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,1,3,4 however, emphasize the late age of onset of unilateral acoustic tumors unassociated with other pathological stigmata. Cushing1 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 cerebellopontile-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 presented1,3,4 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 therapeusis 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

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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.
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5th cranial nerve. A piece of the tumor was removed for rapid section diagnosis and was found to be acoustic neuroma. The tumor and its capsule were removed in a piecemeal fashion until only the rostral and lateral parts of the capsule remained. Separation of the capsule from the side of the brain stem was unusually simple for a tumor of this size. The anterior portion was then carefully dissected away from the 5th cranial nerve, leaving that structure intact. The lateral portion invading the bone was then completely removed. The 7th and 8th nerves could not be identified with certainty during the dissection.

Histological Diagnosis. Permanent sections were reviewed by Dr. Raymond D. Adams, and the classical picture of acoustic neuroma was again reported (Fig. 2).

Postoperative course was uneventful. Examination 3 months later revealed an active boy who ate and slept well and who talked in an almost normal tone. Nightmares suffered preoperatively had disappeared. Left nerve deafness and complete left facial paralysis were the only deficits present. Tests of coordination and cerebellar function were within normal range, and the Romberg sign was negative.

DISCUSSION

The size and position of this tumor are of some interest. While Cushing drew attention to some unusual tumors 7 cm. in length, the ordinary large neuromas seldom reach this length and the tumor described in this report could certainly be considered as an example of a large tumor. The usual course of these tumors is prolonged and it is not unreasonable to believe that the inception of tumor growth preceded, by some time, the onset of symptoms in 1948 or 1949. This would place the date of inception of tumor growth before this child's 6th year of life.

The normal spinal fluid protein may be related to the extreme lateral position of the tumor. A good bit of tumor was found disrupting the petrous bone and a comparatively small amount of neuroma was seen to be protruding into the subarachnoid space of the angle. Tumor did not extend through the notch and a complete removal of the neoplasm was surprisingly easy.

CONCLUSION

An unusual case of unilateral acoustic neuroma is presented, occurring in an 83/4-year-old boy without stigmata of von Recklinghausen's disease.

REFERENCES