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Cranial CT of Neurofibromatosis

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The results of computed tomography (CT) of the orbits and brain of 29 patients with neurofibromatosis were reviewed to determine the contribution of CT scanning to the diagnosis of this disorder. In the presence of orbital symptoms, CT confirmed the presence of suspected lesions such as optic nerve gliomas, demonstrated asymptomatic and atypical lesions, and displayed concomitant intracranial involvement of the optic chiasm. CT of the brain revealed a multiplicity of abnormalities such as astrocytomas, intracerebral calcification, hydrocephalus, and congenital lesions.

Neurofibromatosis is an inherited disorder resulting in hamartomatous or neoplastic changes in the derivatives of the primary germ layers that may affect any organ system. Some authors categorize neurofibromatosis into central, peripheral, or mixed types according to tumor site. The incidence of central nervous system tumors in 223 patients with neurofibromatosis was 6 times that of the general population [1].

Multiplicity is the predominant characteristic of central nervous system lesions accompanying neurofibromatosis. Lichtenstein [2] presented an overall schema of these lesions and stated that they represent "foci of hyperplasia and neoplasia of the supportive derivatives of the primitive ectoderm." Some of the more commonly encountered central nervous system tumors include optic gliomas, acoustic schwannomas, meningiomas, piloid astrocytomas, and ependymomas.

The plain radiographic findings of this disease have been comprehensively reviewed [3–5], but we know of only one report dealing with cranial computed tomographic (CT) findings [6]. The purpose of this investigation was to review our experience with cranial CT in patients with neurofibromatosis to determine the contribution of CT scanning in the diagnosis of this disorder.

Materials and Methods

From 1973 to 1978, 148 patients with the diagnosis of neurofibromatosis were seen at University of Iowa Hospitals and Clinics. Of these, cranial CT was requested only for those 29 patients with appropriate symptoms. There were 29 examinations of the brain and 16 examinations of the orbits.

We reviewed the medical records of patients undergoing CT and confirmed the diagnosis of neurofibromatosis using the criterion of six or more café au lait spots, each greater than 1.5 cm in diameter [1]. One case (case 21) did not have cutaneous pigmentation but was included because of characteristic radiographic findings and a facial plexiform neuroma.

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The most common symptoms (table 1) were visual loss (eight cases) and seizures (seven cases). The other 14 cases demonstrated a variety of symptoms including hearing loss, endocrine disturbance, hemiparesis, ataxia, depression, proptosis, and peripheral neuropathy.

An EMI Mark 1 scanner, later updated to the EMI 1005 configuration, was used. One examination was performed with an 80 × 80 matrix and the remainder were performed with the 160 × 160 matrix. Intravenous contrast enhancement was performed in 21 examinations.

We recorded the gross and histologic descriptions of the lesions that had been biopsied and noted the results of contributory radiographic examinations (table 2). We did not evaluate these patients for macrocranium, a finding reported in up to 75% of children with neurofibromatosis [7, 8].

TABLE 1: **Predominant Symptoms of 29 Patients with Neurofibromatosis**

Symptom	CT Findings (No. Patients)	
	Abnormal	Normal
Visual loss	7	1
Seizures	5	2
Hearing loss	2	2
Endocrine disturbance, rule out hypothalamic mass	1	1
Hemiparesis	1	0
Proptosis	3	0
Ataxia	2	0
Other	0	2
Total	21	8

Results

Eight patients had normal CT examinations. CT of the other 21 patients (72%) showed one or more abnormality of the orbits or brain (table 2). The average age of patients with abnormal examinations was 12 years (range, 2½–61). A family history of neurofibromatosis was found in nine patients and subcutaneous nodules were demonstrated in 14 patients.

