and tympanic membranes were normal and the bony labyrinth did not exhibit any pathological findings. All turns of the cochlea were filled with loosely arranged connective tissue. A sparse inflammatory infiltrate accompanied this into the vestibule. The scala tympani of the basal turn and the vestibule were also partially filled with new metaplastic bone ie non-specific labyrinthitis ossificans. (Fig. 1). The findings in the spiral ganglion have been described previously (Pollak and Felix, 1985). Despite there being no organ of Corti and no nerve fibres in the osseous spiral lamina, the spiral ganglion was normal (21,700 spiral ganglion cells estimated). The posterior ampulla was also filled with connective tissuue and new bone but contained no tumour. A tumour was found, the main mass of which came from the distal portion of the singular nerve. (Fig. 2) Further small nests of tumour cells were found in the adjacent connective tissue in the vestibule. This tumour had the appearance of an acoustic schwannoma with Antoni type A tissue with nuclei arranged in the characteristic whorls, though there were no definitive palisades. (Fig. 3) There were nests of tumour cells in the surrounding connective tissue and adjacent labyrinth with the same appearance but were too small to show a distinctive whorlled pattern. They were not connected to the main tumour. (Figs. 4a, b)

Discussion

To explain the finding of a singular nerve tumour with adjacent labyrinthine nerve cell nests as a single pathology three differential diagnosis can be considered. They are (a) an acoustic schwannoma as an incidental finding, (b) a traumatic schwannoma secondary to the bacterial trauma to the labyrinth and (c) neurofibromatosis.

Acoustic schwannomas have been described by Stewart *et al.* (1975) as incidental temporal bone findings. They are usually intrameatal but may occur more laterally and have been reported in the vestibule by Wanamaker (1972) and in the cochlea by Karlan *et al.* (1972). They are not common in the labyrinth; Sataloff *et al.* (1988) have reviewed the 23 intralabyrinthine acoustic schwannomas reported in the literature up to 1988. Acoustic schwannomas are by definition solitary tumours, though this is blurred by the finding of more than one tumour in the same vestibular nerve as reported by Luetje *et al.* (1983). In our case, the main part of the tumour gives an appearance which, if found alone, would be easy to diagnose as a typical schwannoma. The presence of the multiple separate nests of similar cells inside the labyrinth, in an area usually

