Meningeal Dissemination of Retinoblastoma: CT Findings in Eight Patients

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Thirty-seven patients with retinoblastoma were evaluated prospectively by clinical examination, lumbar puncture, and CT. Eight (22%) of the 37 were found to have meningeal dissemination. The tumor was bilateral in three patients. Two cases showed no CT signs of local recurrence. Headache, nausea/vomiting, and restlessness were the most common symptoms. CT scans in these patients showed diffuse meningeal contrast enhancement, nodular masses, ependymal-subependymal enhancement, and ventricular dilatation.

Our series of eight patients with meningeal spread illustrates a considerable range of dissemination patterns referable to retinoblastoma. One case illustrated the CT finding of multiple epidural metastases.

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Retinoblastoma is a tumor arising from primitive embryonal retinal cells. It is the most common malignant intraocular neoplasm of infancy and childhood. Reported worldwide prevalence ranges from 1 per 15,000 to 1 per 30,000 live births. When the lesion extends beyond the eye, mortality approaches 100%. However, by early diagnosis, the 5-year survival rate is 92% [1].

As a rule, retinoblastoma spreads through the optic nerve or the choroid, but such invasion is not a reliable predictor of a metastatic pattern. Virtually all patients succumbing to retinoblastoma show brain or skull involvement, and one half to two thirds also exhibit spread to distant organs [2].

This report describes CT findings in eight retinoblastoma patients with demonstrable meningeal dissemination, two of whom showed no signs of local recurrence.

Subjects and Methods

From March 1985 to May 1989, 37 cases of retinoblastoma with unilateral or bilateral involvement were diagnosed at our centers. Patients' ages ranged from 9 months to 8 years.

All patients were studied by CT (CT/T 8800 General Electric Medical Systems, Milwaukee; CT/MAX General Electric Medical Systems, Tokyo; and TCT-600-S Toshiba Medical Systems, Tokyo), and postcontrast CT scans were obtained in all but one patient. Scanning was performed in an axial plane with 10-mm sections or with 5-mm sections and overlapping for the orbit.

Results

Of the 37 patients in our study population, eight (four boys and four girls) had demonstrable meningeal dissemination by CSF analysis and CT scanning, and one case was also demonstrated by myelography. Three of the eight patients were under 3 years old, three were 3 years old, one was 6 years old, and one was 8 years old. The tumor was bilateral in three patients. Five of the 37 patients were asymptomatic; two of them showed intracranial nodular masses throughout the

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optic pathways. All eight cases of meningeal metastases were symptomatic.

The most common symptoms (Table 1) were headache (in four cases) and nausea and vomiting (in three cases). Three patients also exhibited restlessness. Time lag from tumor diagnosis to the appearance of metastatic lesions ranged from 6 to 32 months. During this time, patients received radiotherapy and chemotherapy.

CT scans showed enhancement after contrast injection, particularly within cisternal spaces, In the posterior fossa, enhancement was often bilateral, extending superiorly around the brainstem, predominantly in the quadrigeminal and perimesencephalic cisterns. It was also evident along the sylvian and interhemispheric fissures (Figs. 1 and 2).

Periventricular spread of tumor cells was observed in two patients and was manifested by ependymal and subependymal nodular enhancement (Table 2 and Figs. 3 and 4). A rounded mass of tumor was seen in the third ventricle (Fig. 5) in four cases, and in the suprasellar cistern in three patients. In six cases CT scanning disclosed ventricular dilatation with periventricular lucency, which also was the only finding in the single patient who did not receive IV contrast injection. In one case there were multiple epidural metastases (Fig. 6), indicating that the mode of spread in this instance was hematogenous and therefore different from leptomeningeal spread as in the other cases.

