CASE REPORTS AND TECHNICAL NOTES

AN UNUSUAL CASE OF UNILATERAL EIGHTH NERVE TUMOR

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The occurrence of bilateral acoustic tumors, or acoustic tumors associated with von Recklinghausen's neurofibromatosis, is well known in the younger age groups.\(^2\),\(^3\) The most authoritative authors on acoustic neuroma,\(^1\),\(^2\),\(^4\) however, emphasize the late age of onset of unilateral acoustic tumors unassociated with other pathological stigmata. Cushing\(^1\) states that "... the symptoms of an acoustic tumor rarely occur before the third decade of life, ... Hence one may be justified in the conclusion that a patient under 20 with a cerebellorontile-angle syndrome in all likelihood has a lesion other than an acoustic tumor." The case of unilateral acoustic nerve tumor presented here, however, proves to be a remarkable exception to that rule, for the age of onset was 6 years and the neuroma was removed and verified histologically when the child was 8½ years old. Furthermore, this patient showed none of the associated stigmata of von Recklinghausen's disease, either by physical examination or family history. Some reports of unilateral acoustic neuroma occurring in the second decade have been presented\(^1\),\(^2\),\(^3\) but none have been reported in the first decade of life.

Additional unusual features of this case include a normal spinal fluid protein and a massive destruction of the involved petrous pyramid (Fig. 1). These features did not alter the outline of therapeutics in this 8½-year-old boy, but they made the probable histological diagnosis of his posterior fossa tumor an intriguing clinical problem.

CASE REPORT

This 8½-year-old white male was admitted to the Boston Floating Hospital on June 10, 1951, and discharged on July 3, 1951.

Late in 1948, he had exhibited a mild transitory left facial weakness, which disappeared completely. In August 1949 there developed a sudden paresis of the left face which was persistent. Late in 1949, the patient noted the gradual onset of deafness in the left ear and he found he could no longer hear a watch tick in that ear. At this time the left eye was observed to close incompletely and the left side of the face seemed full. His symptoms were gradually progressive and in July 1950 he began to experience episodes of vomiting associated with vertigo. In April 1951 he began to deviate to the right while walking, and he tilted his head to the right. He also had trouble in keeping his balance when standing still.

Examination. There were no cutaneous or subcutaneous stigmata of von Recklinghausen's disease detected after thorough clinical investigation. Significant findings included no papilledema or signs of increased intracranial pressure. There was paresis of the left lateral rectus muscle and conjugate deviation to the right or left was poorly maintained. A slow bilateral horizontal nystagmus, with quick component in the direction of gaze, was also present. The corneal reflexes were normal. There was a severe left facial weakness, the forehead and both orbicularis groups being equally involved. Deafness on the left was of the nerve variety. There was no impairment of hearing on the right. The patient perceived painful stimuli in the left
palate, and the tongue and uvula moved in midline. There was slight bilateral unsteadiness in the finger-to-nose test while the heel-to-knee test showed marked overshooting, especially on the left. There was no weakness or sensory deficit. Deep tendon reflexes were equal, though somewhat depressed, bilaterally. Plantars were extensor. The patient would fall either to the right or left in the Romberg position and he walked in a grossly wobbly fashion with his eyes closed.

X-rays of the skull showed a massive destruction of the medial half of the left petrous bone (Fig. 1). Lumbar puncture in the lateral recumbent position showed an initial pressure of 178 mm. with clear CSF. There were no cells; the colloidal gold curve was normal; total protein was 31 mg./100 cc.

Operation. On June 15, 1951, the left side of the posterior fossa was explored through an inverted U-shaped incision. The left cerebellopontine angle contained an encapsulated tumor roughly the size of a small lemon. It was situated quite laterally, eroding and infiltrating the petrous pyramid. The mass extended superiorly to the tentorial notch and seemed to compress the...

Fig. 1. Roentgenogram showing massive destruction of medial half of left petrous bone.

Fig. 2. Photomicrograph of acoustic neuroma removed.