Orbital Lesions

Of the 10 patients with demonstrated orbital abnormalities, six had discrete optic nerve lesions consistent with glioma (cases 1, 3, 5–7, and 9). One case (case 1) was confirmed by biopsy. Exploration of the opposite optic nerve in this patient showed tumoral swelling that was not detected by CT. The optic chiasm was normal by CT, pneumoencephalography, and operative inspection. In three of the six cases of optic glioma (cases 5–7), a soft-tissue mass of the optic chiasm was demonstrated by CT (fig. 1). CT in one case (case 2) showed a diffuse density of the retrobulbar tissues with scleral contrast enhancement, findings not typical of optic glioma. After biopsy and orbital exenteration, the histologic evaluation was malignant schwannoma (fig. 2).

CT in another case (case 4) demonstrated a diffuse retrobulbar mass with the additional finding of globe enlargement. A plexiform neurofibroma that had infiltrated the globe was found at operation.

A densely calcified mass along the optic nerve in case 8 was correctly diagnosed as a meningioma of the optic nerve sheath (fig. 3A). During resection of this lesion, an en plaque meningioma of the frontal lobe was discovered which, even in retrospect, was not evident on CT. CT in this case also revealed several sites of punctate calcification with no associated contrast enhancement (fig. 3B). These areas were asymptomatic and were not biopsied.

CT in one case (case 21) demonstrated extensive unilateral abnormalities of the skull and extracranial soft tissues (fig. 4). The left greater wing of the sphenoid was dysplastic, resulting in the absence of the posterior orbital wall and enlargement of the middle cranial fossa. The ipsilateral sylvian fissure was wide, suggesting that the volume of brain tissue was not increased but was merely accommodating the larger capacity of the temporal fossa. A subcutaneous soft-tissue density extended from the left ear to the left orbit. The left globe was displaced inferiorly. A diffuse soft-tissue mass that enhanced slightly after contrast infusion was present in the left retrobulbar area and obliterated the optic nerve. Several operations have been performed to debulk the facial tumor and to decompress the orbit; the histologic reports have shown plexiform neurofibroma in both locations.

Plain skull films were also available for review in seven patients with orbital abnormalities. One examination was normal (case 2). Four cases showed enlargement of the optic canal (cases 1, 4, 5, and 9); case 8 demonstrated a calcified orbital mass and case 21 confirmed the sphenoidal dysplasia.

Intracranial Abnormalities

We found intracranial lesions in 16 patients (cases 5–20). In case 12, CT showed bilateral enhancing masses in the cerebellopontine angle cisterns, and plain skull examination demonstrated bilateral erosion of the internal auditory canals. Although not confirmed histologically, the diagnosis of bilateral acoustic neurinoma is highly probable. CT identified soft-tissue masses in the region of the optic chiasm in seven patients (cases 5–7, 10, 11, 18, and 20). Biopsy results of case 11 indicated a gangioglioma (fig. 5).

CT in six cases demonstrated intraaxial masses. Four showed attenuation less than that of brain tissue (cases 9, 14, 16, and 19) and two showed contrast enhancement (cases 11 and 15). Three cases were biopsied revealing low grade astrocytoma (cases 14–16).

Abnormalities of the ventricular system were seen on three examinations. Two demonstrated hydrocephalus and one demonstrated agenesis of the corpus callosum and a midline frontal cystic mass having the appearance of an arachnoid cyst. We did not encounter the characteristic calvarial lucency in the lambdoid suture [4].

Discussion

When evaluating patients with neurofibromatosis who develop visual loss or proptosis, we found CT of the orbits

