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## Radiology of Primary Intracranial Yolk-Sac (Endodermal Sinus) Tumors

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Most primary intracranial germ-cell tumors are germinomas and teratomas. Less frequent are embryonal carcinomas, choriocarcinomas, and yolk-sac (endodermal sinus) tumors in various admixtures and degrees of purity. Teilum [1] first used the term *endodermal sinus tumor* in 1959 to describe those tumors (whether ovarian, testicular, or extra-gonadal) that originate in primitive germ cells of extraembryonic origin and demonstrate selective overgrowth of yolk-sac elements. The reported incidence of intracranial germ-cell tumors ranges from 0.4% to over 3.4% of intracranial neoplasms [2]. From a review of two pure and three mixed cases of endodermal sinus tumor out of 4,000 intracranial tumors, Albrechtsen et al. [3] concluded that intracranial yolk-sac carcinoma is a true pathologic entity expected to constitute over 0.1% of intracranial tumors and about 10%

of those situated in the midline about the pineal and hypothalamic region.

Of the 14 previous reports of 17 pure yolk-sac tumors found in the world literature, none has appeared in the radiologic literature [3–16]. The purpose of the present communication is to call these rare tumors to the attention of radiologists, with emphasis on a brief review of their neuroradiologic manifestations, and to report an additional, very rare case presenting in the subependymal lateral ventricular and suprasellar area.

### Case Report

A 16-year-old girl was evaluated in December 1979 for long-standing short stature and delayed sexual development. Physical

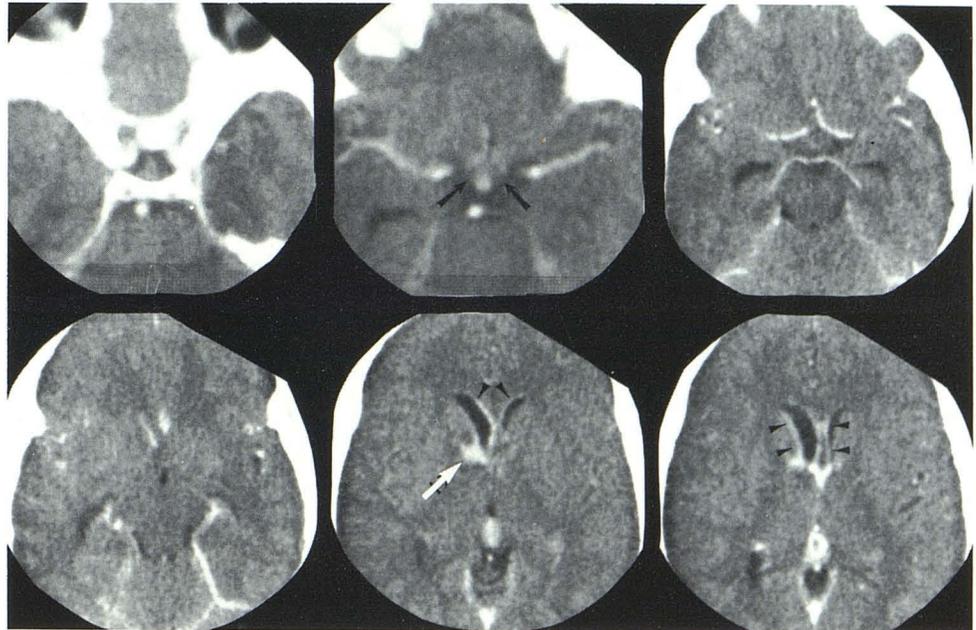


Fig. 1.—Initial contrast-enhanced CT scans. Small mass in suprasellar cistern (*black arrows*). Second contrast-enhancing lesion, without demonstrable contiguous extension, in right caudate region (*white arrow*). Pathologic subependymal contrast enhancement of frontal horns on right more than left (*arrowheads*). These lesions were isodense on noncontrast study. Somewhat prominent enhancement of pineal region was unchanged during 10 months of follow-up.

