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PSYCHIATRIC DIAGNOSES IN HYPOTHALAMIC HAMARTOMA PATIENTS

COMMENTS

Since initating the hypothalamic hamartoma consortium at our institution in 2003, we have operated and treated more than 100 patients with this condition. Barrow has now acquired the largest institutional experience with the surgical treatment of these rare developmental abnormalities. Overall, improvements in surgical technique have improved outcomes, eradicating or ameliorating the intractable epilepsy too often associated with these lesions.

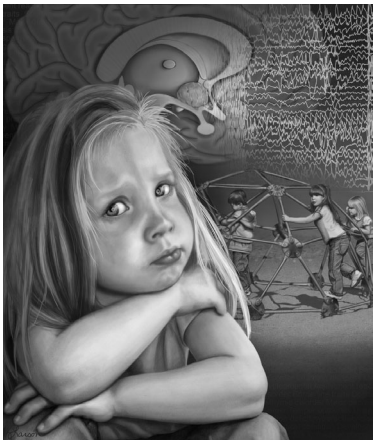
Nonetheless, the behavioral and psychiatric disorders associated with hypothalamic hamartomas can be as devastating to patients and their families as the seizures. Not all patients may regain a normal level of function. As of yet, however, no neuropsychological tool is available for the assessment of children and adults with a hypothalamic hamartoma. Despite efforts in the neuropsychological community to relate these patients' psychiatric and behavioral disorders to their seizure history, no significant relationship has been found. The lack of this information impedes the development of neuropsychological treatments or rehabilitation best suited to help patients and families.

Dr. Prigatano, Director of the Division of Neuropsychology, and colleagues are therefore pursuing this under-researched aspect of hypothalamic hamartomas. In this issue, they report the results of a preliminary study documenting the variability of the diagnostic terms used to describe 57 children and adults with hypothalamic hamartomas and refractory epilepsy. Their goal was to explore the variability of diagnostic terms applied to these patients by previous clinicians to identify any relationships between these patients' psychiatric and behavioral diagnoses and estimates of their intellectual functioning. As this group notes, systematic neuropsychological studies are needed help explain the various psychiatric disturbances seen in patients with this unfortunate condition.

Also in this issue, Aliabadi et al. review a case that will alert clinicians to the diagnostic similarities between diffuse idiopathic skeletal hyperostosis and ankylosing spondylitis and how to distinguish between the overlapping clinical features associated with these two conditions. Despite the clinical similarities of these two pathologies, their treatments differ considerably. Consequently, an early and accurate diagnosis is crucial to the successful management of patients with severe cervical ankylosis.

In another article, Wait and coworkers present clinical images from a patient originally treated for symptoms of tectal compression and intermittent hydrocephalus. Postoperatively, the patient developed a de novo simple cerebellar cyst. This entity was new to our institution and does not appear to have been reported previously. Finally, readers will find helpful indices of all the articles published in 2005 and 2006. We hope our readers find these articles interesting and out of the ordinary. As always, we are pleased to share our clinical findings with students of the neurosciences. However, we need your help too. Please consider using the enclosed self-addressed and stamped envelope to forward a tax-deductible donation that will help us continue to provide this journal free of charge. Thank you for your help.

Robert F. Spetzler, MD
Editor-in-Chief



This issue's cover illustrates a child with emotional and social difficulties associated with a hypothalamic hamartoma. This benign tumor, which involves the hypothalamic nuclei and causes gelastic seizures, can lead to various behavioral, psychiatric, and cognitive disorders. See the article by Prigatano et al. on page 4. The illustration is by Kristen Larson.

Psychiatric Diagnoses Applied to Children and Adults with Hypothalamic Hamartomas and Refractory Epilepsy

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Records of 57 children and adults with a history of HH and refractory epilepsy were reviewed. The psychiatric diagnoses and behavioral descriptions made by previous examiners were recorded. An attempt was made to relate those diagnostic observations to the patient's level of estimated intelligence when examined. Most patients were described as having a psychiatric or behavioral disorder (47 of 57 patients; 82.5%). The problems were heterogeneous, ranging from mild mood disorders to psychosis; oppositional-defiant disorder and attentional deficit disorder were frequently mentioned. There was, however, no relationship between patients' estimated level of intelligence and their psychiatric diagnosis, except in cases of mental retardation. Patients with estimated normal intelligence tended not to receive a psychiatric diagnosis (5 of 15 patients; 30%).

Key Words: epilepsy, hypothalamic hamartoma, intelligence, psychiatric disorders

Abbreviations Used: HH, hypothalamic hamartoma; IQ, intelligent quotient

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The behavioral and psychiatric disorders associated with HH and refractory epilepsy can be as devastating to patients and families as their cognitive disorders and associated epilepsy. To date, efforts to relate the psychiatric and behavioral disorders of these patients to their seizure history have failed to find any significant relationships.[10] Detailed case analyses of aggressive behavior in HH patients with epilepsy have emphasized the multifactorial nature of the aggressive or angry outbursts of HH patients and the multiplicity of diagnoses that can be applied to a given HH patient.[6] However, most reports explicitly or implicitly suggest that the cognitive limitations of patients underlie their limited coping skills.[2,4,6,10] Thus, there might be a relationship between patients' cognitive status and their psychiatric and behavioral characteristics.

The limited literature on the psychiatric features of HH patients also suggests that the diagnosis of attentional deficit disorder and oppositional-defiant disorder are common in this patient group.[10] Again, however, multiple diagnostic terms have been used to describe these patients.

