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MR Imaging of the Brain in Patients with Diabetes Insipidus

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Diabetes insipidus is a clinical syndrome characterized by the excretion of copious volumes of dilute urine combined with persistent intake of abnormally large quantities of fluid. There are two general forms of the disease, central (vasopressin deficient) and nephrogenic (vasopressin resistant). Diabetes insipidus of central origin most often results from lesions in the hypothalamic-neurohypophyseal axis. Twenty-six cases of central diabetes insipidus were evaluated with the use of high-field-strength MR imaging. A wide variety of precipitating conditions were found, including Langerhans cell histiocytosis, neoplasia, trauma, and infection. A thickened pituitary infundibulum was seen in most patients, and an absence of high intensity signal in the posterior pituitary lobe on T1-weighted images was seen in every case. Analysis of stalk morphology; associated brain findings; and correlation with the patient's age, sex, history, and radiographs of other body parts improved diagnostic specificity.

When combined with clinical information, MR imaging is able to provide a specific diagnosis in almost all cases of central diabetes insipidus.

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Diabetes insipidus (DI) is a disorder characterized by abnormally large volumes (>30 ml/kg per day) of dilute urine (<250 mosmol/l) and persistent thirst [1]. Probably the most common causative defect is inadequate secretion of antidiuretic hormone (ADH), a disorder variously termed neurogenic, central, or hypothalamic DI. DI can also result from renal insensitivity to the antidiuretic effects of ADH (nephrogenic DI). Patients with central DI who fail to ingest adequate volumes of water (hypodipsia) are at risk for developing life-threatening hypernatremia, and constant care may be required to ensure fluid-electrolyte balance [2]. Symptomatic polydipsia and polyuria can also result from inappropriately excessive fluid intake in the absence of any underlying defect in ADH biosynthesis or release (psychogenic polydipsia), although this cause is very rare [2].

Earlier studies [3] suggested that idiopathic central DI accounted for approximately 50% of all cases. More recent work suggests, however, that head injury and surgery in the sellar area are responsible for a higher percentage of cases. In a study of 119 patients with long-standing (>6 months) central DI, Moses [1] reported that 50% developed DI after head injury or surgery on the pituitary or hypothalamus, whereas 25% had idiopathic central DI. In 8% of the patients, DI developed from cancer metastases to the hypothalamus, 6% of the cases resulted from malignant brain tumors, 6% were related to nontraumatic encephalomalacia, and 3% were related to Langerhans cell histiocytosis [4]. The hereditary forms of central DI are extremely rare, accounting for about 1% of all cases [5]. Other rare causes include thrombotic thrombocytopenic purpura [6], pituitary apoplexy [7], postpartum pituitary necrosis [8, 9], Wegener granulomatosis [10], and systemic blastomycosis [11]. Infiltration of the hypothalamus by amyloid or sarcoidosis has also been reported [2]. Imaging studies (CT or MR) of the hypothalamic-pituitary

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TABLE 1: Clinical and MR Findings in Patients with Diabetes Insipidus (DI)

Cause of DI/Case No.	Age (yr)	Sex	Type of DI; Signs/Symptoms	MR Findings in Hypothalamus/Pituitary	MR Enhancement	Pathologic Documentation/ Further Pathologic Findings
Langerhans cell histiocytosis						
1	29	F	Acute	Symmetric, thickened stalk; no hyperintensity in post pituitary on T1	Homogeneously enhanced thickened stalk	Stalk biopsy
2	14	F	Acute	Symmetric, thickened stalk; no hyperintensity in post pituitary on T1	Enhanced studies not performed	Mastoid biopsy
3	1	F	Acute	Symmetric, thickened stalk; no hyperintensity in post pituitary on T1	Homogeneously enhanced thickened stalk	Skin biopsy
4	57	F	Acute	Symmetric, thickened stalk; no hyperintensity in post pituitary on T1	Homogeneously enhanced thickened stalk and hypothalamic nodule	Stalk biopsy
5	32	F	Acute	Symmetric, thickened stalk; no hyperintensity in post pituitary on T1	Enhanced studies not performed	Lung biopsy
6	28	F	Acute	Symmetric, thickened stalk; no hyperintensity in post pituitary on T1	Enhanced studies not performed	Lung biopsy
Germinoma						
7	19	M	Acute; Parinaud syndrome	No hyperintensity in post pituitary on T1; thickened upper stalk	Enhancement of hypothalamus and R trigonal subependymal nodule; heterogeneously enhanced pineal mass	Pineal biopsy
8	19	M	Acute; Parinaud syndrome	No hyperintensity in post pituitary on T1; thickened upper stalk	Enhanced studies not performed	Pineal biopsy
9	12	M	Acute; Parinaud syndrome	No hyperintensity in post pituitary on T1; thickened upper stalk	Enhanced studies not performed	Pineal biopsy
10	13	M	Chronic	No hyperintensity in post pituitary on T1; thickened upper stalk	Enhancement of hypothalamus and optic chiasm	Hypothalamic mixed germ cell tumor
Craniopharyngioma						
11	10	M	Acute	Hypothalamic mass; no hyperintensity in post pituitary on T1	Enhanced cystic mass occupying third ventricle	-
12	8	F	Acute; headache	Hypothalamic mass; no hyperintensity in post pituitary on T1	Enhanced studies not performed	-
Hypothalamic glioma						
13	12	M	Acute; visual field defect	Hypothalamic mass; no hyperintensity in post pituitary on T1	Enhanced studies not performed	-
Tuberculosis						
14	17	F	Acute transient; seizures	Symmetric, thickened stalk; no hyperintensity in post pituitary on T1	Abnormal enhancement in basal cisterns	-
Sarcoidosis						
15	24	M	Chronic	Symmetric, thickened stalk; no hyperintensity in post pituitary on T1	Enhanced studies not performed	-
Metastasis						
16	57	M	Transient; visual defect	Sellar/suprasellar mass; no hyperintensity in post pituitary on T1	Inhomogeneously enhancing mass	Oat cell lung carcinoma
17	69	M	Transient; visual defect	Sellar/suprasellar mass; no hyperintensity in post pituitary on T1	Inhomogeneously enhancing mass	Adenocarcinoma
Sellar surgery						
18	42	F	Transient	Postop sellar change; marked structural distortion with high signal due to methemoglobin in sella	Enhanced studies not performed	-
19	37	M	Transient	Postop sellar change; marked structural distortion with high signal due to methemoglobin in sella	Enhanced studies not performed	-
20	47	M	Transient; fever	Postop sellar change; marked structural distortion with high signal due to methemoglobin in sella	Multiloculated enhancing pituitary mass	Pituitary mass

