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CT of Pineal Region Tumors

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The computed tomographic (CT) features of pineal region tumors were analyzed in 60 histologically proven tumors. This is the largest reported series of histologically verified pineal region tumors studied with CT. The tumors were classified as germ-cell tumors, glial tumors, pineal parenchymal tumors, and meningiomas. Preenhancement germinomas revealed characteristically high-density areas with calcification; uniform enhancement was seen after injection of contrast material. When present, pineal calcification was engulfed by the tumor. Teratomas, present only in male patients, revealed areas of mixed densities (e.g., calcification and fatty areas) and did not show significant contrast enhancement. Spontaneous intraventricular rupture was noted in one case. Unlike other tumors, the original pineal calcification could be recognized in two-thirds of glioma cases and was displaced anteriorly and superiorly in most. Gliomas were hypodense to isodense on precontrast scans and enhanced in a nodular and a ring fashion. Benign pineal parenchymal tumors showed iso- to hyperdense areas with nodular enhancement after injection of contrast material. Pineoblastomas were well defined hyperdense masses without calcification on precontrast scans. After injection of contrast material, they showed well defined enhancement with occasional small, central lucencies. Meningiomas were hyperdense in most cases, uniformly enhanced in a homogeneous fashion, and showed a tentorial attachment.

Pineal region tumors constitute 2% of intracranial tumors. Masses in the region of the pineal body form a group whose anatomic location has usually precluded histologic verification before treatment [1]. The pineal body has a close relation to the quadrigeminal plate cistern, posterior part of the thalamus, posterior third ventricle, and vein of Galen. Lesions may infiltrate these adjacent structures by contiguous spread, and sometimes it may be impossible even at necropsy to determine the primary site. This has often resulted in grouping of the tumors in this area as pineal region tumors [1]. In general, documented by surgical and autopsy series, about 25% of the tumors of the pineal region are benign and encapsulated. These lesions are generally considered insensitive to radiotherapy, and because of their defined borders are amenable to surgical removal [2].

In the past, neurologic management of pineal region tumors has been strongly influenced by unfavorable results after direct surgical intervention. Mortality ranged from 29% in one series [3] to 70% in another [4]. In 1960, a representative series was reported of third ventricular tumors at Neurological Institute of New York [5]. Among the pineal tumors there was total removal in five cases, resulting in three deaths. Keeping in mind the above results, a retrospective study of histologically proven pineal tumors studied by computed tomography (CT) was undertaken to correlate CT appearance with various histologic tumor types.

Materials and Methods

CT scans of 60 patients with histologically proven pineal region tumors were studied. There were 44 males and 16 females. CT scans were obtained before and after enhancement