TABLE 1: Symptoms at Diagnosis of Meningeal Metastasis in Eight Cases of Retinoblastoma

Symptoms	No. of Patients
Headache	4
Nausea/vomiting	3
Quadriparesia	2
Movement disorders	1
Hyporeflexia	2
Endocranial hypertension	2
Coma	1
Restlessness	3

TABLE 2: CT Findings in Eight Cases of Retinoblastoma with Meningeal Spread

Finding	No. of Patients
Local recurrence	6
Meningeal contrast enhancement	7
Nodular masses	4
Ependymal-subependymal enhancement	2
Local osteolysis	1
Exocranial lesions	1
Ventricular dilatation	6

Discussion

Retinoblastoma metastasis invades the CNS in some 50% of cases and may involve brain parenchyma, subarachnoid space and meninges, or spinal cord. Intracerebral metastasis has been documented in 25–41% of postmortem examinations [3]. Meningeal spread alone is rare. Meningeal metastasis in retinoblastoma appears most commonly with uncalcified tumors that exceed orbital boundaries, particularly when they extend into the skull [4].

Although all of our 37 cases exhibited orbital calcifications, eight of them had meningeal spread. Two patients showed no local recurrence at the time of meningeal spread. The reported time lag between tumor excision and the appearance

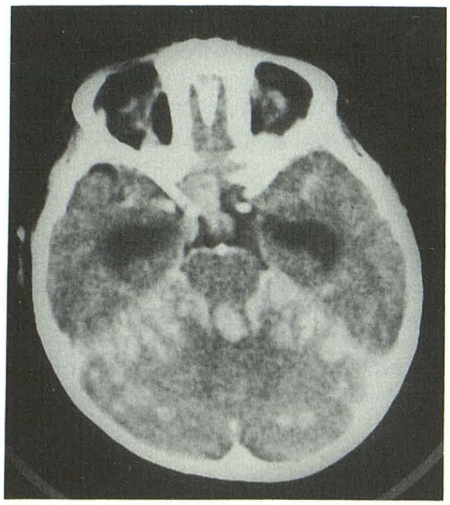
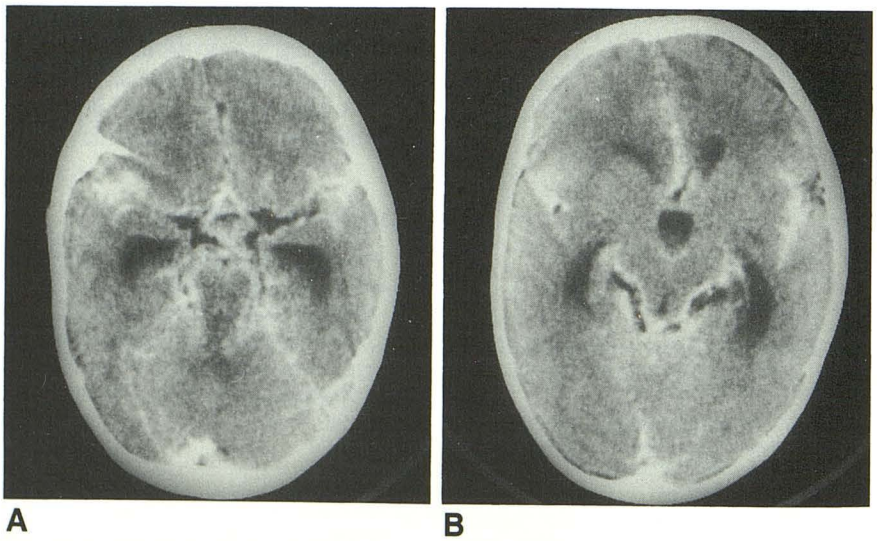


Fig. 1.—Axial CT scan of retinoblastoma after contrast injection shows diffuse enhancement in cerebellopontine cistern, quadrigeminal cistern, suprasellar cistern, cerebellum, fourth ventricle, and tentorium.



A B

Fig. 2.—*A* and *B*, Diffuse postcontrast enhancement in sylvian fissure, interhemispheric fissure, suprasellar cistern, and quadrigeminal cistern. Ventricular dilatation is moderate.

