CLINICAL OBSERVATIONS ON TUMORS OF THE PINEAL REGION

JIRO SUZUKI, M.D., TOKUO WADA, M.D., AND MASAYOSHI KOWADA, M.D.

Hind Surgical Clinic, Tohoku University, Sendai, Japan

(Received for publication September 25, 1961)

SINCE Blaé's²⁵ original report, tumors of the pineal region have presented a fascinating challenge in diagnosis,¹⁷ pathology,¹⁶,¹⁸ physiology,⁵,¹³,²¹ and therapy.²,⁴,⁸,²¹ In most areas, as Cushing⁸ stated, "... they are so few in number and so variable in their histological composition that their life history and the possible relation of any one type to the condition known as 'pubertas praecox' is imperfectly understood." However, as previously reported,¹⁴,²¹ these tumors occur relatively frequently in Japan. The present report concerns the clinical features, therapy and results in 24 such cases. The pathological classification was: pinealoma, 15; "double" tumor, 5; teratoma, 2; astrocytoma, 1; and meningioma, 1.

As often noted,¹ the greatest incidence is in males under the age of 25 (Fig. 1). In this series, 21 of the patients were males and were under 25 years of age. The oldest patient, a woman with meningioma, was aged 31 years. The initial symptom was headache in 21 cases, usually associated with nausea and vomiting. Other complaints were: diplopia, 22; visual disturbances, 15; tinnitus, 9; dysacusis, 8; and vertigo, 8. The duration of symptoms was less than 6 months in 14 cases and was over a year in only 2 instances, the longest being 26 months.

Examination revealed papilledema in 21 cases, with secondary optic atrophy in an additional case. Limitation of upward gaze (Parinaud's syndrome) was noted in 16 patients, half of these having absent pupillary reactions. Some form of disturbance of the 3rd cranial nerve was found in 18 patients, in all but 2 being bilateral. By contrast, the trochlear nerve was involved in only 4 instances, and the abducens in 2. The 7th and 8th cranial nerves were affected in 4 and 7 cases respectively, and the 5th in 5. Nystagmus of various types was present in 9 instances. Ataxia was evident in 16 cases, of cerebellar type in 6. In 7 instances there were sensory disturbances, usually lateralized. Hemiplegia occurred only twice but abnormal Babinski's responses appeared in 5 cases and hyperreflexia with clonus in 8.

Disturbances in the psychic and mental spheres were not uncommon. Clouding of consciousness occurred in 7 cases, usually episodic in relation to severe headaches. Evidences of excitability and impatience were found in 10 patients, while 4 became dull and apathetic. Metabolic abnormalities were of various types, both obesity and emaciation being observed; the latter was especially striking in 8 instances. Polyuria and frequency were not unusual. Pubertas praecox was present in 2 patients while in a third similar changes developed following removal of an astrocytoma in the pineal region. Vascular hypotension below 100 mm. Hg systolic was noted in 3 instances.
TREATMENT

The first documented surgical approach to a pineal tumor was made in 1913 by Krause through the subtentorial space without success. During the past 40 years the usual procedure has been either that of Dandy, through the splenium of the corpus callosum, or the transventricular approach of Van Wagenen. Horrax, in 1937, reported a successful removal of a 70-gram pinealoma by a modification of the latter method. More recently the trend has been toward radiation therapy after some form of ventricular shunt, because of the high mortality associated with attempts at surgical excision. Assessing the operative hazards, it is apparent that they fall into three categories: preparation, the operation itself, and postoperative management. The problems, and our methods toward their solution, will be considered under the above headings.

Preparation is directed primarily toward early reduction of increased intracranial pressure, which was present in 22 of the 24 cases. This is accomplished by controlled ventricular drainage over a period of 4 or 5 days, with appropriate antibiotic protection. Since tissue swelling may be present in the tumor-bearing area, cortisone is given daily in amounts of 100–150 mg. Many of the

Spinal-fluid findings usually were confirmatory of increased intracranial pressure, only 2 patients having pressures of 150 or less. Figures between 150 and 300 were obtained in 7 instances, 300 to 450 in 5, and over 450 in 11. The fluid was bloody and xanthochromic in only 1 case, that of a tumor infiltrating the ventricle. The protein was elevated by the Pandy test in 16 cases.

Electroencephalography was done in 15 instances with only 2 normal results. The others showed frequent slow waves, usually of high voltage and generalized, except in 4 patients who presented lateralizing neurological signs.

Roentgenography disclosed calcification in the pineal region in 15 instances, including a teratoma containing teeth and cartilage (Fig. 2). Spreading of sutures and increased convolutional markings were present in 4 patients under 10 years of age. Ventriculography revealed symmetrical hydrocephalus and a defect in the pineal region of the 3rd ventricle in all but 1 case (Fig. 3). Slight displacement or irregular enlargement of the adjacent lateral ventricles were noted in some instances of infiltrative tumors. Angiography supplied valuable information concerning collateral venous channels for the veins of Galen in some cases.