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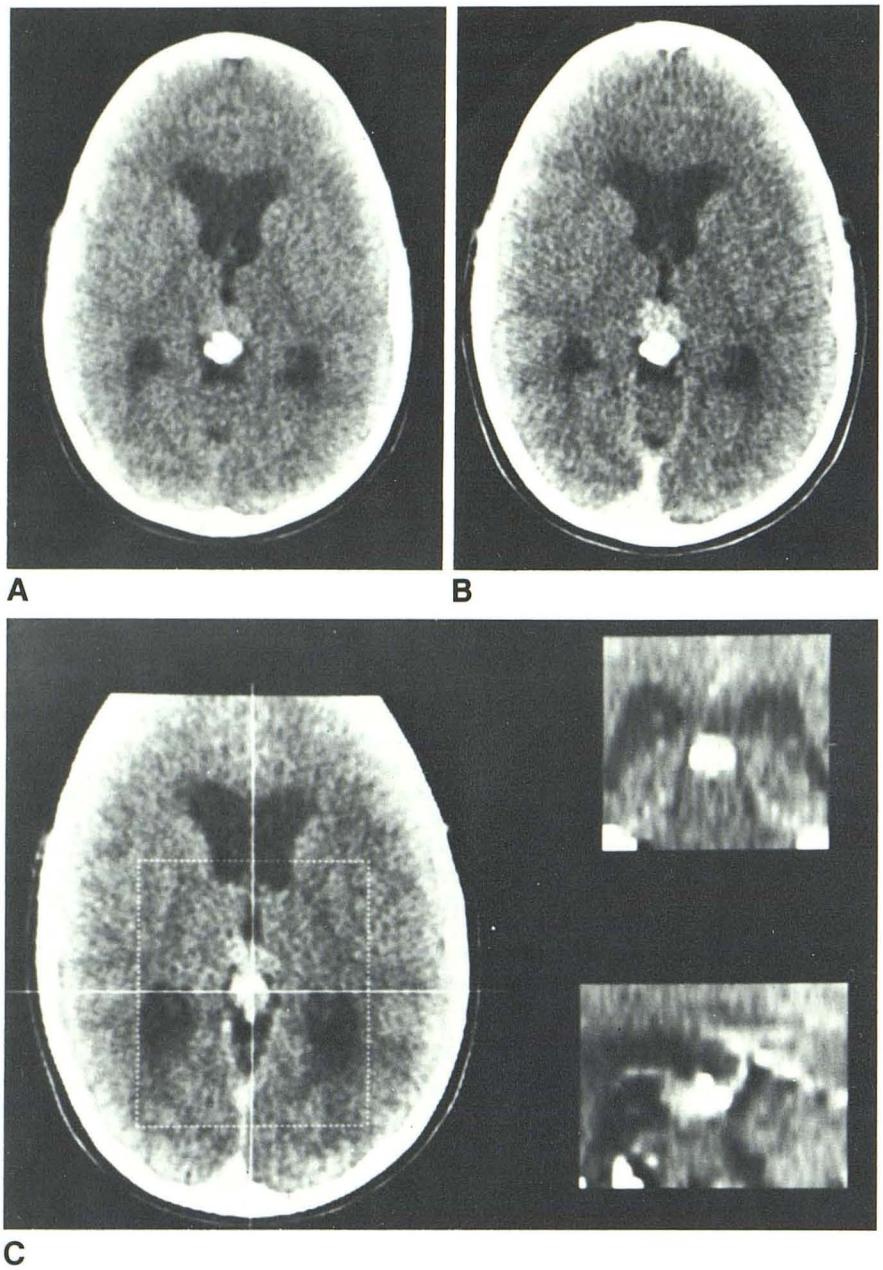


Fig. 1.—Germinoma. **A**, Precontrast scan. Calcified prominent pineal gland is associated with high-density mass anteriorly. Mild hydrocephalus. **B**, Postcontrast scan. Enhancement in high-density region. **C**, Reconstructed coronal and sagittal views. Calcified and enhancing lesion in pineal area.

in almost all patients. Coronal views were obtained whenever possible and were considered necessary (12 of 60). Angiograms were obtained in most patients (53 of 60) to identify vascular supply and venous drainage. It was not our intent to offer specific CT and angiographic correlations, but rather to correlate CT and histology of pineal region tumors. Age and gender were important factors in the differential diagnosis, since certain pineal region tumors, such as teratomas, germinomas, and pineoblastomas, occur either exclusively or predominantly in males.

Results

Pineal region tumors were divided into four broad categories according to the pathologic classification of Rubinstein [6]. They were divided into germ-cell tumors, glial tumors, pineal parenchymal tumors, and meningiomas.

Germ Cell Tumors

Under this category there were four subdivisions: germinomas, teratomas, embryonal cell carcinoma, and choriocarcinoma.

Germinomas. There were 16 cases of pineal germinoma, 13 in males and three in females. In 15 cases the lesion was denser than brain on unenhanced CT scans. The other patient did not have an unenhanced study. Moderate-to-marked enhancement was noted in 14 cases, slight enhancement in one