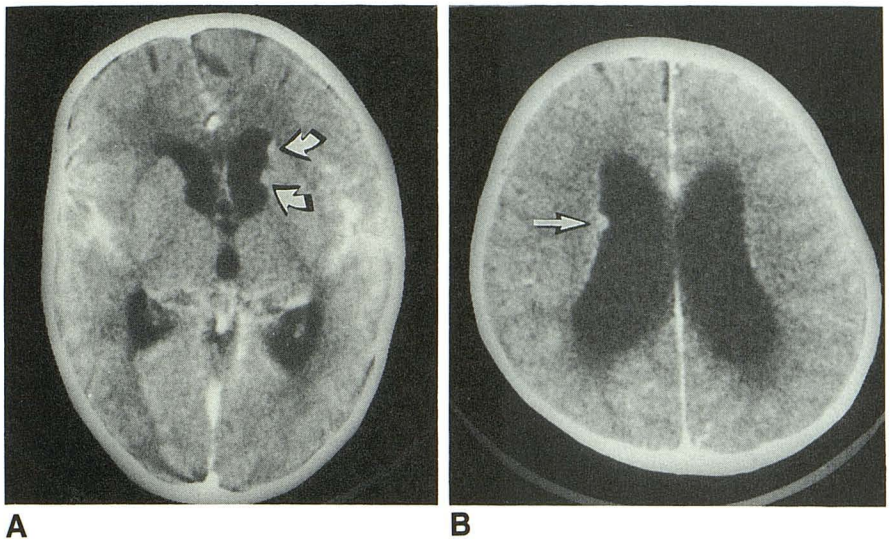


Fig. 3.—*A* and *B*, Postcontrast CT scans show subependymal nodules in frontal horns (*A*) and body (*B*) of lateral ventricles (*arrows*). Ventricular dilatation with mild periventricular lucency is also present.

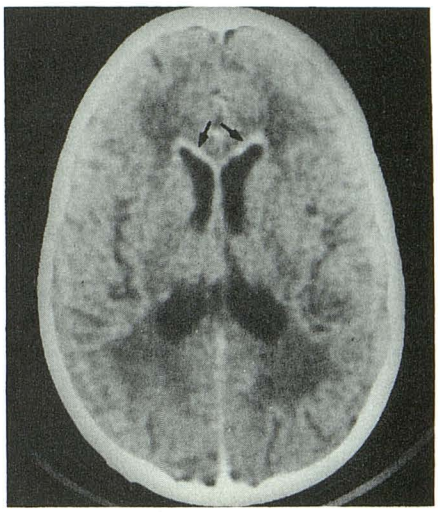


Fig. 4.—Postcontrast CT scan shows ependymal enhancement at frontal horns of lateral ventricles (*arrows*). Also note periventricular lucencies with mild ventricular dilatation.