TABLE 2: **CT Abnormalities in Neurofibromatosis**

Case No.	Age (yrs), Gender	Symptoms	CT Findings	Other Studies	Confirmation
1	3½, F	Visual loss	R optic nerve mass	Rad: enlarged R optic canal; PEG: neg	OP: R optic nerve mass, neg. chiasm, L optic nerve swelling; BX: astrocytoma
2	2½, F	Visual loss	Infiltrating L orbital mass	Rad: neg	Malignant schwannoma
3	3½, F	Visual loss	L optic nerve mass	PEG: neg chiasm	. . .
4	17, F	Proptosis	Infiltrating R retrobulbar mass; enlarged R globe	Rad: R orbital enlargement	Plexiform neurofibroma with infiltration of choroid layer of the eye
5	3, F	Proptosis	L optic nerve mass; R optic nerve thickening; chiasmal mass	Rad: enlarged R and L optic canals	. . .
6	9, F	Visual loss	R optic nerve mass; chiasmal mass	. . .	. . .
7	17, F	Primary amenorrhea	R optic nerve mass; chiasmal mass	. . .	. . .
8	20, M	Visual loss	Calcified R optic nerve mass; R cerebellar calcification; L choroid plexus calcification at foramen of Munro	Rad: calcified R orbital mass	BX: meningioma
9	61, F	Visual loss	R optic nerve thickening: low attenuation R frontal lesion	Rad: enlarged R optic canal	. . .
10	5, M	Visual loss	Chiasmal mass	. . .	. . .
11	18, F	Seizures, visual loss	Chiasmal, 3d ventricular mass, hydrocephalus	Ventriculography: mass, anterior 3d ventricle	BX: ganglioglioma
12	19, F	Hearing loss	R and L cerebellopontine angle masses	Rad: R and L internal auditory canal erosion	. . .
13	15, F	Ataxia	Hydrocephalus; calcific density in temporal horn of R lateral ventricle	. . .	Irradiation of chiasmal mass age 5 years
14	16, M	Seizures	Low attenuation L temporal mass	. . .	Astrocytoma
15	20, F	Hemiparesis	L temporal enhancing mass	Angiography: L temporal mass	Astrocytoma, grade I or II
16	24, M	Seizures	Low attenuation frontal convexity mass	Angiography: avascular mass	Low grade astrocytoma, pilocytic type
17	13, M	Ataxia	Hydrocephalus	No mass on PEG or angiography	. . .
18	36, F	Hearing loss	Calcified chiasmal mass R thalamic calcification	. . .	. . .
19	30, M	Seizures	Midline frontal low attenuation mass; agenesis of corpus callosum	Angiography: avascular mass; PEG: no communication with ventricles; agenesis of corpus callosum	. . .
20	5, F	Seizures	Chiasmal mass; R temporal encephalomalacia	. . .	CSF cytology; primary nonglial tumor; status postirradiation
21	18, F	Proptosis	Subcutaneous facial and retrobulbar mass; sphenoidal dysplasia	Rad: dysplasia of sphenoid and maxilla; enlargement of orbit and temporal fossa	BX: orbit and face; plexiform neurofibroma

Note.—R = right; L = left; Rad = plain radiography; PEG = pneumoencephalography; neg = negative; OP = operative findings; BX = biopsy findings.

useful in confirming the presence of an optic nerve glioma. An unsuspected lesion of the asymptomatic orbit may also be revealed and the intracranial component may be demonstrated in most cases.

Our findings agree with the CT description of optic nerve gliomas given by Byrd et al. [9]. These are: diffuse thickening of the intraorbital part of the optic nerve, fusiform enlargement of the optic nerve (fig. 1A), or a discrete, focal mass arising from the optic nerve. According to Glaser [10], involvement of the optic chiasm is present initially in many cases and does not represent extension of the orbital lesion. The findings of suprasellar involvement consist of a soft-tissue mass filling the suprasellar cistern (fig. 1B) and/or bilaterally symmetrical area of contrast enhancement in the hypothalamus.