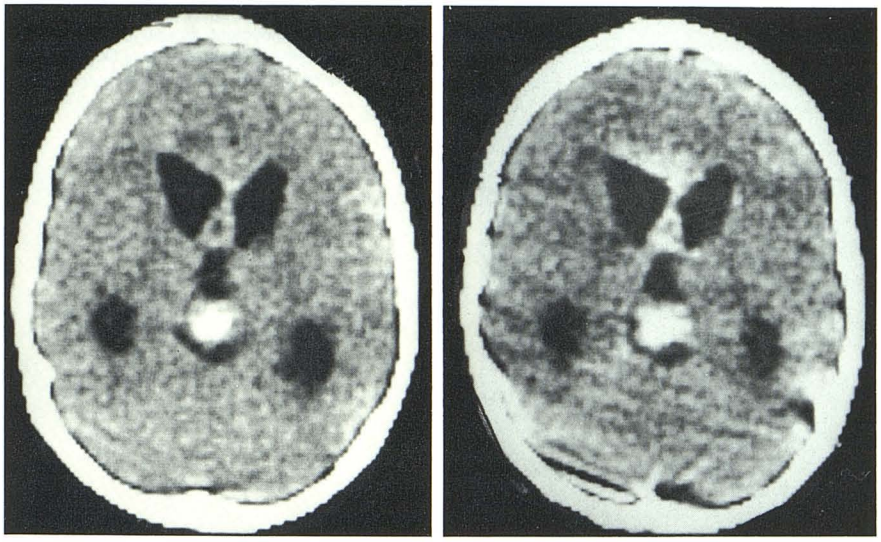


Fig. 2.—Germinoma with seeding. **A**, Precontrast scan. Prominent pineal calcification with hyperdense mass associated with periventricular high density and hydrocephalus. **B**, Postcontrast scan. Enhancing pineal mass with periventricular enhancement consistent with tumor seeding.

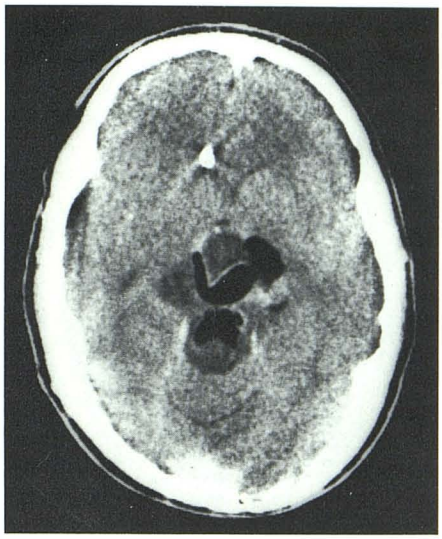
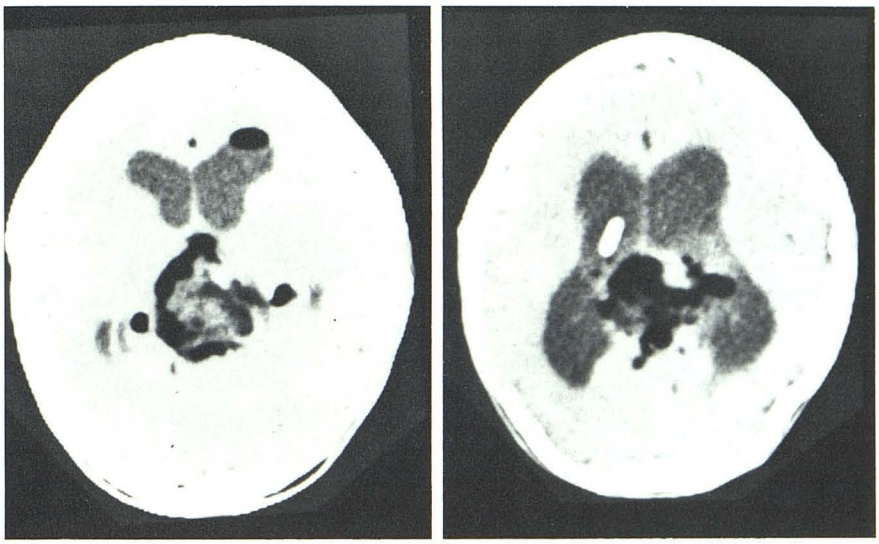


Fig. 3.—Teratoma. Postcontrast axial view. Mixed densities, including fatty areas and cystic areas associated with linear calcifications. No significant enhancement. Shunt tip is in right lateral ventricle.

Fig. 4.—Spontaneous rupture of teratoma. **A**, Wide window at frontal horn level. Fatty densities in pineal area, frontal horn, and pericallosal cistern. **B**, Fatty tumor has ruptured into lateral ventricles. Shunt tip in right lateral ventricle.



case, and no enhancement in one case. A prominent and slightly enlarged pineal calcification was noted in six cases and was engulfed by enhancing tumor in all (fig. 1). The prominent pineal gland calcification was recognized by its discrete character and its close-to-midline location. On the other hand, parenchymal calcification of the tumor was recognized by a coarse, nodular pattern on either side of the pineal body. This was observed in 10 cases. Meningeal and ependymal seeding was found in five cases and was demonstrated by CT in the subarachnoid space and the ventricular wall (fig. 2). Spinal seeding was detected by myelography.

