Evolution of a primary intrasellar germinomatous teratoma into a choriocarcinoma

Case report

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A case of intrasellar teratoma with a germinal structure in a 10-year-old girl is described. A few months after intracranial surgery the tumor differentiated into a choriocarcinoma and finally spread to multiple cerebral, pulmonary, and renal metastases. In the course of choriocarcinomatous evolution, very high urinary levels of luteinizing gonadotropin (HCG) developed, but there was no clinical or anatomical evidence of precocious puberty.

KEYWORDS □ teratoma □ choriocarcinoma □ metastasis

TUMORS with a dysgerminal or seminomatous structure are sometimes, although very rarely, found within the skull in the sites preferred by teratomas; that is, in the pineal region and in the posterior portion of the third ventricle,1,5,6 in the anterior half of the third ventricle and the infundibulochiasmal region,1,5,6,8 and even more rarely within the sella.2,4,6 Suprasellar and sellar varieties are usually called "ectopic pinealomas," but this term is controversial; Russell,6,7 in particular, regards these tumors and even anisomorphic pinealomas of the epiphysial region as atypical teratomas.

The case we are reporting is interesting because of the rarity of the tumor's intrasellar site and because we were able to follow its biological and pathological evolution from a teratoma with dysgerminal features into a choriocarcinoma; the unusual endocrine features of the case are also worth discussing.

Case Report

This 10-year-old girl was admitted on January 20, 1968, because of a 2-year history of progressively severe polydipsia and polyuria. In the 2 months preceding admission, visual acuity in the left eye declined steadily, and vomiting and severe headache developed 5 days before admission.

Examination. The child's general condition was poor. Visual acuity was 1/10 in the left eye and 10/10 in the right; she had left temporal hemianopia and an upper temporal quadrant defect in the right eye. Both optic discs were pale with sharp margins and turgid veins. The lumbar cerebrospinal fluid (CSF), electroencephalogram (EEG), and chest films were all normal. The sella was large and its walls thinned. A pneumoencephalogram done in steps by lumbar route showed the pattern of a sellar tumor with modest suprasellar...
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Fig. 1. Left: Photomicrograph of the biopsy specimen taken at operation shows on the right a typical pinealomatous structure (alveoles of large polygonal cells, separated by thin fibrillary bands with lymphocyte-like cells). On the left, tubular cavities lined by tall columnar cells are visible. H & E, × 175. Right: Autopsy specimen of the intrasellar recurrence shows irregularly anastomosed strings of cells presenting the characteristics of both cytotrophoblast and syncytiotrophoblast. H & E, × 250.

growth. The lumbar CSF on this occasion contained 116 cells/mm³, and the Pandy test was strongly positive.

Operation. Right frontal craniotomy revealed a tumor originating in the sella and expanding above it, thus protruding between the optic nerves. The tumor, which was very hard and about 1.5 cm in diameter, was removed piecemeal but totally.

Histological Examination. There was diffuse proliferation of large polygonal cells, grouped for the most part in alveoles, and separated by thin fibrillar bands in which there was abundant infiltration of small cells. The cells were relatively monomorphous with scant clear cytoplasm and spherical nucleus; mitoses and monstrous tri- and binucleate cells were fairly frequent. At some points tubular cavities coated with tall columnar cells recalling respiratory or gastrointestinal epithelium were clearly visible (Fig. 1 left). The histopathological diagnosis was teratoma with germinomatous features.

The postoperative course was uneventful. On discharge the patient still had marked diabetes insipidus, and the visual acuity on the left was now 4/10.

Second Admission. Forty-five days after the operation the patient was readmitted in a state of extreme emaciation and asthenia, with paralysis of the right third cranial nerve, deficits of the sixth nerve, and of the first division of the fifth nerve on the same side. The most interesting abnormal data emerged from the radioimmuno- and bio- assays of the urinary gonadotropins. Although the patient was prepubertal and hypophysectomized, repeated determinations disclosed surprisingly high values of a luteinizing hormone (LH)-like substance, which on closer examination proved to be human chorionic gonadotropin (HCG). Initially, in February, the luteinizing gonadotropin HCG values had been borderline, but by mid-May the values ranged from 80,000 to 100,000 IU/24 hours with occasional peaks of over 200,000 to 300,000 IU/24 hours. The average follicle-stimulating hormone (FSH) concentrations were 28 to 30 IU/24 hours, with peaks of 40 and over. This high level of gonadotropin activity was accompanied by marked depression of all the other hormonal activities, as shown by the following laboratory data: urinary 17-ketosteroids: 0.56 to 0.76 mg/24 hrs; urinary 17-hydroxycorticosteroids 2.7 to 3.9 mg/24 hrs; urinary pregnandiol: 0.15 mg/24 hrs; estrone: 1.2 mg/24 hrs; estriol 2.2 mg/24 hrs.

Two completely negative chest x-ray films.
were followed by others revealing multiple pulmonary nodules that increased in size as the level of HCG in the urine rose. The patient's condition grew steadily worse until she died 5 months after operation.

Postmortem Examination. Necropsy revealed an extensive recurrence of the intrasellar and suprasellar tumor with diffuse erosion of the base of the skull. There were multiple encephalic metastases with evidence of necrosis and hemorrhage. The pineal gland was intact both on gross and microscopic examination. Micronodular metastases like those in the brain were identified in both lungs, liver, and left kidney. Inspection of the genital organs and breasts revealed no sign of puberty; this was confirmed by histological examination of the endometrium and ovaries.

Histological examination of the intrasellar recurrence revealed a pattern typical of choriocarcinoma. Irregularly anastomosed strings of cells presented the characteristics of both cytotrophoblast and of syncytiotrophoblast; the cells were extremely polymorphous with frequent karyokineses, many of them also atypical (Fig. 1 right). At the margin of the neoplastic nodule there was a residue of hypophyseal gland. This histological pattern thus differed greatly from that of the biopsy, both because of the absence of seminomatous and organoid structures and because of its malignant evolution.

Discussion

Germinomas of the diencephalic-chiasmal region, "ectopic pinealomas," account for less than a third of all intracranial tumors of this nature. In about one-fifth of these cases there is also involvement of the sella turcica, which appears abnormally large both on skull films and at operation. This sellar involvement in "ectopic pinealomas" is regarded as secondary. Three well-documented cases in which a tumor of this type was intra- and suprasellar do not disclose whether the site of origin was intrasellar or diencephalic. All three had uniform histological features of anisomorphous pinealomas or germinomas, with no teratomatous features.

In the case of Russell the pineal body contained a dermoid cyst; in the two cases of Ghatak, et al., the pineal body was macro- and microscopically intact.

The only case in the literature comparable with the one we have described of primary intrasellar germinoma is Case 4 in the series of Kageyama and Belsky. The points in common with our case are the patient's prepubertal age, the 1- to 2-year history, symptoms that gained fatal momentum only in the last few weeks, moderately enlarged sella turcica on skull films, necrotic-hemorrhagic tumor entirely within the sella, and terminal metastatic spread. The histology was also similar, with teratomatous, pinealomatous, and choriocarcinomatous types of tissue discovered in both cases. In our case the choriocarcinomatous tissue appeared only later, coinciding with the rapid clinical deterioration. The two cases differed in sex, signs of precocious puberty (absent in our case), gonadotropin levels and pineal characters.

A point worth emphasizing in our case was the absence of precocious puberty in spite of the very high HCG levels; this may have been due either to the small quantity of FSH produced or to unresponsiveness of the target organs.

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References


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