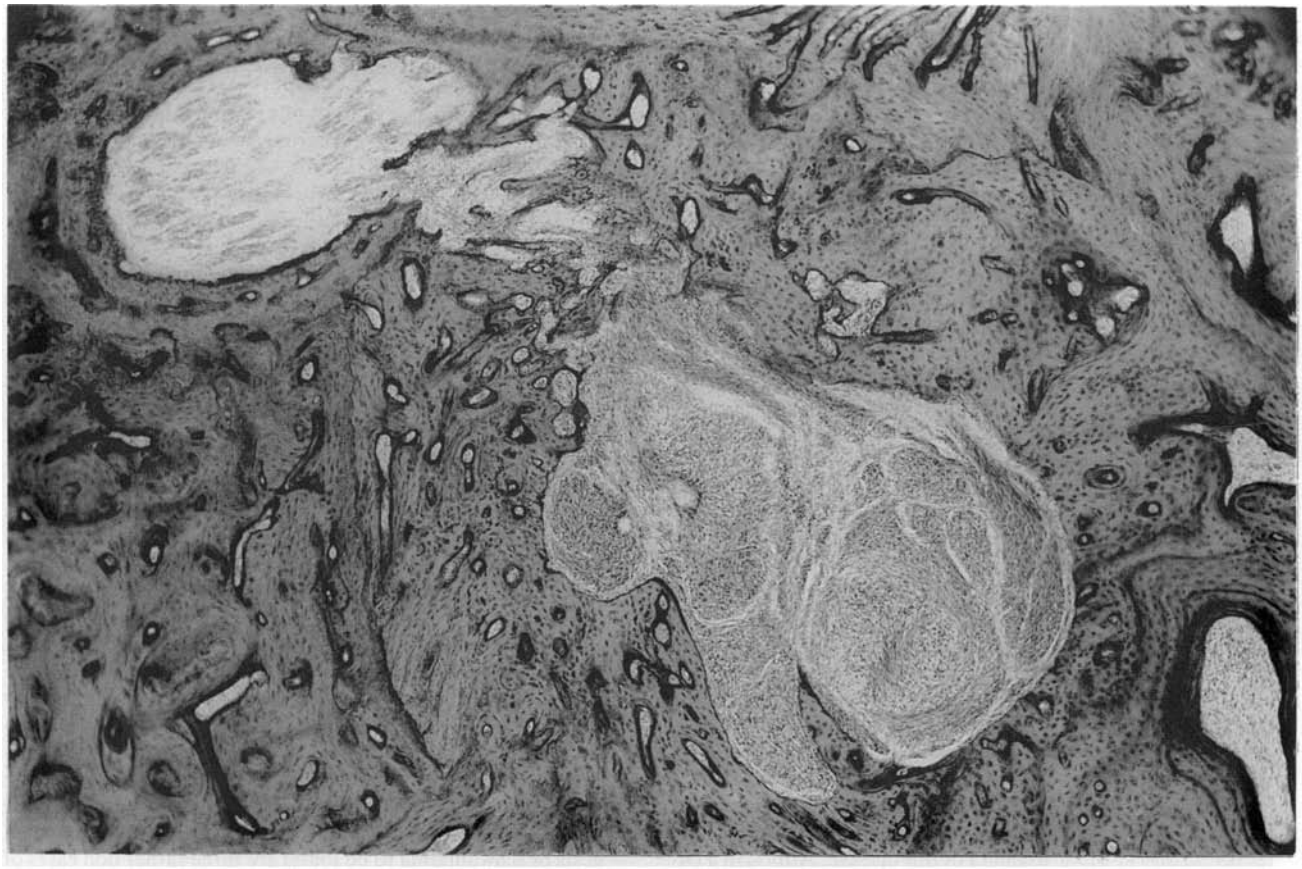

FIG. 2

The tumour as seen in the section in Figure 1. It is in connection with the singular nerve. The typical whorls of an acoustic neuroma are seen. (Original magnification ×15)

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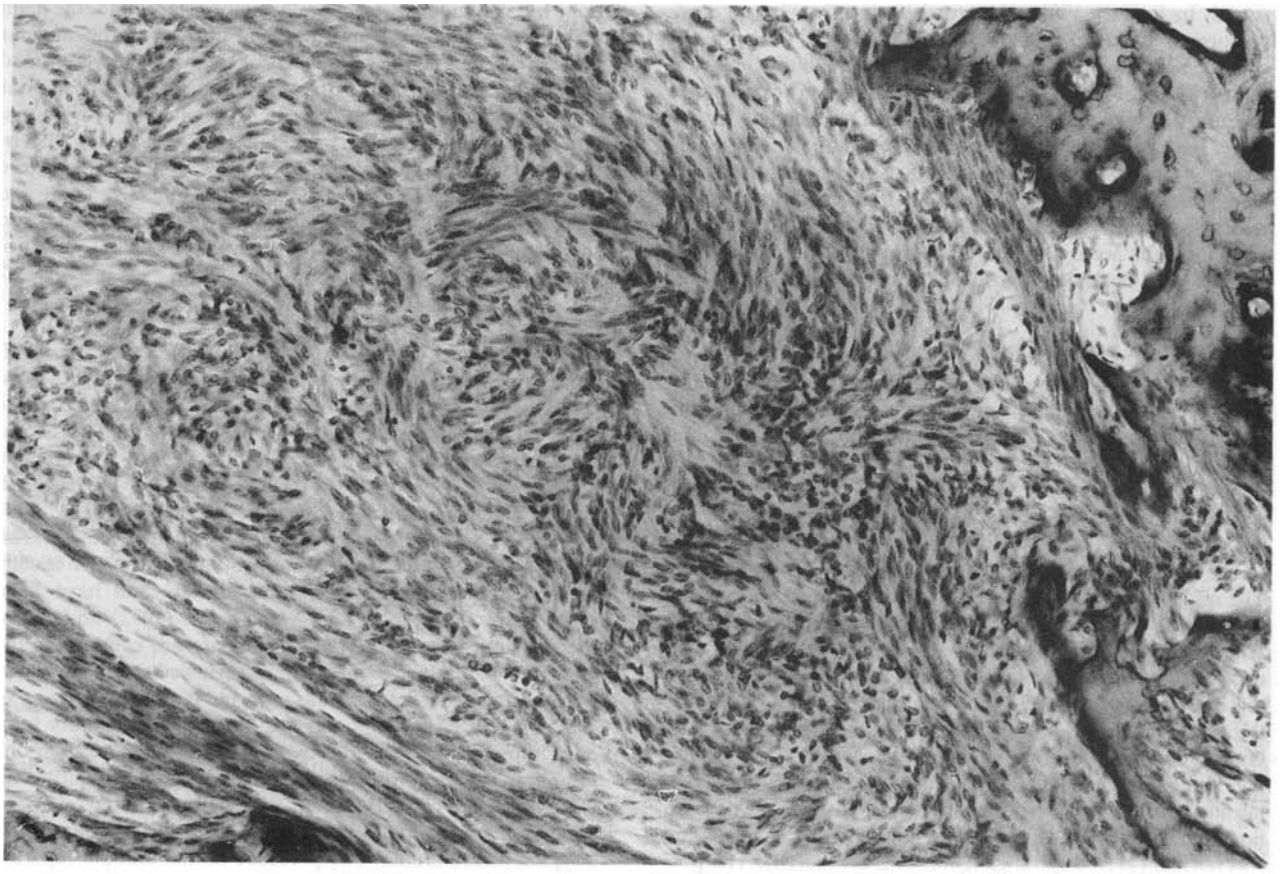