Teratomas. There were five cases of teratoma; all were in males. The teratomas were all sharply demarcated from the surrounding brain. Another distinct feature, fatty substances with a low CT attenuation, was identified in all five cases. No other tumor in this series showed these fatty densities. In four of the five cases there was tumor calcification showing a mixture of linear and nodular densities (fig. 3). No significant contrast enhancement was observed. In one case, a small area of malignant transformation was detected histologically. In this case, the initial CT interpretation was of a teratoma. On follow-up, there was subarachnoid seeding in the suprasellar area and periventricular enhancement. Spontaneous intraventricular rupture of a teratoma was seen in one case (fig. 4).

Embryonal cell carcinoma and choriocarcinoma. There was

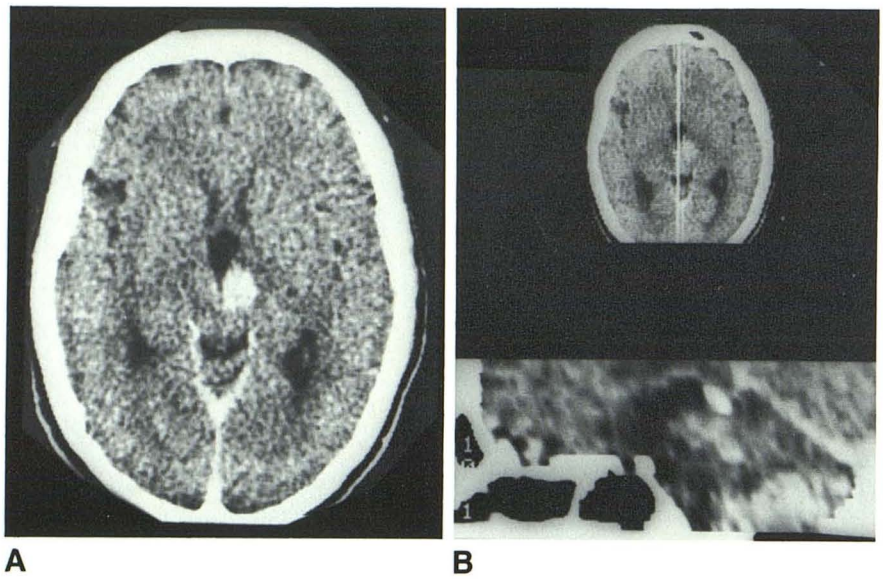


Fig. 5.—Nodular glioma. **A**, Postcontrast axial scan. Nodular enhancement around posterior third ventricle. **B**, Sagittal reconstruction. Nodular enhancement below calcified pineal gland.

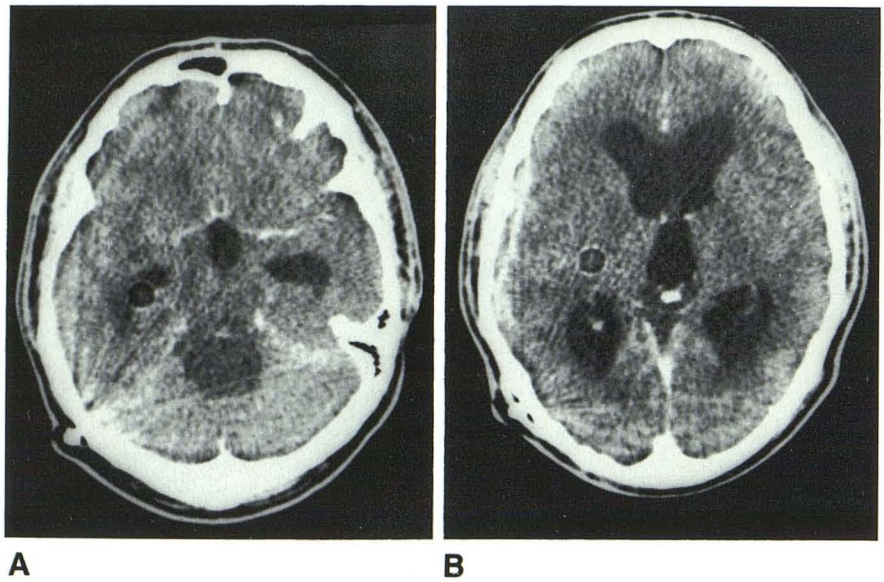


Fig. 6.—Cystic glioma. **A**, Postcontrast scan. Lucent, nonenhancing cystic lesion projecting into posterior fossa, without fat densities. **B**, Higher level. Calcified pineal displaced superiorly by inferiorly located tumor.

one case each of embryonal cell carcinoma and choriocarcinoma; both tumors were hyperdense on unenhanced scans, but enhanced markedly after administration of contrast material. There was no abnormal pineal calcification or parenchymal calcification within the tumor. In this group angiography provided a very useful clue for the diagnosis, showing pronounced vascularity.