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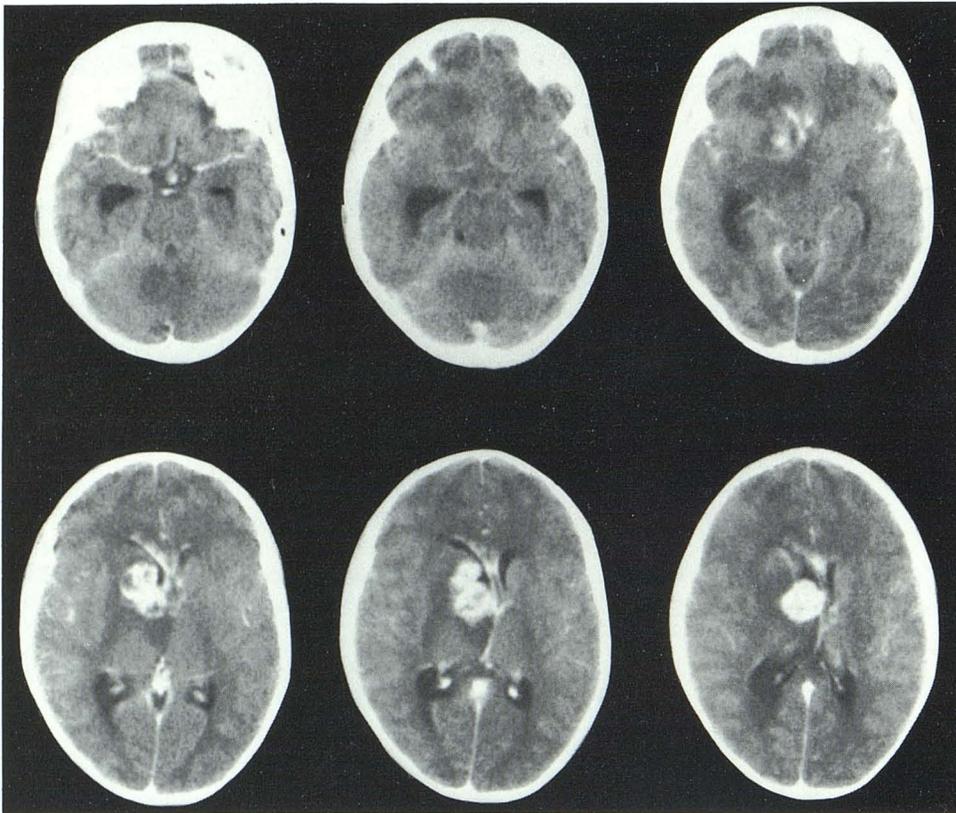


Fig. 2.—Contrast-enhanced CT scans 4 months after fig. 1. Marked interval enlargement of contrast-enhancing lesion in right caudate and foramen of Monro region with associated mass effect and hydrocephalus. Increase in pathologic subependymal contrast enhancement. No definite interval change in immediate suprasellar region.



Fig. 3.—Right internal carotid angiography, frontal view. Moderate neovascularity of intracerebral mass (arrow). No arteriovenous shunting was demonstrated. Vertebral angiography was normal.

examination was unremarkable except for the short stature and Tanner stage I pubertal development. The bone age was 13 years. Endocrine studies showed a mild increase in prolactin activity, mild diabetes insipidus, and deficiencies of growth hormone, FSH, and LH. Cyclical, monthly episodes of nausea and vomiting with dehydration led to hospital admission in May 1981, when right-sided papilledema was seen. Formal visual field examination was normal. Serum alpha-fetoprotein and human chorionic gonadotropin evaluations were normal. Cerebrospinal fluid analysis demonstrated 17 white blood cells per high power field. A brain computed tomographic (CT) study was abnormal (fig. 1). Follow-up CT with direct coronal views in June 1981 showed no change.

In September 1981, the patient was admitted again because of increasing lethargy and dehydration related to persisting monthly episodes of nausea and vomiting. CT showed a large mass in the area of previous abnormality deep within the brain (fig. 2). Angiography demonstrated moderate vascularity of the lesion (fig. 3). Biopsy by a transcortical approach with the aid of intraoperative sonographic guidance revealed a yolk-sac (endodermal sinus) tumor (fig. 4). Radiation therapy in the form of a 5,040 rad (50 Gy) tumor dose was delivered during 28 days. However, the patient developed lower extremity weakness and urinary retention during January 1982. Myelography elsewhere demonstrated multiple spinal intradural, extramedullary metastases including a complete block at the T10 level. Cranial CT in February 1982 revealed marked improvement in the previously demonstrated abnormalities, but interim development of a new right frontal convexity mass (fig. 5). Despite complete spinal axis and additional brain irradiation, the patient died in March 1982 before initiation of chemotherapy. Permission for an autopsy was not granted.