FIG. 3
Close up of the tumour. (Original magnification ×64)

devoid of nerves, is incompatible with the diagnosis of an acoustic schwannoma. The patient is also very young for this diagnosis.

Traumatic neuromas of the vestibular nerves are recognized following surgical labyrinthectomy. Hilding and House (1965), Pulec (1974), Linthicum *et al.* (1979) and Ylikoski and Belal (1981) have all reported cases. They presumably can also occur after bacterial damage to the labyrinth. The over zealous reparative attempts of a peripheral nerve that lead to a traumatic neuroma are well documented by Weller and Cevros-Navarro (1977) who describe a gradual development for many years after injury. The nature of vestibular traumatic neuromas is less well defined. Findings in the labyrinth that have been called traumatic neuromas by the above authors include: multiple small neuromas, single small neuromas, single masses of Schwann cells filling the vestibule, and less discrete masses of tissue filling the labyrinth consisting of intertwined nerve fibres. Not all nerve proliferations within the labyrinth following trauma have been considered traumatic neuroma. Schuknecht (1982) reported a case three months following labyrinthectomy in which bundles of interlacing nerve fibres were obscured in the supporting stromata of the utricular macula and cristae ampullares. He did not consider this to be a traumatic neuroma. Given this uncertainty about the nature of, or even the existence of, vestibular nerve traumatic neuroma, the diagnosis is hard to make or exclude in our case.

Multiple intratemporal tumours are more characteristic of neurofibromatosis type 2. Nager (1969) illustrated this with a case possessing many individual tumours including one in the cochlea and another in the vestibule. The present case had no family history or other stigmata of this disease. Although a circumscribed schwannoma can exist within a neurofibroma, the pattern of separate cell nests in the vestibule is not that of neurofibromatosis. The singular nerve has not been reported as, and would be unlikely as, an isolated site for neurofibromatosis.

The present tumour does not fit into any one of these categories, nor into the many other rare diagnoses that were considered such as cranial fasciitis which could have caused a lesion of not dissimilar histological pattern (Lauer and Enzinger, 1980). The one case with certain similarities that has been reported previously is from a study by Stewart *et al.* (1975) who looked for occult acoustic schwannomas of the vestibular nerve in the temporal bone collection at the Massachusetts Eye and Ear Infirmary. Of the five cases, one in an 84-year-old man had two tumours, one of the superior vestibular nerve and one of the singular nerve. Importantly, there were also some small nests of tumour cells in the Scarpa's ganglion and in the saccular macula. This case had received external beam radiotherapy twelve years previously with subsequent extensive osteoradionecrosis of the temporal bone. There was no other clinical evidence of neurofibromatosis and the vestibular nerves in this case had degenerated. This case from the Massachusetts Eye and Ear Infirmary and our own have certain factors in common. There is a significant history of trauma in both; in the Massachusett's case from external beam irradiation with subsequent osteoradionecrosis and in our own from labyrinthitis secondary to meningitis. They both have tumours characteristic of an acoustic schwannoma in the singular nerve. The most unusual finding that they have in common is of cell nests of nerve tumour in the labyrinth. We have a situation where there are the appearances of both an acoustic schwannoma of the singular nerve and a number of traumatic neruoma occurring together. The only other reports of small nests of schwannoma to be found are three rather odd cases of intracanalicular acoustic tumours described by Kasantikul *et al.*, in 1980, all of whom presented with a pre-operative facial palsy. These are described as having prominent dilated vas-