Table 1 continues

TABLE 1—Continued

Cause of DI/Case No.	Age (yr)	Sex	Type of DI; Signs/Symptoms	MR Findings in Hypothalamus/Pituitary	MR Enhancement	Pathologic Documentation/ Further Pathologic Findings
Transection stalk after trauma						
21	17	F	Transient; amenorrhea	Transection of stalk; aberrantly placed hyperintensity in median eminence of post pituitary after resolution of DI	Enhanced studies not performed	—
22	14	M	Transient	Transection of stalk; no hyperintensity in post pituitary on T1	Enhanced studies not performed	—
Erdheim-Chester disease						
23	39	M	Chronic	No hyperintensity in post pituitary on T1; thickened upper stalk	Homogeneously enhanced thickened median eminence	—
Unknown						
24	25	F	Acute	Symmetric, thickened stalk; no hyperintensity in post pituitary on T1	Enhanced studies not performed	—
25	29	F	Acute	Symmetric, thickened stalk; no hyperintensity in post pituitary on T1	Homogeneously enhanced thickened stalk	—
26	52	M	Chronic	Empty sella; downward displaced chiasm; no hyperintensity in post pituitary on T1	Enhanced studies not performed	—

Note.—post = posterior; T1 = T1-weighted images; R = right; postop = postoperative.

region are important in the diagnosis of DI. MR imaging is particularly useful in documenting the presence or absence of a structural lesion, as opposed to assigning a diagnosis of idiopathic DI for which only symptomatic therapy is prescribed.

We report our MR imaging findings in 26 patients with clinically proved DI.

Materials and Methods

We retrospectively studied the MR findings in 26 patients (14 males and 12 females 1–69 years old; mean, 27.7 years of age) seen during the period 1986–1990 with clinical evidence of DI. Clinical histories were correlated with imaging findings. All patients were imaged on a 1.5-T General Electric system. Eleven patients were scanned before and after IV administration of gadopentetate dimeglumine. Surgical specimens were obtained from the hypothalamic-pituitary area or other affected body sites in 19 patients.

The MR technique consisted of unenhanced T1-weighted spin-echo (SE) scans, 600/20/4 (TR/TE/excitations), through the sella in sagittal and coronal planes with 3-mm slices and a 0.5-mm interslice gap. Enhanced studies were obtained in the same planes. Images were reconstructed with two-dimensional Fourier transform on a 256 × 256 image matrix. Axial T2-weighted images (2000/40,80/2) of the whole brain were obtained also in most patients. Additional axial T1-weighted images (600/20/4) of the brain were obtained in 11 patients after contrast administration.

Unenhanced and enhanced images were reviewed by two neuro-radiologists. The size, shape, and signal intensity of the pituitary stalk, hypothalamus, and pituitary gland and any associated abnormalities were evaluated. MR findings were correlated with clinical history and surgical pathology.

Results

Results of MR examinations, correlated with the clinical symptoms and signs, associated imaging findings, and surgical pathology, are summarized in Table 1.

Langerhans Cell Histiocytosis

Six female patients 1–57 years old with acute onset and persistent DI were found to have uniformly thickened pituitary stalks 3–4 mm wide in the coronal plane and 3–17 mm wide in the sagittal plane (Fig. 1) (stalks in normal controls were less than 2.8 mm in both planes). One patient had a destructive mastoid lesion. Another had clinical Letterer-Siwe disease (with skin rash and hepatosplenomegaly). Two patients had honeycomb-like interstitial lung disease and the other two had thickened pituitary stalks alone. In none of the patients was high signal intensity seen in the posterior lobe. Enhanced MR was performed in three patients. In two there was normal enhancement of the stalk (cases 1 and 3); in the third (case 4), an enhancing nodule in the hypothalamus proved to be Langerhans cell histiocytosis.