Glial Tumors

There were 16 cases of glioma, 10 in males and six in females. Twelve cases revealed hypodense areas, two were isodense, one had mixed density with coarse calcification, and one had hydrocephalus with no detectable mass. Normal pineal calcification was observed in 11 patients in this group. The pineal calcification was displaced upward and forward in 10 cases and posteriorly in one. In the other four cases, no pineal calcification was detected. The pineal calcification appeared to be normal and was identified in the midline as a discrete density. The patient with no detectable mass and hydrocephalus on CT had a magnetic resonance (MR) scan at another institution; it revealed a definite mass in the pineal region. After intravenous injection of contrast material, moderate-to-marked enhancement was noted in 11 cases, mild enhancement in four, and no abnormal enhancement in one (the case in which a lesion was evident on MR). The enhancement pattern was a single nodule in six cases (fig. 5), a "ring" in six cases, and no enhancement pattern in one case. Two cases appeared "cystic" with no enhancing cyst wall (fig. 6).

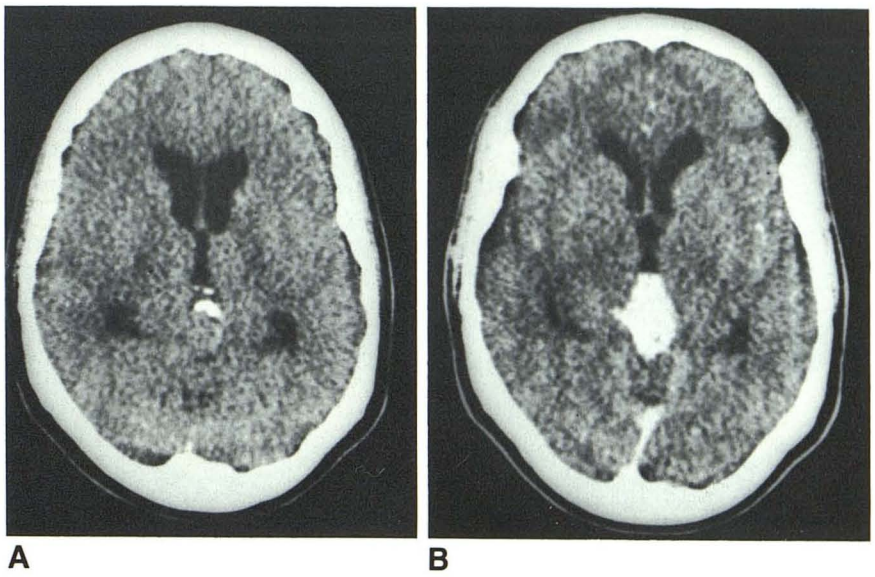


Fig. 7.—Pineocytoma. **A**, Precontrast scan. Partly calcified and isodense mass in pineal area associated with mild hydrocephalus. **B**, Postcontrast scan. Nodular enhancement.

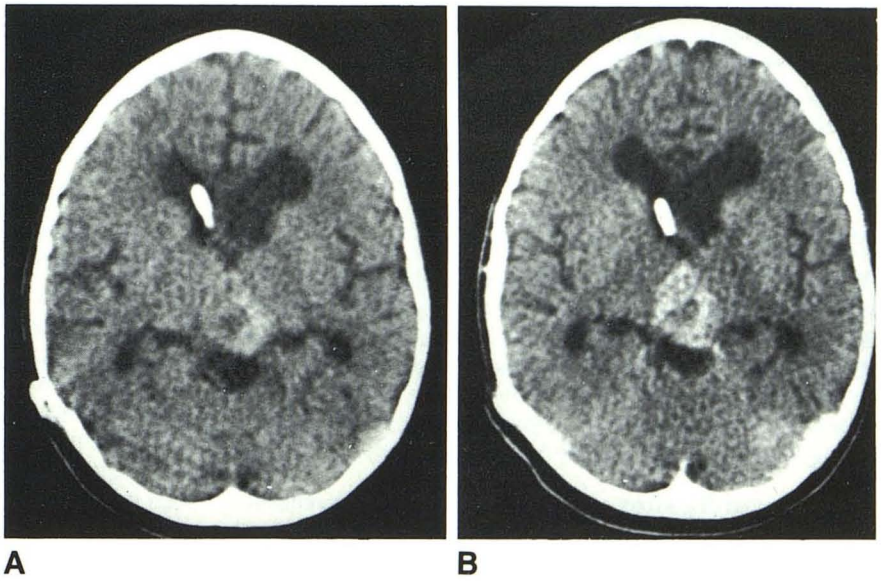


Fig. 8.—Pineoblastoma. **A**, Precontrast CT scan. Hyperdense mass with central lucency in pineal area, mild hydrocephalus, and shunt tip in right frontal horn. **B**, Postcontrast scan. Enhancement with central lucency probably representing necrotic area.