**Discussion**

Of the 17 previously reported primary intracranial, pure yolk-sac tumors, 13 presented in the pineal region and only four in the suprasellar region. The patients were 9–24 years old. To our knowledge, the present case report is the first description of the radiologic findings of such a tumor presenting in a subependymal lateral ventricular and suprasellar location. Information about CT findings is available for only six of the 17 reported tumors. Five showed relatively homogeneous contrast enhancement, one showed ringlike enhancement, and all presented as a midline or paramedian

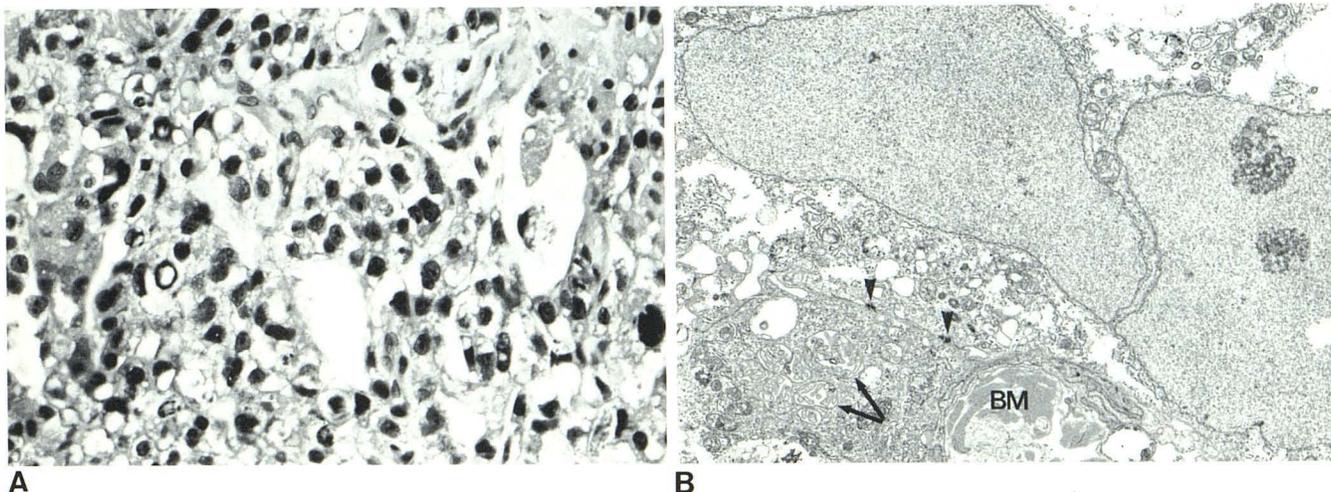


Fig. 4.—A, Tumor cells with large nuclei, prominent nucleoli, and abundant clear or vacuolated cytoplasm form solid sheets or glandlike spaces. Intracellular and extracellular hyaline globules were PAS-positive (H and E  $\times 470$ ). B, Endodermal cells have large nuclei and prominent nucleoli with threadlike nucleolonema. Cytoplasm contains variable numbers of organelles. Adjacent

cells are joined by desmosomes (arrowheads). Intercellular lumina (arrows) contain microvilli projecting into them from adjacent cells. Basement membranelike material (BM) lies between tumor cells. Similar material was also seen intracellularly ( $\times 7,500$ ).

Fig. 5.—Contrast-enhanced CT scan after radiation therapy. New enhancing mass in anterior part of right frontal lobe. Disappearance of subependymal contrast enhancement. Minimal ventricular deformity without definite pathologic contrast enhancement in region of previously demonstrated tumor near foramen of Monro. Scattered subarachnoid Pantopaque.



mass in the pineal region. Angiographic information available from four of the cases (also all in the pineal region) indicates that typically there is moderate to high neovascularity, similar to that seen in malignant teratomas or ependymomas. Obstructive hydrocephalus is common. There is a propensity for ependymal spread and subarachnoid dissemination, including spinal "drop metastases."

The inevitable progression of these tumors despite surgery and radiation attests to their aggressiveness. The small number of reported cases prevents a reliable conclusion regarding appropriate therapy.

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