Dysplasia of the greater wing of the sphenoid may be

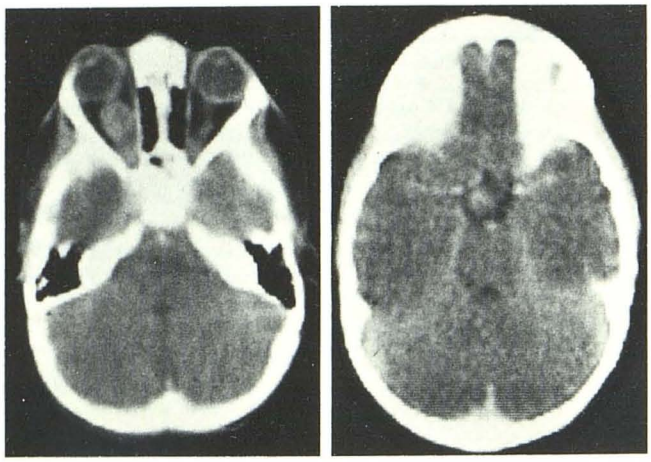

Fig. 1.—Case 5. Serial sections. **A**, Contrast-enhanced. Fusiform mass involves left optic nerve; diffuse thickening of right optic nerve. **B**, Contrast-enhancing soft-tissue mass in suprasellar cistern.

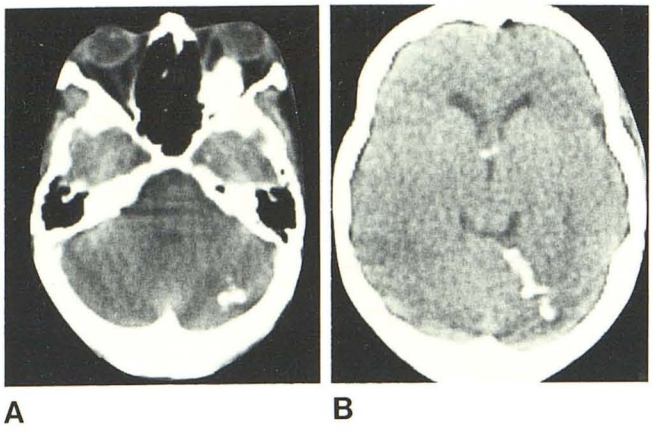

Fig. 3.—Case 8. Optic nerve sheath meningioma. Unenhanced views. **A**, Densely calcified mass along right optic nerve. Calcific area in right cerebellar hemisphere. **B**, Higher section. Extension of cerebellar calcification. Punctate calcification in region of foramen of Munro.

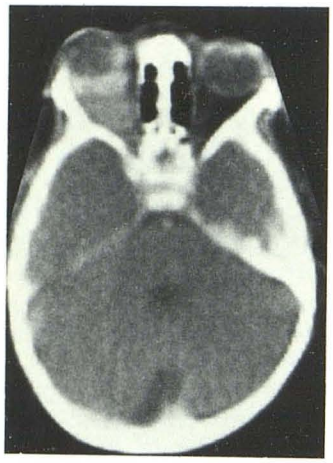

Fig. 2.—Case 2. Malignant schwannoma. Contrast-enhanced scans. Diffuse retrobulbar mass and thick scleral enhancement.

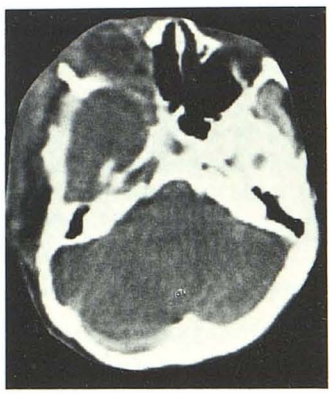

Fig. 4.—Case 21. Plexiform neurofibroma. Contrast-enhanced view. Extensive left subcutaneous facial mass. Inferior displacement of left globe, large left temporal fossa, and absent posterior wall of orbit.

recognized in patients with proptosis [11–13]. CT was especially helpful in predicting a lesion other than the expected optic glioma in three cases (cases 2, 4, and 8), thereby changing the course of the patient's management.

As in the cases autopsied by Pearce [14], the most common intracranial tumor in our series was astrocytoma. Neither the arachnoid cyst nor the associated agenesis of the corpus callosum are recognized as lesions found in neurofibromatosis and they may have occurred coincidentally. A similar case, not identified as neurofibromatosis, was illustrated in a text by Gonzalez et al. [15].