Pineal Parenchymal Tumors

There were 13 cases of pineal parenchymal tumors; they were subdivided into benign-type pineocytomas (eight cases) and malignant-type pineoblastomas (five cases). Of the eight pineocytomas, five were in males and three in females. Pineocytomas were isodense to hyperdense relative to brain on unenhanced CT scans in seven cases; no preenhancement CT scan was obtained in one case. The tumor enhanced as a well defined, homogeneous nodule in seven cases (fig. 7), and no enhancement occurred in one case. One case had an abnormally large pineal calcification, one case had a normal pineal calcification, and six cases showed no pineal calcification.

The five cases of pineoblastomas were all in males. All had a well defined mass without calcification. Four cases showed moderate enhancement with a small central lucency, unlike the benign pineocytomas (fig. 8). Ependymal seeding in the lateral ventricle was seen in one case.

Meningiomas

There were eight cases of meningioma occurring in the pineal region, four in females and four in males. There was a case of meningeal sarcoma in this group. Six cases were hyperdense on precontrast CT and two cases had no precontrast CT scan. All cases showed homogeneous intense enhancement. Three cases had coronal CT, which demonstrated the tentorial attachment (fig. 9).

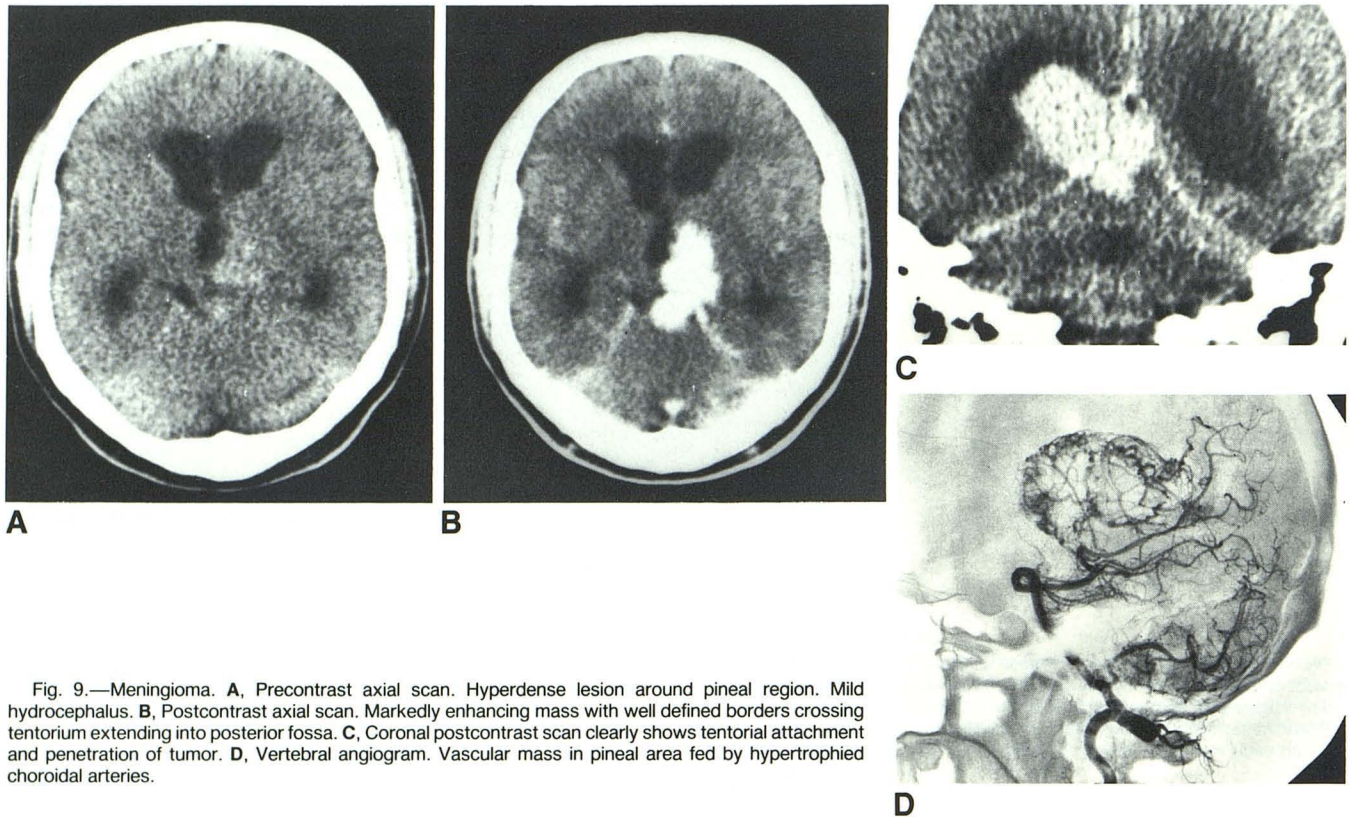


Fig. 9.—Meningioma. **A**, Precontrast axial scan. Hyperdense lesion around pineal region. Mild hydrocephalus. **B**, Postcontrast axial scan. Markedly enhancing mass with well defined borders crossing tentorium extending into posterior fossa. **C**, Coronal postcontrast scan clearly shows tentorial attachment and penetration of tumor. **D**, Vertebral angiogram. Vascular mass in pineal area fed by hypertrophied choroidal arteries.

Discussion

We are reporting the largest series of histologically proven pineal tumors studied by CT. Previous reports [7–10] included a smaller number of histologically documented lesions, specifically 33, 22, nine, and seven cases, respectively.

Germinomas

The cell origin of germinomas is usually ascribed to primitive germ cells that are said to migrate over wide areas of the embryo during early fetal life. The pineal area is the most common site of intracranial germinomas. Many arise in the midline in the hypothalamic region and are frequently called ectopic pinealomas or suprasellar germinomas. Most germinomas are reported to occur in the second and third decades of life, and this was true in our series also. Germinomas of the pineal region show an overwhelming male preponderance. Germinomas of the suprasellar region show a lesser preponderance of males [6]. Germinomas are the most common tumor of the pineal region, accounting for 50% of all neoplasms in this area [6]. In our series, this neoplasm was the most common single type, accounting for 16 of 60 cases. In the series reported by Jooma and Kendall [7], germinoma was the most common tumor, constituting 17 of 33 cases. The incidence of calcification in the germinomas in our series was higher than in the series of Jooma and Kendall, probably because our patients were studied with high-resolution CT.