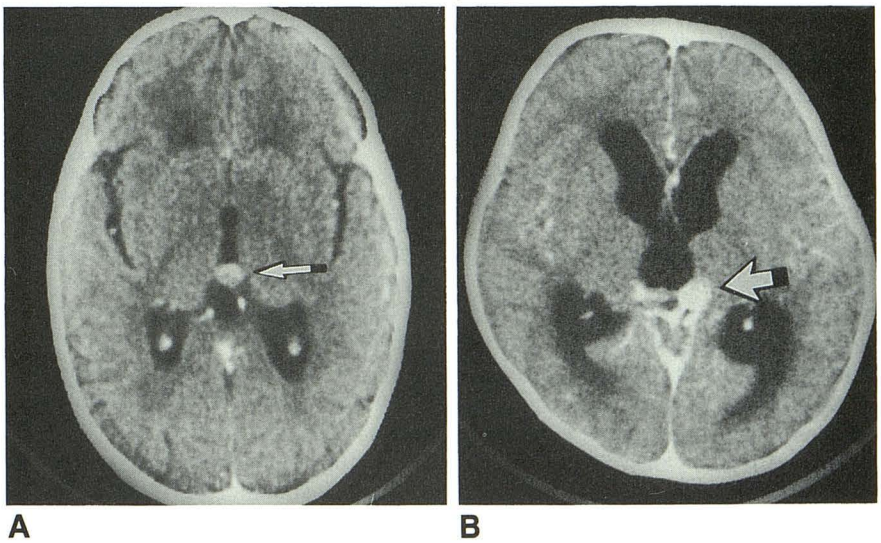


Fig. 5.—*A* and *B*, Postcontrast CT scans show nodular lesions in third ventricle (*arrows*).

of metastatic lesions has been extremely variable, ranging from 3 weeks to 24 months [4]. In our series the interval was 6 to 32 months.

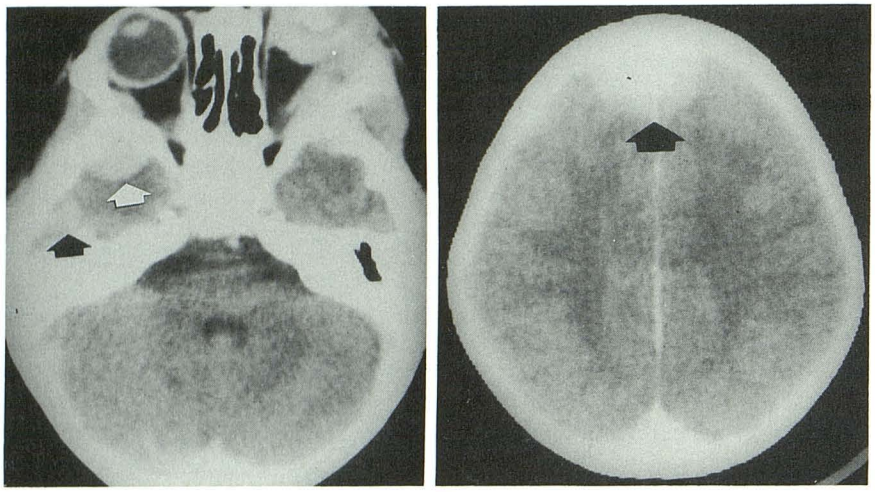
Prognosis for retinoblastoma patients with meningeal metastasis is invariably poor: six of our patients died within 5–20 days, regardless of the chemotherapeutic schedule employed; one is still alive at 6 months after surgery; and the remaining patient was lost to follow-up.

Both CT and MR imaging are valuable diagnostic techniques for the follow-up of tumors likely to metastasize to the leptomeninges [5] as well as for the control of patients diagnosed and treated for retinoblastoma. Five of our 37 cases were asymptomatic but had lesions demonstrable on CT that were compatible with intracranial spread of the underlying neoplastic disease.

Our treatment schedule for patients with retinoblastoma that exceeds the limits of the eyeball consists of (1) orbital and chiasmic radiotherapy (linear accelerator), (2) CT control 30 days later, and (3) repeated CT scanning every 6 months for at least 2 years.

Ascherl et al. [6] described the CT findings in patients with widespread malignant meningeal neoplasms as showing gyral enhancement not accompanied by surrounding edema; sulcal and cisternal obliteration, and enhancement in the perimesencephalic, suprasellar, and sylvian fissures; and ependymal-subependymal enhancement. They also noted ventricular dilatation with or without periventricular lucency.

In our series, we found a variable combination of these lesions. In a single case there was patent multiple epidural metastasis resembling intracranial involvement referable to



A B

Fig. 6.—*A* and *B*, Retinoblastoma in left eye. Postcontrast CT scans show multiple epidural metastases (*arrows*) and calcification and enhancement in left globe.

neuroblastoma and rhabdomyosarcoma, which to our knowledge has not been previously documented in the literature.

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