8. Granular Cell Tumors of the Neurohypophysis

Granular cell tumors (myoblastomas) are found in a wide variety of cutaneous, oral, and visceral sites (for review see Moscovic and Azar, 1967; Strong et al., 1970). In the nervous system they have been found in the peripheral nerves (Bangle, 1953; Buldzolovich, 1968; Carstens, 1970; Fust and Custer, 1948, 1949; Garancis et al., 1970), the cervical spinal leptomeninges, the cerebral hemispheres (Markesbery et al., 1973), and the pituitary. Two examples of metastatic, intracerebral neoplasms believed to have been granular cell tumors have also been recorded (Meredith et al., 1958; Schwidde et al., 1951).—Most often granular cell tumors of the posterior lobe or of the pituitary stalk are encountered incidentally at autopsy as microscopic nodules of large, pale, granular cells. A frequency of 1.8 to 17% has been reported (Buston et al., 1962; Hamperl, 1937; Harland, 1956; Kiyono, 1926; Löffler, 1929; Luse and Kernohan, 1955a; Mink et al., 1955; Popovitch et al., 1970; Priessl, 1922; Rap and Zarsaka, 1970; Shanklin, 1947, 1953; Simonds and Brandes, 1925; Sternberg, 1921). Occasionally they become large and produce clinical symptoms. The first such case has been described by Lüthi and Klingler (1951). Since then additional patients have been reported making up a total of 21 cases (Bara and Lantos, 1968; Burston et al., 1962; Daron et al., 1956; Glazer et al., 1956; Harland, 1956; Jenevein, 1964; Iliescu, 1969; Korbine and Ross, 1973; Lima et al., 1960; Liss and Kahn, 1958; Mink et al., 1955; Poppen and Packard, 1966; Rubinstein, 1972; Satyamurti and Huntingdon, 1972; Sekino et al., 1969; Symon et al., 1971; Talerman and Dawson-Butterworth, 1966; Ulrich et al., 1974).

In spite of extensive clinical and pathological investigation, including histochemical and electron microscopic studies, the histogenesis and true nature of the condition remain unknown. Theories include a non-neoplastic product of degeneration and regeneration (Ewing, 1940; Pour et al., 1973; Whitten, 1968; Willis, 1960); a storage or metabolic disorder involving histiocytes (Azzopardi, 1956; Baraf and Bender, 1964; Gray and Gruenfeld, 1937; Leroux and Delarre, 1939; Shear, 1960); and a neoplasm with disturbance of metabolism of myogenic origin (Abrikossoff, 1926, 1931; Christ and Ozzello, 1971; Klemperer, 1934; Krieg, 1962; Murphy et al., 1949; Murray, 1951), of neural origin (Alkek et al., 1968; Ashburn and Rodger, 1952; Bangle, 1953; Carstens, 1970; Caputo et al., 1972; Fisher and Wechsler, 1962; Fust and Custer, 1948, 1949; Garancis et al., 1970, Sobel et al., 1971, 1973b), of fibroblastic origin (Coggins, 1952; Pearse, 1950), or of mesenchymal origin (Moscovic et al., 1967; Sobel et al., 1973a).

We have examined material of two cases. The tissue was removed...
during surgery from the first and obtained at autopsy from the second. The material was fixed in unbuffered formalin in both cases before osmium fixation and plastic embedding were carried out. Both cases have been published separately.—The first patient (Ulrich et al., 1974) (no. 10/72) was a 35 year old female suffering since the age of 33 years from secondary amenorrhea and recently from a rapidly progressing organic psychosis. The neurologic examination also demonstrated a bitemporal hemianopsia. The gonadotrophin excretion was normal in spite of amenorrhea. The basal metabolic rate was decreased (−16%). The X-ray examination showed a normal sella and a suprasellar space occupying mass with compression of the third ventricle during pneumoencephalography. The CSF contained an increased protein content of 55 mg/100 ml. The solid, soft, well vascularized tumor was removed during surgery through a transventricular approach. The postoperative course was characterized by several epileptic seizures, diabetes insipidus, central hyperthermia, and electrolyte disturbances which could be controlled by appropriate fluid infusions. Three months after surgery she suffered again from an status epilepticus and died of cardiac arrest during bronchoscopy which was performed because of suspected aspiration. The autopsy showed a communicating hydrocephalus with a large opening in the floor of the third ventricle. There was no residual tumor. The pituitary was inconspicuous.

The second patient (no. 463/72) was the first case with a space occupying granular cell tumor published in the literature (Lüthi and Klingler, 1951). He survived surgery for 22 years. The detailed autopsy findings will be published elsewhere (Wegmann and Landolt, 1975).—In 1950 at the age of 34 years the patient originally was admitted to the neurosurgical clinic because of increased fatigue and slowly increasing visual disturbances. The clinical examination showed a severe visual loss on the left side with an atrophic optic papilla and bitemporal hemianopsia. There were no endocrinological symptoms. The skull X-rays showed an enlarged sella with a partially destroyed dorsum. An intrasellar tumor with suprasellar extension could be demonstrated with pneumoencephalography. The CSF protein content was reported to be normal. The solid, well vascularized tumor was removed on a transfrontal approach. After surgery the patient suffered from an pituitary insufficiency which made a full, lifelong substitution therapy with cortisone and thyroxine necessary. The postoperative diabetes insipidus of moderate severity (urine production up to 4,500 ml) disappeared spontaneously after 6 months. The patient needed psychiatric care since 1956 because of a recurrent psychosis with suicidal tendencies. In spite of this the patient was able to work as an unskilled worker performing only light work. His strength decreased somewhat in the last months of his life. He died of