The characteristic CT features of germinoma are of a well defined, hyperdense, occasionally calcified tumor occurring in a young male, engulfing the pineal gland, and enhancing homogeneously. Pathologically, germinomas are highly cellular neoplasms, usually containing no hemorrhage, necrosis, or cystic degeneration [11]. The hyperdense areas on CT may correlate with the densely cellular nature of these tumors. A prominently calcified pineal gland engulfed by tumor tissue, a characteristic feature of germinomas in our series, was not seen in any other pineal tumors. Germinomas are histologically malignant, and infiltrative spread is likely to occur along the floor and walls of the third ventricle. Seeding along the lateral ventricles and cerebrospinal fluid (CSF) spaces is common. Five of our 16 cases revealed CSF seeding either at the initial or later stages of the tumor.

Teratomas

The pineal is the most common site of intracranial teratomas. In an analysis of 94 reported cases of teratomas involving the brain [12], 39 cases were in the pineal area. The incidence of teratomas is highest in the second and third decades, as was true for germinomas. Teratomas also occur predominantly in males; all our five cases were in males.

Pathologically, teratomas have well defined borders with variegated cut surfaces. Teratomatous elements such as cartilage, bone, and hair are often seen. All five teratomas

demonstrated fat densities and four revealed calcification. Unlike the coarse, stippled calcification of germinomas, teratomas possess a mixture of linear and nodular calcifications. Spontaneous rupture of teratomas into ventricles has been reported only once in the CT literature [13]. Our case is the second. Besides teratomas, dermoids and epidermoids are also known to rupture spontaneously. In brief, the CT features of teratomas should include hypodense areas of various fats and linear or nodular calcification, either in the periphery or in the center of the tumor. The calcific nodules are usually larger than those observed in germinomas. The other less frequently occurring germ-cell tumors are choriocarcinomas and chorioepitheliomas. There was one case of each, and no specific CT characterization can be made except that they were hyperdense, enhanced after intravenous administration of contrast material like other germ-cell tumors, and contained no fat densities. They were very vascular angiographically.