Possible explanations for the calcified lesions encountered in three cases include intraventricular meningioma [16] and hamartomatous lesions [17, 18]. These foci of meningiomatosis are common in neurofibromatosis. They differ from frank neoplasms and they may be calcified.

One case of hydrocephalus may have resulted from irradiation to a chiasmal mass 10 years before the current examination. In the other case, no mass was demonstrated by angiography or pneumoencephalography, and the etiology of the hydrocephalus is unexplained. Hydrocephalus due to aqueductal stenosis has been observed. In one case [6], periaqueductal gliosis was present. In another, aqueductal stenosis was caused by polypoid ependymal granulations.

In contrast to our data, Salvolini et al. [6] reported five cases of retrobulbar expanding masses for which the histology was not specified and they did not mention encountering lesions of the optic chiasm. They described one falx meningioma, five cases of acoustic neuroma, five cases of hydrocephalus, and four other intracranial masses. Their material showed a similarly high incidence of abnormal examinations (26/31 cases).

In our series, 21 of 29 symptomatic patients had abnormal CT scans. In 14 patients, a lesion concordant with the symptomatology was found. In the other seven cases CT demonstrated an asymptomatic abnormality in addition to the clinically suspected lesion. Although plain skull exami-

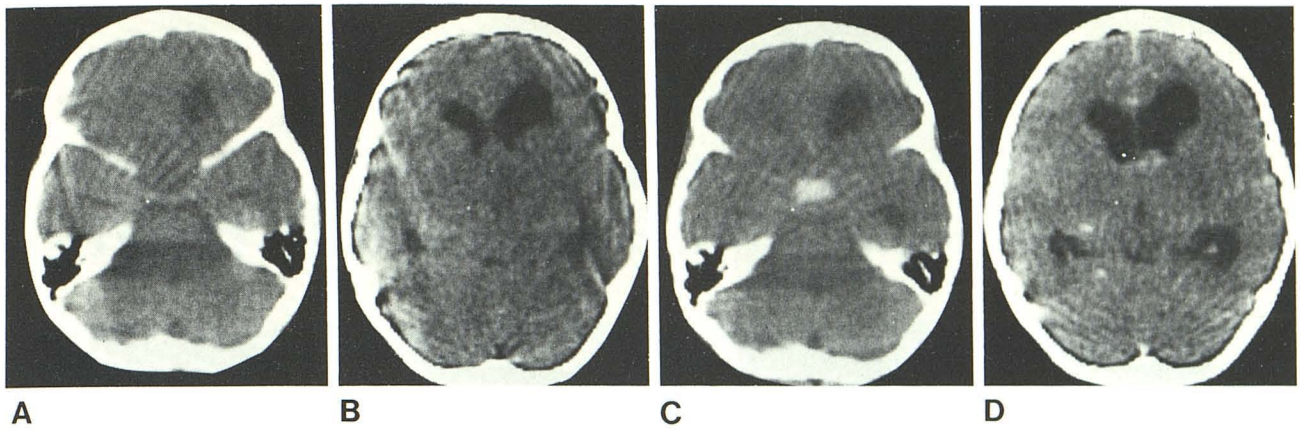

Fig. 5.—Case 11. Ganglioma. **A** and **B**, Unenhanced scans. Suprasellar mass, obliteration of anterior third ventricle, and dilated lateral ventricles. **C** and **D**, Contrast-enhanced scans. Enhancing mass of optic chiasm and partial enhancement of third ventricular mass.

nations were abnormal in six of seven cases in the orbital group, CT provided more specific information regarding the soft-tissue component of the lesions.

Our study population did not include patients who were totally asymptomatic and therefore we were unable to assess the overall incidence of a "central" neurofibromatosis. However, the number of silent lesions discovered in our series and the higher incidence of central nervous system tumors with neurofibromatosis suggests that CT may have a worthwhile role as a screening examination in this disorder.

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