Germ Cell Tumor

Abnormally enlarged upper pituitary stalks were seen in four patients 12–19 years old with acute onset and persistent DI. Coexisting lobulated pineal masses were present in three patients. Abnormal contrast enhancement of the subependymal lining in the anterior recess of the third ventricle from CSF metastasis was seen in one patient (Fig. 2). In another patient, an isolated hypothalamic mass showed enhancement (Fig. 3).

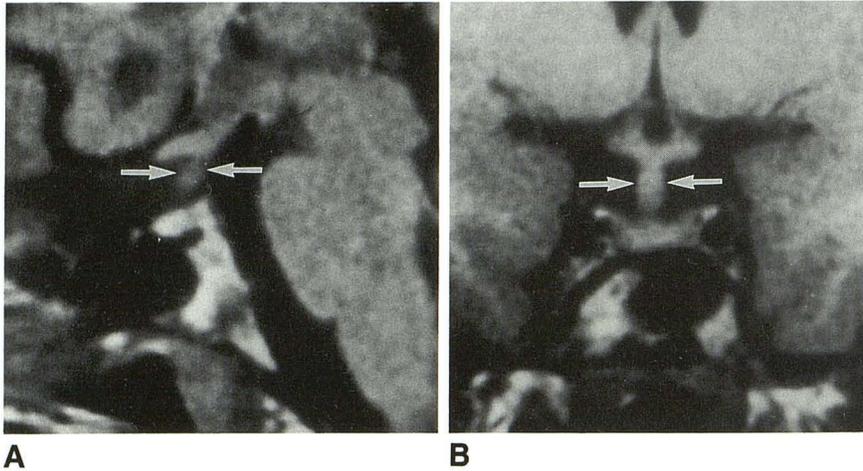


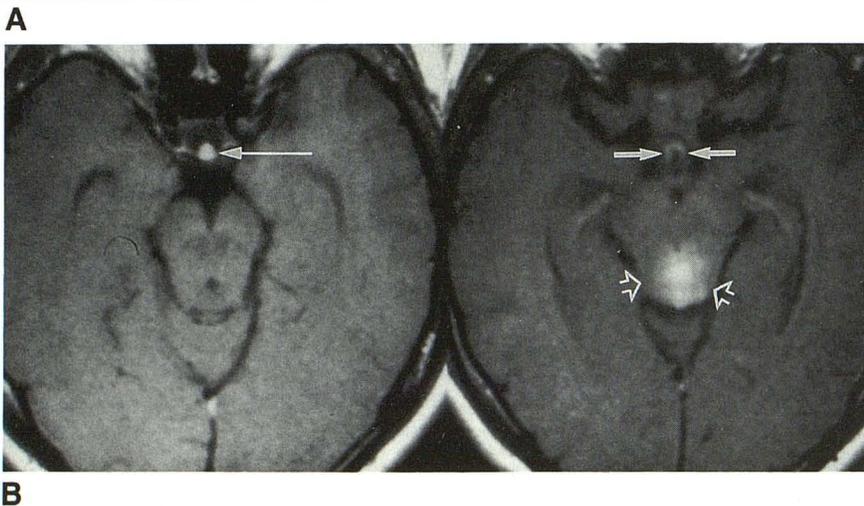
Fig. 1.—Case 5: Uniformly thickened pituitary infundibulum from Langerhans cell histiocytosis. A and B, Sagittal (A) and coronal (B) T1-weighted images (600/20) reveal uniformly thickened (4 mm) pituitary infundibulum (arrows). No posterior pituitary hyperintensity was observed.



Fig. 2.—Case 7: Hypothalamic metastasis from pineal germinoma.

A, Sagittal SE 600/20 images before (left) and after (right) infusion of gadopentetate dimeglumine show heterogeneous enhancement of pineal mass (open arrows) and enhancing nodule in median eminence (long solid arrow). Pituitary infundibulum is asymmetrically thickened (short solid arrows).

B, Axial SE 600/20 images after infusion of gadopentetate dimeglumine reveal enhancing pineal tumor (open arrows) as well as subependymal metastasis in anterior recess of third ventricle (short solid arrows) and thickened pituitary infundibulum (long solid arrow).



Craniopharyngioma

Craniopharyngiomas were found in two patients (10-year-old boy and 8-year-old girl) with acute onset and persistent DI. In one patient (case 12), a large, rather homogeneous sellar and suprasellar mass was seen that obliterated the anterior third ventricle. Another patient (case 11) with a history of craniopharyngioma was admitted because of sudden onset of DI. After enhancement, MR revealed enhancing sellar and

suprasellar masses with a large multicystic lesion that filled the posterior third ventricle, representing the recurrent craniopharyngioma.

Hypothalamic Glioma

A 12-year-old boy (case 13) with acute onset of DI and a visual field defect was evaluated by MR. A 3-cm hypothalamic

Fig. 3.—Case 10: Hypothalamic mixed germ cell tumor.

A, Sagittal SE 600/20 image reveals hypothalamic mass involving optic chiasm (straight arrow) and median eminence (curved arrow). Pituitary infundibulum is asymmetrically thickened.

