CLINICAL RECORDS

BICKERSTAFF'S ENCEPHALITIS

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It is well-known that brain stem lesions may give rise to VIIIth nerve symptoms. The exact ætiology is, however, very often obscure. It is felt that it is of interest to report the following case because of its relative rarity and also because the presentation was very like that of an advanced acoustic neuroma.

Case Report

History: Mrs. A.H., (47), was admitted on *6.5.65*. She had been well apart from a bilateral recurrent otitis externa till 4 weeks prior to admission. She had a few days of non-specific malaise and then she suffered from a left-sided facial palsy which took 2 days to develop, and shortly after this her general practitioner noticed a herpetic lesion on the left cheek. At this time she also lost taste on the left side of the mouth. Two weeks later she suddenly developed double vision and felt a sensation of dysequilibrium for a few moments. One week later she began to have difficulty in swallowing and her relatives noticed that her speech was slurred. Two days prior to admission she suddenly became very dizzy and was confined to bed with this and vomiting. Over the last few weeks she had gradually become more deaf in the left ear.

Physical Examination: On admission she was pale and drowsy and had severe vertigo and vomiting. The pulse was regular and she was normotensive. The fundi were normal with no papillœdema and the pupils reacted normally to light and accommodation. There was a defect of conjugate eye movement and a bilateral VIth nerve palsy more marked on the left side. She had a complete left-sided lower motor neurone palsy of the facial nerve. The sensation on the left side of the face was altered and although the left corneal reflex was absent this was probably due to motor impairment. The left side of the palate did not move and there was difficulty in swallowing. Other cranial nerves were intact. She had signs of cerebellar impairment, i.e. dysmetria in the left hand and dysdiodochokinesis. There was some diminution in motor power on the left side of the body and the plantar response was flexor, but she had intermittent clonus of the left ankle. Her gait was unsteady even with the eyes open and she veered to the left side.

Otoneurological Examination: She had bilateral mild otitis externa but no vesicles were seen. Both drums were intact and the upper respiratory tract was normal. There was a grade 3 nystagmus to the right.

Position tests with electronystagmography revealed mainly pendular waves due to her drowsy state. When aroused, however, she showed nystagmus to the

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right in four of the standard six positions. A curious feature was that the nystagmus with the eyes closed had a speed of 5°/sec. but with the eyes open the speed rose to 11°/sec. The most important feature, however, was that in the upright position, direction changing nystagmus (i.e. to the left and right intermittently) was recorded.

There was no function on the left side with a modified Hallpike caloric and also with the Dundas Grant method.

With full masking there was no hearing at all in the left ear. The hearing in the right ear was normal.

Investigations: Urine, hæmatology and biochemical results were normal as was the Wasserman reaction. X-ray of the chest, skull and petrous bones showed no abnormality. On the electroencephalogram there was a slight right fronto-temporal abnormality. Lumbar puncture showed no abnormality. Lumbar air encephalogram showed no displacement of midline structures of either temporal horn. From the vesicle, herpes simplex was isolated, but other virology studies were negative to mumps S and V, and also to lymphocytic choriomeningitis. No virus was isolated from the cerebro-spinal fluid after three passages in baby hamster kidney tissue culture.

Progress: When first seen, it was thought that this was either a case of acoustic neuroma because of the deafness, canal paresis, direction changing nystagmus, VIIth and Vth nerve signs, or, less likely, a severe Ramsay Hunt syndrome. Craniotomy was not considered since there was no evidence of increased intra-cranial pressure or X-ray changes.

While further investigations were proceeding the patient began to get better.

Further otoneurological examinations on *18.5.65* and *22.5.65* showed that the hearing had recovered to a 40 db. loss in the left ear; no recruitment was present as tested by the Fowler loudness balance test and Békésy audiometry, and the discomfort threshold was not lowered.

The nystagmus with the eyes open ceased and was only recorded behind closed eyes at a speed of some 4°/sec. in four out of the six positions. There was still a small degree of direction changing nystagmus in the upright position. Although there was still a left canal paresis, function was present with a maximum eye speed of 10°/sec. with water at 30° C and 4°/sec. with water at 44° C.

When the patient was sent to convalesce on *1.6.65* the Vth and IXth nerve signs had resolved and the facial palsy was minimal; the dysphagia had cleared, the speech was no longer slurred, the eye signs had all but resolved and there was only slight unsteadiness in the gait.

Due to the fact that this condition recovered so well with the otoneurological findings pointing to a retrocochlear lesion, it was decided that the only thing it could be was a swelling of the myelin sheaths which had subsided. Thus the diagnosis of the benign encephalitis of Bickerstaff was made.

When seen on *21.6.65* the patient had made a complete recovery.

Comment

A syndrome consisting of a prodromal malaise with a downward progression of mid-brain disturbance till almost total suppression of all functions having

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brain stem innervation occurs with no cardiac or respiratory embarrassment was first described by Bickerstaff and Cloake (1951). They described three examples which gradually recovered to normal and suggested that it might be a virus encephalitis.

In 1957, Bickerstaff described the findings in eight further cases, one of which died and had an autopsy. This showed widespread swelling and ballooning of myelin sheaths in the brain stem in the less compact fibre tracts as seen in cerebral œdema. Astrocytes showed "twinning" and bulkiness of cytoplasm which is also seen in cerebral œdema. The nerve cells were not affected apart from an occasional cell in the IIIrd nerve nucleus showing wrinkling of the nuclear membrane and commencing cytolysis. There was diffuse but slight loss of Purkinje cells in the cerebellum but this was probably a *post-mortem* change.

All eight reported cases had drowsiness and headaches, defective conjugate movements, diplopia and nystagmus. The ages varied from 10 to 47 and there were five males and three females. Seven of the eight had total ophthalmoplegia, six of the eight had a total bilateral Vth nerve palsy and eight of the eight had total VIIth nerve palsies. Six of the eight had deafness but this was not enlarged upon and six of the eight had IXth, Xth, XIth and XIIth nerve palsies. Electroencephalography showed abnormalities in both cases tested. Cerebrospinal fluid pressure was normal in all, five of the eight had lymphocytic pleocytosis, four of the eight had a protein of more than 60 mg. per cent. and three of the eight had had a Paretic Lange curve. Complete recovery occurred from 1–17 months.

More recently, Pantazopoulos and Constantinides (1965) described four cases which fitted this syndrome complex but they described the cases as subacute brain stem polioencephalitis and disseminated encephalomyelitis. All these cases recovered.

In view of the fact that the herpes simplex virus was isolated in the described case it would be attractive to blame this virus. The only reported autopsy however, showed none of the perivascular cuffing considered so typical of virus infections of cerebral tissue.

The clinical pattern is somewhat similar to that of infective polyneuritis, the mode of onset, progression, stationary state at maximal disability and rapid recovery of gross paralysis with slower final recovery from all symptoms. Infective polyneuritis is now considered to be an auto-immune disease.

Summary

A case showing most of the characteristics of Bickerstaff's encephalitis is described. The classical clinical picture is mentioned as are the autopsy findings.

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