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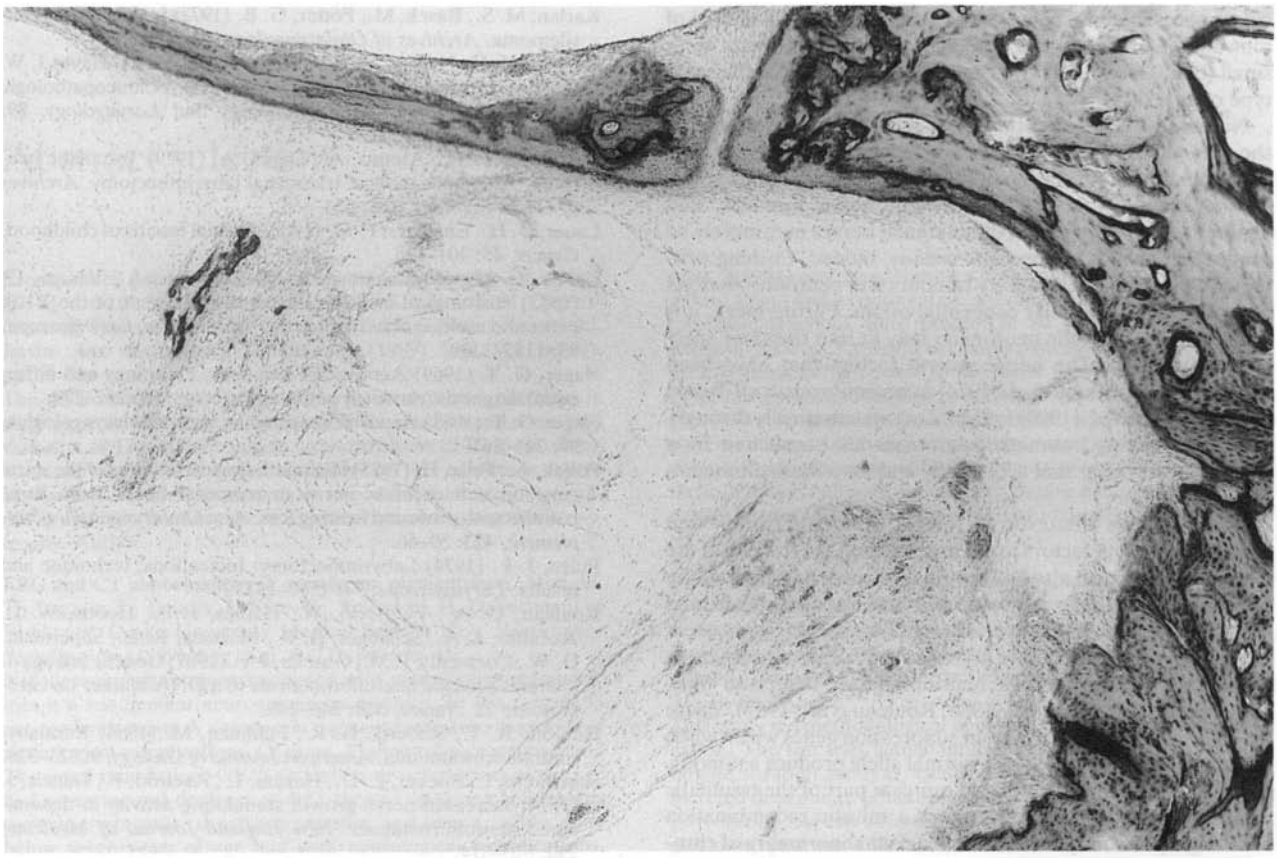

FIG. 4a

Some of the multiple nests of tumour cells in the vestibulum. The stapes foot plate is in the upper left part of the picture. (Original magnification ×15)