B, Coronal SE 600/20 image after infusion of gadopentetate dimeglumine shows strong but slightly heterogeneous enhancement of hypothalamic mass with involvement of median eminence (arrows).

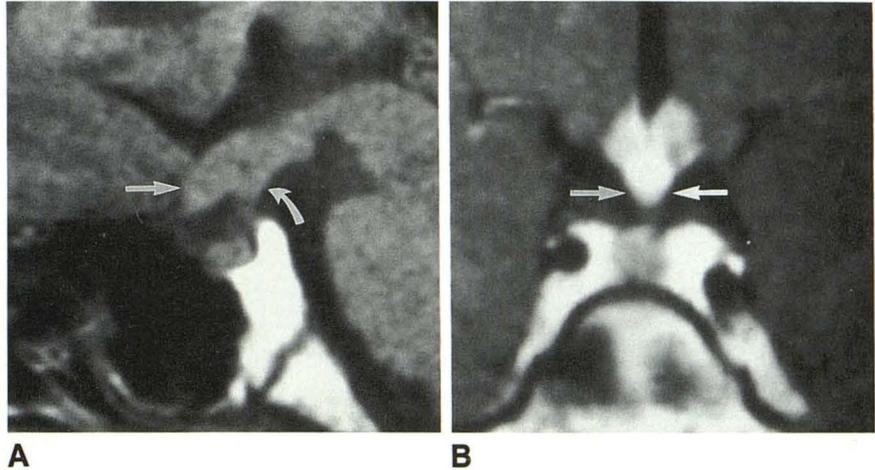
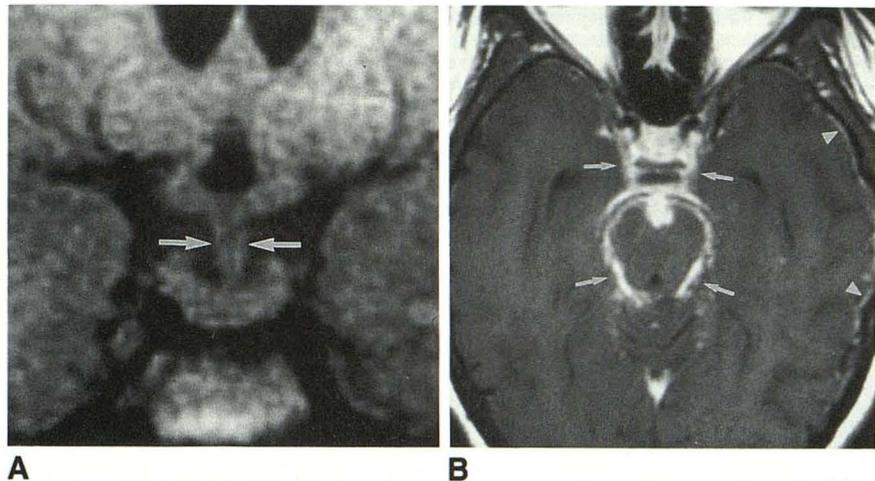


Fig. 4.—Case 14: Uniformly thickened stalk from tuberculosis.

A, Coronal SE 600/20 image reveals uniformly thickened (4 mm) pituitary infundibulum (arrows).

B, Axial SE 600/20 image after infusion of gadopentetate dimeglumine shows abnormal enhancement of suprasellar and circumscencephalic cisterns (arrows) as well as of leptomeninges (arrowheads).



lesion was detected. It was isointense on T1-weighted images and hyperintense on T2-weighted images. There was some mass effect on the chiasm.

Tuberculosis and Sarcoidosis

The patient with tuberculosis was a 17-year-old girl admitted with seizures and acute onset of DI. MR showed a uniformly thickened stalk (Fig. 4A). After administration of contrast material, there was diffuse enhancement in the basal cisterns (Fig. 4B). A spinal epidural abscess was noted also. CSF revealed tuberculosis bacilli. After antituberculous treatment, the DI resolved. The other patient (a 24-year-old man) was known to have a history of pulmonary sarcoidosis with chronic DI. MR revealed a uniformly thickened stalk, presumably representing neurosarcoidosis.

Metastases

Two patients (a 57-year-old man and a 69-year-old man) with transient DI and visual defects were evaluated. MR showed destructive, inhomogeneously enhancing sellar and suprasellar masses in both cases (Fig. 5). Systemic examinations revealed the primary carcinomas.

Postoperative Sella

Transsphenoidal hypophysectomy for pituitary adenoma had been performed recently in three patients (cases 18–20). Transient DI was noted in each case. T1-weighted MR images within 2 weeks after surgery in cases 18 and 19 showed an area of signal hyperintensity in the pituitary fossa with associated mass effect. This most likely represented methemoglobin accumulation in the surgical bed, a fairly typical feature of the early postoperative sella (Fig. 6). No treatment was given in case 18 or 19. In case 20, a patient with severe headache and fever, enhanced MR images revealed a multiloculated enhancing sellar mass (Fig. 7); pituitary abscess was suspected. DI disappeared after drainage.

