Intrasellar Germinomas: A Form of Ectopic Pinealoma*

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Tumors histologically identical to the seminomas and dysgerminomas of the gonads occur intracranially and are commonly known as “atypical teratoma” or “germinoma.”6,9,11 These tumors are also often designated as “pinealomas” because of their frequent occurrence in the pineal region and their apparent histological resemblance to the fetal pineal glands at some stage of development.5 Despite the fact that neoplasms of the pineal parenchymal cells, although rare, are morphologically distinct from the germinomas.6,9,11 Similar tumors, when they occur in locations other than the pineal gland, mostly in the chiasmal region, are referred to as “ectopic pinealomas” and have been defined as a distinct group from the clinical as well as therapeutic points of view.5,10 Germinomas arising in the hypothalamic region constitute less than a third of the total intracranial tumors of similar nature3 and are usually limited to the suprasellar region about the optic chiasm with invasion of the neighboring structures to variable extents.14 Extension of the tumor to the pituitary fossa resulting in radiologically detectable enlargement of the sella turcica occurs only on rare occasions.5,10 In fact, the lack of sellar enlargement has been considered one of the features distinguishing hypothalamic germinomas from pituitary tumors.7 Detailed pathological studies of the intrasellar involvement by germinomas are rare; most reports deal with cases in which this involvement was observed during surgery.1,6,13-15

We are reporting two cases of germinoma in the region of the hypothalamus and sella turcica, studied at autopsy. In each instance the tumor occupied most of the sella turcica, which was markedly expanded, and involved the hypothalamus and third ventricle. Both neoplasms closely resembled a pituitary

tumor clinically as well as on macroscopic examination.

Case Reports

Case 1. A 19-year-old boy began having visual difficulties accompanied by transient episodes of frontal headaches in September, 1965. Neurological examination at that time revealed no abnormality except for a questionable weakness of the right sixth cranial nerve. No visual field defect was present. He remained stable until the summer of 1966 when his vision began to fail rapidly; within 3 months he was unable to read. In addition, he complained of fatigue, loss of hair, and chronic constipation. He underwent various endocrine function tests and was found to be suffering from panhypopituitarism. At that time, he was blind in the right eye and had a left temporal hemianopia. No other neurological abnormality was present. Skull x-ray films revealed a markedly enlarged sella turcica.

Operation. In October, 1966, the patient underwent a right frontal craniotomy; a large tumor was seen involving the pituitary fossa and hypothalamus. Biopsy of the tumor disclosed a germinoma whose details are given below. The patient never regained consciousness and died 8 days after the operation.

Autopsy findings. The unfixed brain weighed 1500 gm. The right cerebral hemisphere was swollen. A large fleshy tumor was present in the retrochiasmal region extending posteriorly to the interpeduncular fossa (Fig. 1). The tumor displaced the optic chiasm anterosuperiorly and completely obscured the structures in the floor of the third ventricle. Inferiorly, the tumor occupied most of the sella turcica which was enlarged in all dimensions. A portion of the pituitary gland was grossly recognizable as a thin rim of brownish tissue compressed by the tumor against the sellar floor (Fig. 2). The tumor extended to the cavernous sinus bilaterally. Serial coronal sections of the

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Fig. 1. Case 1. Germinoma in the retrochiasmal region extending to the interpeduncular fossa. The optic nerves are infiltrated by the tumor (*black arrow*). The hemorrhagic area represents the site of biopsy (*white arrow*).

cerebrum disclosed invasion of the anterior two-thirds of the third ventricle by the tumor (Fig. 3). Aside from an area of fresh hemorrhage at the site of biopsy, the tumor was soft, and grayish in color with small areas of necrosis. A large fresh infarct with breakdown of tissue was seen in the region of the right basal ganglia (Fig. 3). The pineal......