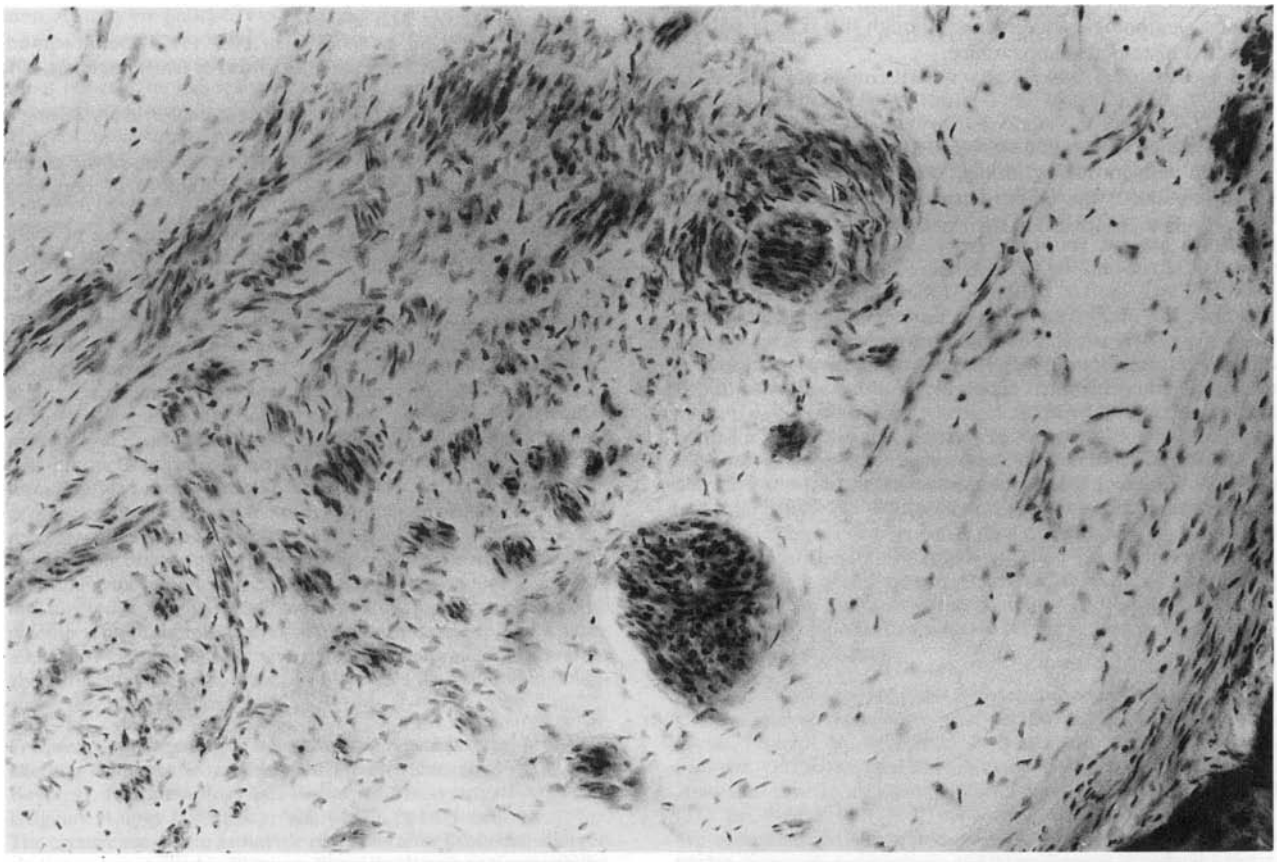

FIG. 4b

Close up of some of the multiple nests of tumour cells in the vestibulum. (Original magnification ×60)

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cular spaces in dense connective tissue with a few small foci of schwannoma close to the vascular walls. The tumours were small (0.5, 0.6 and 1.0 cm). They do not appear to be the same type of case as ours.

Nager, in 1985, stated that there is a general acceptance of the idea that bilateral eight nerve tumours are but a monosymptomatic manifestation of neurofibromatosis and that it is possible that solitary, bilateral eight nerve tumours, and generalized involvement of other cranial nerves may merely be graduation of one and the same malady. Indeed, Cushing writing in 1932 considered solitary tumours and neurofibromatosis to be doubtless of similar congenital origin. Furthermore, it is possible that traumatic neuromata may fit into the same spectrum of disease. The nerve growth factors that have been detected by Schenkein *et al.* (1974) in neurofibromatosis Type 1 and by Kanter *et al.* (1980) in Type 2 act systematically throughout life, whereas traumatic neuromata are stimulated by a nerve growth factor that acts locally and for a limited time following the injury.

In these cases whatever the events or predisposition that caused the growth factors to act in producing the tumour of the singular nerve, must also have caused cell nests to grow into the labyrinth. In the Massachusettes Eye and Ear case, the factors also found targets in the superior vestibular nerve. From recent developments in genetics we know that both neurofibromatosis Type 2 and solitary acoustic neurinoma have defects in chromosome 22 (Seizinger *et al.*, 1986; Rouleau *et al.*, 1987). These are likely to be recessive tumour suppression genes which when defective and not masked by a normal allele produce a tumour. It is possible to speculate that in our case part of the traumatic neuroma being formed underwent a mitotic recombination allowing a previously formed but masked abnormality of chromosome 22 to take effect, the nerve growth factor present not only stimulating the traumatic neuroma but also an acoustic neurinoma. Such speculation leads onto a possible role for trauma in the aetiology of some acoustic schwannomas.

There is always the alternative possibility of course that two separate pathologies do co-exist, though this is very unlikely from the histological appearance.

Conclusion

The finding of a second case of singular nerve tumour in conjunction with intra-labyrinthine tumour leads to speculation that tumours of the eighth nerve may have some common underlying mechanisms in their pathogenesis.

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