Post-Traumatic Transection of the Stalk

Two patients (cases 21 and 22) had been injured in car accidents and had a history of transient DI. One patient (case 21) also had amenorrhea. In case 21, MR performed 2 years after the accident showed a transected stalk with bright signal at the proximal stalk stump. In this patient, DI had resolved completely by the time of MR imaging (Fig. 8). In the other patient (case 22), MR performed 3 months after the car accident showed a ruptured anterior third ventricle and a

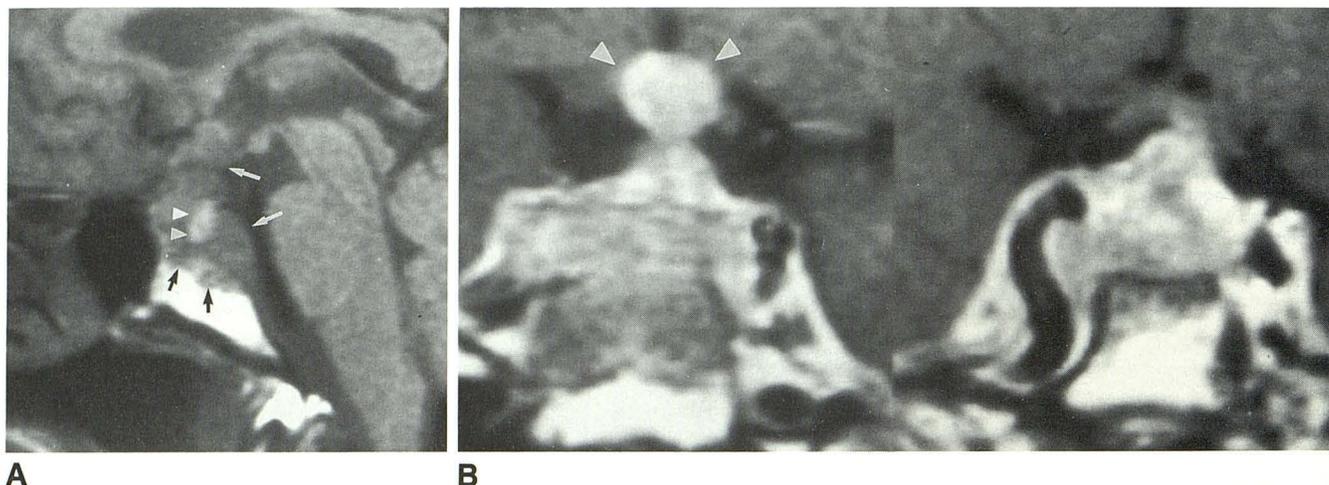


Fig. 5.—Case 16: Sellar and suprasellar mass from lung cancer.

A, Sagittal SE 600/20 MR image reveals an inhomogeneous sellar mass with suprasellar extension (white arrows). Irregular border of pituitary floor (black arrows) is attributable to tumor invasion. Central high signal intensity (arrowheads) most likely represents methemoglobin from intratumoral hemorrhage.

B, Coronal SE 600/20 contiguous MR images after infusion of gadopentetate dimeglumine show inhomogeneous enhancement of this sellar and suprasellar mass. Hypothalamus is involved (arrowheads).

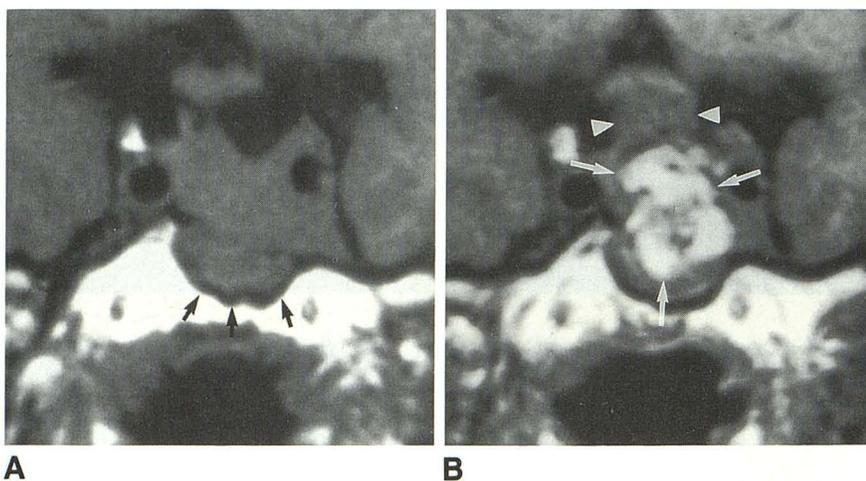


Fig. 6.—Case 18: Pre- and postoperative sella.

A, Preoperative coronal SE 600/20 MR image shows irregularly shaped pituitary macroadenoma with depression of floor of sella (arrows).

B, Postoperative coronal SE 600/20 MR image 2 weeks after surgery. Large hyperintense area in sella with associated mass effect (arrows) probably represents methemoglobin accumulation in surgical bed. Tumor or residual gland appears to have been displaced into suprasellar region (arrowheads).

completely transected stalk. The bright spot was not identified at any location. The patient continued to have DI for several months after the MR study.

Erdheim-Chester Syndrome

A 39-year-old man had an 8-year history of bone pain and DI. Enhanced MR showed a thickened enhancing upper pituitary stalk (Fig. 9A); the bone scan showed symmetric diffuse osteoblastic changes in peripheral long bones (Fig. 9B). Bone biopsy showed changes typical of Erdheim-Chester disease.