Pineal Cell Tumors

Tumors arising from the pineal parenchymal cells are probably less common than germinomas and teratomas [6]. However, in our series, pineal parenchymal cell tumors outnumbered teratomas. The term "pinealoma" would, strictly speaking, be appropriate only for tumors arising from the pineal parenchymal cells. Those arising from the pineal parenchymal cells are divided into more mature pineocytomas and more primitive pineoblastomas. The pineocytoma is usually well circumscribed, noninvasive, slowly growing, may be found at any age, and both genders are equally affected. The CT features of pineocytomas include isodense and hyperdense areas on unenhanced CT. Enhancement was homogeneous without apparent breakdown or necrosis in the tumor. In males pineocytomas cannot be distinguished from noncalcified germinomas. When pineal calcification is seen in a female, the lesion is more likely a pineocytoma.

Pineoblastomas are usually frankly invasive and highly malignant. Histologically, the tumor is densely cellular. Hemorrhage and calcification are not found, but cystic degeneration is observed occasionally. Cystic degeneration was noted in our series as ring enhancement with central low density. Extension of pineoblastomas to involve the cerebellar vermis is not uncommon and often raises the question of medulloblastoma. Histologically, pineoblastomas and medulloblastomas are very similar. On CT scans, pineoblastomas are characterized by a lucent center, which differentiates them from the benign pineocytoma.

Glial Tumors

Since the normal pineal gland includes fibrillary astrocytes, it is possible that it may be the source of glial tumors such as astrocytomas and glioblastomas. However, by the time these tumors are examined pathologically, they are so extensive that the precise origin may be difficult to identify. Many of these tumors probably arise from the quadrigeminal plate or from the wall of the third ventricle [6].

In 22 cases of pineal region tumors studied by Zimmerman et al. [8], there were eight gliomas and seven germinomas. In another series [7], there were three gliomas out of 33 cases. In our series gliomas constituted 16 of 60, the second most common tumor after germ-cell tumors. Most gliomas in our series displaced the pineal gland anteriorly and superiorly, which suggested the tumor was extrapineal in origin. The precontrast CT scan usually helped establish the CT diagnosis of gliomas, since they were hypodense relative to brain. The two cases presenting as nonenhancing cysts were apparently unique; such a morphology has not been reported before. Gliomas occurred in males and females in equal numbers, and age distribution did not provide any help in the diagnosis.

Meningiomas

Meningiomas arising in the pineal area are rare. In the series of Zimmerman et al. [8] and Jooma and Kendall [7], no meningiomas were reported. Sachs et al. [14] reported three cases of meningiomas in the pineal region. At that time, there was a total of 20 cases of meningiomas in this location in the literature, including their own three cases. In one series of 20 pineal region tumors, one tumor was a meningioma [2]. There were eight cases of meningiomas in our series. Both males and females were equally affected, although, in general, meningiomas are more often found in females than in males.

Meningiomas occupying the pineal area may arise from the velum interpositum or from the free edge of tentorium, where it is joined by the inferior margin of the falx. The meningiomas in the pineal area were hyperdense in most cases on unenhanced CT and enhanced in a nodular fashion after intravenous administration of contrast material. The characteristic feature of dural attachment was best seen on the coronal view. Angiography helped in identifying the feeders to the tumor and the characteristic homogeneous blush. One case showed sarcomatous changes and there was also spinal seeding into the lumbar subarachnoid space. No other cases had seeding and none had fat densities.

Clinical Management

Stein [2, 15] used an infratentorial supracerebellar approach, which reaches the tumor below the deep venous system. In his series of over 60 cases, there were three deaths and no significant operative morbidity.

Treatment programs in nonresectable tumors are tailored to the histologic nature of the tumor. In germinomas, CSF is evaluated three times for abnormal cells, and a myelogram is obtained. If there is any evidence of CSF seeding, radiation therapy is applied to the entire brain with a boost to the tumor site as well as radiation to the spinal canal. In astrocytomas, radiation is directed to the site of the tumor only. In pineal cell tumors, radiation is directed primarily to the tumor site. In embryonal cell tumors such as embryonal carcinoma and choriocarcinoma, chemotherapy may be the treatment of first choice. In germinomas that escape the effects of radiotherapy, chemotherapy is considered.

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