Ramsay-Hunt Syndrome with complete recovery of cranial nerves

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Introduction

The syndrome characterised by facial paralysis associated with hearing loss, dizziness and an eruption of blisters on the auricle due to Herpes Zoster was described by Ramsay Hunt in 1906. This syndrome is now synonymous with Herpes Zoster oticus. Hunt thought that the symptoms resulted from geniculate ganglionitis caused by the Herpes Subsequently Denny Brown (1944) has shown that there is little if any gar glion involvement, though there is definite lymphocytic infiltration of the nerve. However the inflammatory changes per se are not sufficient to cause paralysis. Crabtree (1968) attributes the frequent involvement of the facial nerve to the fact that it travels through a bony canal for almost 30 mm. in the skull. The inflammatory changes cause sufficient swelling, oedema and accumulation of metabolites within the fixed bony canal which results in ischaemic paralysis. The incidence of Ramsay Hunt syndrome varies with different authors from 2% - 9% of all facial palsies (Francisco Antoli-Candela, Jr. et al 1974). There are many similarities between Ramsay Hunt syndrome and Bells palsy in the clinical behaviour, the predictability from electrical tests and the time of recovery, Atkins and Brain (1933), Taverner (1955), Gregg (1961) and Tonita et al (1971). However the paralysis in Herpes Zoster is more likely to be a complete paralysis.

Case Report

L.T.L., 47 year old Chinese male was admitted as a casualty to the E.N.T. Unit, University Hospital

on the 28th May 1974, with severe right otalgia 6 days. He also had fever 4 days, inability to talk clearly and weakness of the right face 4 days and a rash and pruritus in the right ear on admission. He experienced hyperacusis on the right side and complained of loss of taste on the right side of tongue.

He gave a past history of diabetes, but never had chicken pox.

On examination, he was a well built middle aged male. The significant findings were a swelling of the right side of face with loss of right nasolabial fold. He had a vesicular rash on the right concha with scabbing (Fig. 1). There were also vesicles on the external auditory meatus. There was a complete seventh nerve paralysis on the right side (Fig. 2) and loss of taste in the right anterior 2/3 of tongue. There was also decreased lacrimation on the right as tested by Schirmers Test. There was evidence of hearing loss on the right ear (Fig. 3) and complete vestibular paralysis on the right as evidenced in the electronystagmography (Fig. 4). Viral culture for herpes was negative. Electromyography confirmed early changes of muscle degeneration.

The patient was treated with 200 mg. Cortisone daily for 2 days, followed by 100 mg. daily for 2 days, 50 mg. daily for 2 days, 25 mg. daily for 2 days and finally 10 mg. daily for 2 more days. The face was stimulated with galvanic current, daily massaged and exercised. The diabetes was controlled initially with ½ tablet of Tolbutamide while on steroid therapy, and later controlled on a low caloric diet alone.



Fig. 1 Right ear on admission showing the rash on the auricle.



Fig. 2 Face of the patient on admission.

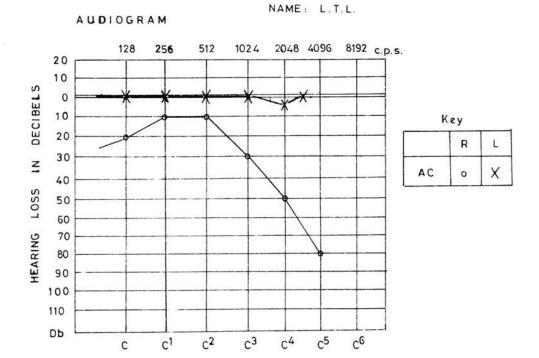


Fig. 3 Audiogram done on 30.5.1974.

The patient was followed up and after eight weeks there was a complete recovery of the face (Fig. 5), hearing (Fig. 6) and vestibular function (Fig. 7).



Fig. 4 Electronystagmograph on 30.5.1974. Right canal paralysis on caloric stimulation.



Fig. 5 Face of patient after 8 weeks.

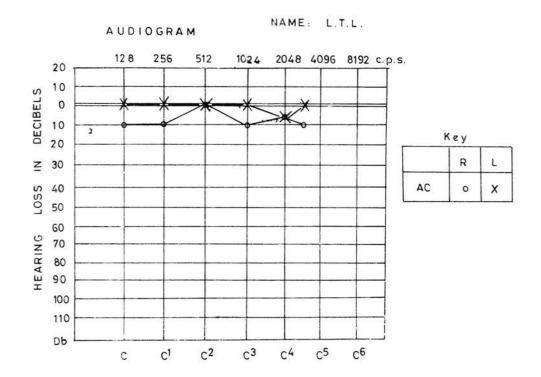


Fig. 6 Audiogram after 8 weeks.

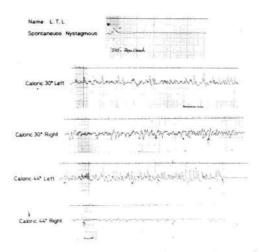


Fig. 7 Electronystagmograph after 8 weeks. Right canal response normal.

Discussion

It is important that all cases of facial paralysis should be accurately evaluated:

Taverner and Adour have shown that Corticosteroids or their precursers (A.C.T.H.) are very effective in the medical management of facial paralysis. The failure in recovery in most cases is because of late detection. Large doses of Cortisone should be given immediately and reduced in decreasing doses for a sufficient period of time. This reduces the oedema of the facial nerve.

Ballance and Dual (1932), and Crabtree (1968) have advocated surgical decompression in certain cases of Herpes Zoster oticus, where electrical tests reveal denervation. If surgical decompression is indicated, the choice of surgical procedures will be indicated by the site of the lesion.

Summary

A case of Ramsay Hunt syndrome with 7th and 8th nerve involvement is described here. Early detection and treatment with large doses of Cortisone had resulted in complete recovery of the cranial nerves within two months.

Acknowledgement

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References

- ADOUR, K.K. (1971) Facial Paralysis, Am. Acad. Ophthal Otolaryng, 76: 1284.
- ATKINS, R.S., and BRAIN, R.T. (1933) Facial palsy and injection with Zoster virus, Lancet, 1: 19.
- BALLANCE, C. and DUAL, A.B. (1932) Operative Treatment of facial palsy, Arch. Otolaryng, 15: 1.
- CRABTREE, J.A. (1968) Herpes Zoster oticus, Laryngoscope, 78: 1853.
- DENNY-BROWN, D. ADAMS R.D. and FITZ-GERALD, P.J. (1944) Pathologic Features of Herpes Zoster, Arch. Neurol. Psych., 51: 216.
- GREGG G. (1961) Some observations on Bell's palsy in Belfast during the period 1949 1958, Arch. Phys. Med., 42: 602.
- HUNT, J.R. (1907) On Herpetic inflammation of the geniculate ganglion: A new syndrome and its complications, J. Nerv. Ment. Dis., 34: 73.
- TAVERNER (1955) Bell's Palsy, Brain, 87: 209.
- TOMIJA, H., HAYAKAWA, W., and HONDO, R. (1972) Vascella Zoster virus in idiopathic facial palsy, Arch. Otolaryng, 95: 364.