Unknown Etiology

Two young women 25 and 29 years old (cases 24 and 25, respectively) had acute onset and persisting DI. MR showed a uniformly thickened stalk in each case. No other abnormal-

ities were noted. In several respects the MR findings were similar to those in case 1, a patient in whom Langerhans cell histiocytosis was diagnosed by biopsy. Since no biopsy data were obtained, these two patients were categorized as unknown. In another patient (case 26), a 52-year-old man with chronic DI, an empty sella and a downwardly displaced chiasm were seen on MR.

In summary, a total of 10 cases of uniformly thickened pituitary infundibulum were seen in six patients with Langerhans cell histiocytosis, one patient with tuberculosis, one patient with sarcoidosis, and two patients with DI of unknown origin. A thickened upper stalk was seen in the four patients with germ cell tumor and the one patient with Erdheim-Chester syndrome. In the remaining patients, the integrity of the infundibulum was either directly compromised by the tumor itself, distorted by surgery, or transected as a result of

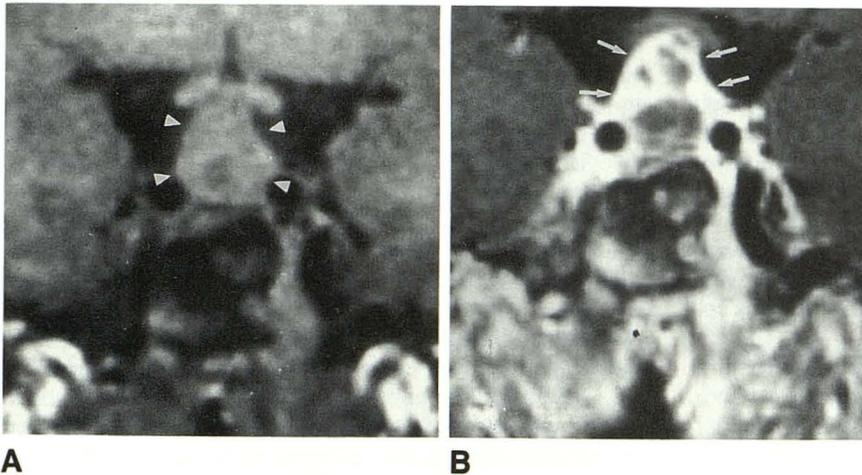


Fig. 7.—Case 20: Postoperative pituitary abscess.
A, Coronal SE 600/20 MR image shows inhomogeneous pituitary mass with suprasellar extension (arrowheads).
B, Coronal SE 600/20 MR image after infusion of gadopentetate dimeglumine shows abnormal multiloculated enhancement pattern of this mass (arrows).

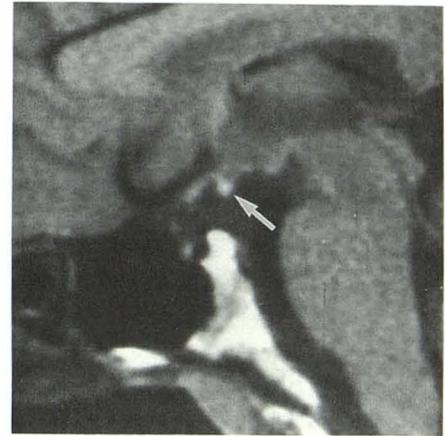
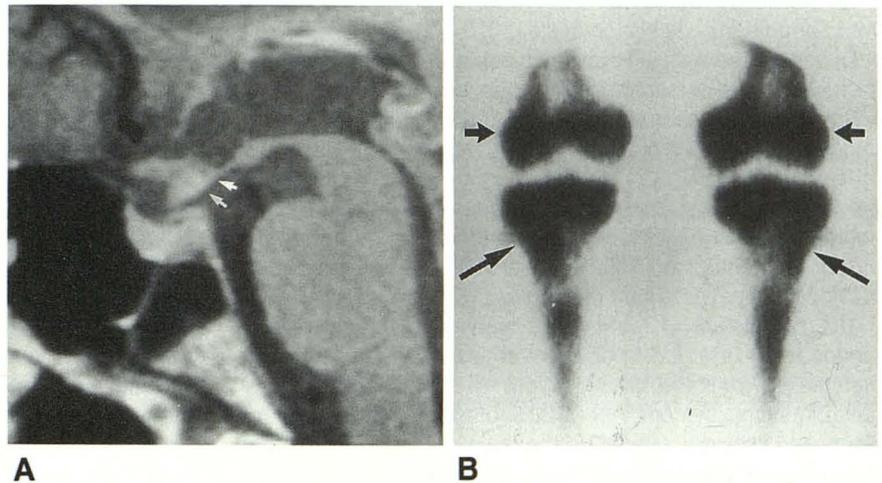


Fig. 8.—Case 21: Transection of pituitary infundibulum resulting from car accident accompanied by transient diabetes insipidus. Sagittal SE 600/20 MR image shows transected pituitary infundibulum and hypoplastic pituitary gland. High-signal focus at median eminence (arrow) represents the same material noted in normal high-signal posterior pituitary. Diabetes insipidus had resolved completely by the time this image was obtained.

