Interstitial Hypertrophic Neuritis of the Eighth Cranial Nerve

Case Report

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Interstitial hypertrophic neuritis was reported in 1893 by Dejerine and Sottas4 as a chronic progressive neuropathy of childhood. Although no true nerves in the body appear to be exempt from the disease, it primarily affects the extremities and this prompted Austin5 to state in his extensive review of the subject that “in no case is cranial nerve deficit prominent in the total clinical picture . . . .” Thus, it seems worthy to report a case which was confused with an acoustic neuroma and operated on as such.

Case Report

A 24-year-old man was first seen in May, 1962, with a chief complaint of double vision for one week. The diplopia had occurred gradually, was more severe late in the day and was maximal on looking to the left. The patient had had two previous episodes of double vision in 1959 and 1961, identical to his present complaint. These had cleared spontaneously after several weeks. Following an upper respiratory infection at age 16, the patient had first noted ringing and decreased hearing in his right ear which persisted but was not progressive. Recently he had noted clumsiness of his hands, the left more than the right, particularly while carrying out his duties as a typist. In January, 1960, he sustained a period of unconsciousness for about 30 minutes following an auto accident, and at that time the right pupil was noted to be larger than the left. No one in the patient’s family has symptoms suggestive of a similar disease.

Examination. The patient was a stocky, muscular, fair-skinned young man. There was a café au lait spot over the left deltoid but no subcutaneous nodules were felt. There was a complete left 6th nerve palsy and anisocoria with normal response to light. He had a partial right 8th nerve deficit with a 50 decibel high frequency hearing loss maximal at 4,000 cycles per second. There was a diminished response to caloric stimulation on the right. Air conduction was greater than bone conduction on both sides and the Weber test lateralized to the left. Taste was poorly perceived bilaterally. Coordination was intact except for a slight terminal tremor on finger-to-nose testing on the right. The Romberg test was negative. Superficial reflexes were normal but deep reflexes were hypoactive.

A lumbar puncture revealed an opening pressure of 220 mm. of cerebrospinal fluid with a protein of 198 mg. per cent and no cells. The electroencephalogram and plain skull x-rays with Stenver’s views were normal. A satisfactory pneumoencephalogram was normal.

For two months the patient continued to experience tinnitus and decreased hearing in the right ear and clumsiness of the left hand. The 6th nerve paresis improved slightly. A repeat lumbar puncture revealed an opening pressure of 230 mm. of cerebrospinal fluid with a protein of 269 mg. per cent and 2 lymphocytes. Audiogram and calories were unchanged.

Operation. On the basis of a clinical suspicion of a small acoustic neuroma, an exploratory suboccipital craniotomy was carried out in August, 1962. A small tumor measuring 1X.5X.5 cm., was found intrinsically involved in the 8th nerve just as it entered the internal auditory foramen. The tumor was avascular and was removed by sacrificing a portion of the 8th nerve. Histologically, the “tumor” was the type of hypertrophic interstitial neuritis described by Dejerine-Sottas (Fig. 1).

Postoperative Course. The patient had no facial weakness and no change in his 8th nerve function on audiography or on caloric testing. Taste was diminished on
both sides of his tongue and could be perceived only posteriorly. The diplopia cleared over the following 3 months. One year later, the patient was unconscious for 2 hours after an auto accident but suffered no sequelae. In the fall of 1964, he noted the gradual onset of a buzzing in the left ear without any change in his hearing. In March, 1965, he again had an episode of double vision similar to previous episodes, and lasting about one month. A lumbar puncture at that time revealed an opening pressure of 200 mm. of cerebrospinal fluid with a protein of 213 mg. per cent.

When last seen in January, 1966, he was asymptomatic except for tinnitus in both ears and the decreased hearing on the right which was unchanged. He had hypoactive deep tendon reflexes and appreciation of vibratory sensation was diminished in both legs. Nerve conduction studies were carried out and revealed slowed conduction in all nerves tested except for the tibial nerve from the knee to the ankle (Table 1). Caloric test results were unchanged but audiography revealed a 40 decibel loss in the higher frequencies in the left ear for the first time.

Discussion

Except for the early cranial nerve deficits, this case fits the classic description of hypertrophic neuritis as summarized by Austin.5 The disease has multiple nerve involvement, a chronic, progressive and relapsing course, pupillary changes, hyporeflexia, and elevated cerebrospinal fluid protein and pressure. Cranial nerve lesions are unusual in the early course of the disease, but Rossollimo2 reported a case with 6th nerve paresis. The cerebrospinal fluid protein is usually elevated in this disease, but seldom over 100 mg. per cent. Symonds and Blackwood8 reported a case in which the spinal cord was compressed by involved spinal nerves, producing a subarachnoid block. These authors suggested this as a possible cause for marked elevations of the cerebrospinal fluid protein. In our case, the protein has remained elevated over the 2 years of follow up but there is no clinical evidence of a subarachnoid block. Elevated pressure has also been previously noted in this disease, but the mechanism is poorly understood. It has been suggested that the elevated protein of any cause blocks reabsorptive pathways.5

The gross pathologic changes consist of fusiform or beaded swellings on peripheral nerves throughout their course. When viewed microscopically in cross section, individual or small bundles of nerve fibers are encircled by concentric rings of cellular connective tissue, giving rise to the typical onion skin appearance (Fig. 1). The myelin sheath is reduced or absent.6 Slowed conduction has been demonstrated in multiple nerves in patients with hypertrophic neuritis with and without clinical evidence of neuropathy.1

This disease can mimic the clinical picture of multiple sclerosis with diffuse lesions and a remitting course, as this case well demonstrates. There have been reported associations of the two diseases,5 but the significance of this remains to be seen. Hypertrophic neuritis has also been reported in association with neurofibromatosis and has been considered a variant of this disorder.1 A recent report describes the pathological changes of hypertrophic neuritis in 2 cases of acromegaly, suggesting this as a cause for some of the neuropathies associated with it.9 The prognosis of hypertrophic neuritis was discussed by Austin and ranged from a progressive downhill course and death in 12 years in one reported case to a 24-year follow up in another case with very little change.2 Treatment of cases with steroids has been moderately successful in inducing remission.1,3

Summary

We have reported a verified instance of interstitial hypertrophic neuritis of the 8th cranial nerve, originally mistaken for a tumor. We have discussed the clinical and laboratory findings characteristic of this disease, and have reviewed related case reports.

References