Fig. 9.—Case 23: Erdheim-Chester disease.
A, Sagittal SE 600/20 MR image after infusion of gadopentetate dimeglumine shows enhancement in a thickened median eminence (arrows) of pituitary infundibulum.
B, ^{99m}Tc bone scan reveals increased osteoblastic activity bilaterally in femur (short arrows) and tibia (long arrows).



mechanical trauma in the region; thus, identification of an isolated thickened infundibulum was not possible.

The normally hyperintense posterior pituitary region was not observed in 22 patients. In three postoperative cases, postsurgical changes made identification of the posterior lobe signal intensity impossible to establish. In another patient with a history of transient DI, an aberrant bright spot was observed at the median eminence after resolution of DI.

Discussion

In the vast majority of cases, the diagnosis of DI is made on clinical grounds. The role of imaging is to determine the cause of DI, and the available data indicate that MR is currently the best imaging technique for this purpose. MR

images clearly demonstrate hypothalamic-pituitary morphology and can detect subtle abnormalities in the region of the sella. A diagnosis of idiopathic DI should be considered only after appropriate MR imaging has been completed and the presence of structural lesions excluded.

Approximately two thirds of patients with central DI have infiltrative lesions involving the hypothalamic-neurohypophyseal axis [1]. Langerhans cell histiocytosis was the single leading cause of DI in the present study, accounting for 23% of cases. All patients with this etiology were females with DI of acute onset that persisted despite treatment. A review of 180 patients with multifocal eosinophilic granulomas indicated that 56% had DI, 25% had skin manifestations, 15% had pulmonary infiltrates, and 15% had otitis media [12]. This suggests that imaging studies of the lungs, temporal bones, or even the peripheral long bones may be useful diagnostic

adjuncts in patients with Langerhans cell histiocytosis (cases 2, 3, 5, and 6). At the same time, it is also noteworthy if a thickened stalk is the only presentation of Langerhans cell histiocytosis (such as in case 1). In two other patients (both females), MR findings were similar. Although these cases (cases 24 and 25) were categorized as DI of unknown origin based on the patients' ages, histories, and imaging results, Langerhans cell histiocytosis may have been responsible for the DI in these cases also. As well, some patients with thickened stalks noted on CT, who were listed as having idiopathic DI [1], may also represent cases of Langerhans cell histiocytosis.

Germ cell tumor was found in the next largest group of patients with DI and was the most common intracranial tumor associated with central DI in children, in agreement with prior reports [13]. All of our patients were teenage boys. Three had a pineal mass with CSF metastasis and Parinaud syndrome (paralysis of upward gaze). MR clearly demonstrated a thickened upper pituitary stalk and lobulated pineal mass. Enhanced MR revealed enhancing CSF metastases (case 7) and suprasellar hypothalamic enhancing mixed germ cell tumor (case 10). In a previous study, dysgerminoma was found to be the most common intracranial tumor associated with neurogenic DI in children, with craniopharyngioma the second most common [13]. DI has been reported in 40% of children with Langerhans cell histiocytosis [13].

DI associated with infiltrative inflammatory processes such as tuberculosis or sarcoidosis is also well established [14]. MR revealed a uniformly thickened stalk resembling that in Langerhans cell histiocytosis, which has been reported by other investigators also [14]. However, the chest radiographs provided crucial evidence for the diagnosis. Transient DI was noted in a patient with tuberculosis meningitis, as evidenced by contrast-enhanced MR. Transient DI has been reported previously in patients with meningitis [15, 16].

The prevalence of metastases within the pituitary gland ranges from 1.8–12% in different series. Most common are metastases from the breast, followed by carcinoma of the gastrointestinal system. About 20% of these metastases are diagnosed clinically with DI as the main presenting symptom [17]. DI of a transient nature may represent metastases to the posterior lobe of the pituitary without significant encroachment on the supraoptic and paraventricular nuclei of the hypothalamus. In the present study, MR revealed the destructive nature of such lesions. Therefore, in older individuals in particular, metastases merit some consideration in the diagnosis.

An association between transient DI and the postoperative sella is not uncommon. It appears to represent selective damage to the posterior lobe without concurrent damage to the hypothalamic neurosecretory nuclei. DI is often associated with persistent anterior lobe dysfunction from surgical damage. The presence of transient DI with fever, headache, visual disturbances, and anterior lobe dysfunction in a patient who recently underwent sellar surgery strongly suggests pituitary abscess [18]. A loculated ring-enhancing sellar mass on MR with clinical symptomatology of DI is diagnostic.

Posttraumatic transection of the stalk with DI has been

described in previous MR studies [19]. In most cases the DI develops within a few days after stalk section and is transient, but in one third of the patients it remains as a chronic complication without concurrent dysfunction of the anterior lobe [20]. Differential diagnosis from congenital pituitary dwarfism is based on the clinical history. It has been reported that dwarfs have a hypoplastic stalk and anterior pituitary lobe [21].

An interesting etiologic combination was seen in the patient with Erdheim-Chester disease and central DI. Erdheim-Chester disease is a rare entity, typically represented by symmetric osteoblastic changes in peripheral long bones with lipid-laden histiocytes and giant cells in bones and visceral organs [22, 23]. One case of Erdheim-Chester disease involving the brain parenchyma was described [24] in which enhancement on MR persisted 7 days after administration of contrast material. However, involvement of the hypothalamus with associated DI has not been reported previously, which is somewhat surprising given the highly infiltrative nature of this disease. Of some interest also is the MR finding in the patient in our study in whom enhancement was no longer seen 1 day after contrast administration. The explanation for this observation is not known.

Another patient had chronic DI and an empty sella with herniation of the optic chiasm. Hypopituitarism, with or without visual field defects secondary to herniation of the suprasellar visual system and third ventricle into the empty sella, has been reported previously [25]. However, the cause of DI in our particular patient remains unknown.

The median age of onset of idiopathic DI in a previous study [1] was 16 years, and the majority of patients (63%) were male. Most of these patients had evidence of anterior pituitary dysfunction, and some had thickened infundibular stalks on CT. Growth hormone deficiency is the most common accompanying anterior pituitary deficit, but inadequate secretion of thyroid-stimulating hormone (TSH) and adrenocorticotropic hormone (ACTH) is also seen [13]. Patients with idiopathic central DI have been shown in pathologic studies to have a striking depletion of Nissl granules and fewer neurons in the supraoptic and paraventricular nuclei of the hypothalamus [5]. Some patients with idiopathic DI have circulating antibodies to vasopressin neurosecretory cells [1], which suggests a possible immunologic basis for the disease. Idiopathic DI is almost always permanent [1]. About 30% of all cases of DI in children are classified as idiopathic [13]. Familial DI is clinically similar to idiopathic DI, but is very rare [5, 13]. Both autosomal-dominant and sex-linked modes of inheritance have been postulated [26].

In children, neurogenic DI affects both sexes equally, and may begin at any age from a few months to adolescence. Polyuria is unusual in neonates, who usually present with chronic dehydration, unexplained fever, vomiting, and constipation, often leading to serious long-term neurologic deficits [13]. In adolescents, polyuria is the main symptom and often has a dramatic onset. Growth hormone deficiency is the most common accompanying anterior pituitary deficiency in children with neurogenic DI, but deficiency of TSH and ACTH secretion may occur also. Inadequate ACTH synthesis and

release may require special attention in patients with DI because it can camouflage the underlying polyuria of DI [13]. Another frequent cause of childhood central DI is a hypothalamic tumor, such as a craniopharyngioma or a hypothalamic glioma [4, 27]. In the present study, MR was able to clearly demarcate the diencephalic mass in different planes (cases 11–13).

In all the patients with DI, the high-intensity signal normally seen in the posterior lobe on T1-weighted MR images was not observed, in agreement with previous reports [28, 29]. In normal subjects, the frequency of the bright signal has varied from 90–100% [28, 29] to as low as 63% [30]. An age-related decline in the rate of detection of the bright signal of approximately 1% per year between 7 months and 85 years of age has also been reported [30]. The source of the hyperintense MR signal in the normal neurohypophysis has been the subject of recent controversy [28–33]. A recent study [33] investigated the effect of two chemical components of the posterior lobe, phospholipids and vasopressin, on the observed MR signal. It was found that a solution containing phospholipid vesicles has T1 and T2 characteristics analogous to those of the neurohypophysis, and that this solution exhibits a single peak that is chemically shifted with respect to water. Vasopressin had no effect on the signal, either in solution or in the vesicles. These data suggest that phospholipids act as a relaxation enhancer of water protons, and that the MR characteristics of the phospholipid vesicles are consistent with the observed MR properties of the neurohypophysis.

The absence of the hyperintensity that is normally seen in the posterior lobe of the pituitary gland is not in itself diagnostically significant, as this bright signal is occasionally absent in some normal subjects [31]. However, we believe that the presence of a bright signal is inconsistent with the diagnosis of DI. We have encountered one case (described below) in which the finding of normal bright signal in the posterior lobe in a patient thought to have DI led to an alternative diagnosis.

We saw one woman who developed DI and preeclampsia late in the third trimester of pregnancy. MR imaging of the hypothalamic-pituitary area was performed 1 month postpartum to exclude a structural lesion. The scan was remarkable in that it was entirely normal, including the presence of hyperintensity in the posterior lobe. We believe that this finding was inconsistent with the diagnosis of vasopressin deficiency. Subsequent testing documented the presence of vasopressinase in her serum. Serum vasopressinase is an enzyme that occasionally appears late in pregnancy [34, 35]. It results in the inactivation of circulating vasopressin and leads to polyuria/polydipsia syndrome, which is indistinguishable from DI [34–36]. This relatively rare diagnosis can be established by incubating pharmacologic preparations of vasopressin with the test patient's serum and demonstrating rapid inactivation of the hormone [34, 35].

In conclusion, central DI can result from a wide variety of diseases. The most common causes of DI appear to be Langerhans cell histiocytosis, germinoma, craniopharyngioma, hypothalamic glioma, granulomatous inflammation, surgery, and trauma. MR is currently the imaging method of

choice in the evaluation of dysfunction of the hypothalamic-neurohypophyseal secretory system. MR imaging, particularly when coupled with clinical information, allows a specific diagnosis in almost all cases of central DI.

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