We recorded the psychiatric diagnostic terms and behavioral descriptions found in the records of 57 children and adults with HH and refractory epilepsy. Our intent was to document the variability of diagnostic terms that previous examiners have applied to this patient population and to determine if a relationship could be established between the psychiatric and behavioral diagnoses applied to these patients and estimates of their current intellectual ability.

Methods

Subjects

Between March 2003 and October 2005, 57 patients with HH and refractory epilepsy were referred for a neuropsychological examination as a part of their preoperative evaluation at our institution. Patients ranged from 5 to 55 years old when examined. An arbitrary cut-off age of 5 years was established so that reliable estimates of intelligence could be obtained on all patients using the same psychometric measures. When evaluated, 32 patients (56.1%) were between the ages of 5 and 14 years old.

Record Review

Before patients underwent neuropsychological testing, their available medical records were reviewed. Psychiatric diagnoses and behavioral disturbances recorded by previous examiners were identified. When records regarding psychiatric history were unavailable or limited, an attempt was made to obtain additional data by obtaining release of information from either the patient or family, as appropriate. Records were available on each patient, but not every patient had a psychiatric diagnosis applied to them. Many patients had more than one psychiatric diagnosis or description of their behavioral abnormalities.

Estimating Level of Intelligence

All HH patients underwent a neuropsychological examination. Many of these patients had significant cognitive and behavioral difficulties that made a lengthy neuropsychological examination impractical, if not impossible.[5] Consequently, an effort was made to administer to each patient at least three subtests of the Wechsler Scale of Intelligence (Wechsler Intelligence Scale for Children-III and IV[7,9] and Wechsler Adult Intelligence Scale-3rd Edition[8] to estimate Verbal Comprehension skills (i.e., Vocabulary), Perceptual Reasoning or Organizational skills (i.e., Block Design), and Processing Speed (i.e., Digit Symbol).

Patients were then classified into three groups. The first group, or Neuropsychology Type 1, were subjects with mean scores on the Vocabulary and Block Design subtests within the average range or higher (e.g., a scaled score of 8 or higher). The second group, or Neuropsychology Group 2, consisted of individuals who had either an average score on Vocabulary or Block Design (i.e., a scaled score of 8 or higher) and a below average score on the other subtest (i.e., a scaled score of 7 or below).

The third group, or Neuropsychology Type 3 patients, were patients who obtained scaled scores of 6 or below on the Vocabulary and/or Block Design subtests. This group performed at a level compatible with at least mild mental retardation. The cognition of an additional group of patients was so severely impaired that they could not adequately take these three subtests. The "testable" mentally retarded patients were identified as Type 3a. The "untestable" mentally retarded patients were identified as Type 3b.

Statistical Analyses

If multiple diagnoses were applied to a given patient, all terms were included in the analysis. A frequency table, which listed the diagnostic and descriptive terms for patients falling within the different neuropsychological types noted above, was established.

A qualitative analysis of descriptive and diagnostic terms that clustered with one another was then performed to determine if there was a relationship between a general class of psychiatric and behavioral disturbances and patients' overall cognitive status. Finally, the frequency that a diagnostic term was applied relative to all of the diagnostic terms was calculated, as well as the frequency with which that diagnostic term was applied to the 57 patients.

Results

Demographic and Clinical Features of Neuropsychology Types

Table 1 summarizes the demographic and selected clinical features of the three neuropsychology types. The three groups did not differ in terms of age, ratio of males to females, handedness, age of seizure onset, duration of epilepsy, or seizure type. However, children with mental retardation had a significantly higher incidence of precocious puberty and larger preoperative HHs than patients with normal intelligence.

Psychiatric and Behavioral Disorders as a Function of Neuropsychology Type

Table 2 lists verbatim the psychiatric terms and behavioral descriptions applied by previous examiners to each of the 57 patients studied. Forty-seven patients (82.5%) had one or more psychiatric and/or behavioral disturbances noted. Only 10 of 57 patients (17.5%) had no psychiatric or behavioral difficulties listed in their medical records. Of these 10 patients, 5 had estimated average intelligence. Multiple terms were applied to this patient group (Table 2), only some of which could be classified using the DSM-IV Manual.[1]

Table 3 summarizes the relative frequency of the diagnostic and descriptive terms that were recorded. Almost half of the sample was described as having mental retardation/developmental delay or borderline intellectual functioning (45.61%). Of 30 patients who were classified as having mental retardation based on current IQ testing, 21 had been classified by previous examiners as showing mental retardation or borderline intellectual functioning.

About one-third of the patients were described by previous examiners as having problems with anger control, episodic dyscontrol, or oppositional behavior (36.84%). These descriptive terms were applied across all levels of intellectual functioning (i.e., neuropsychology types).

The diagnostic terms that indicated attentional deficit/hyperactivity disorder and autistic spectrum disorder also applied across all levels of intellectual functioning. Approximately 16% of the sample were described as having attentional deficit/hyperactivity disorder and 14% as having autistic spectrum disor-

der. Diagnoses seemed to be independent of the patient's current level of intellectual test performance.

More than one-third of the patients were described in behavioral terms that did not easily lend themselves to psychiatric classification. Some of these terms included feeding disorders, self-injurious behavior, stuttering, and unspecified emotional and behavioral problems.

Discussion

This brief report documents that numerous psychiatric and behavioral difficulties have been applied to children and adults who have an HH and refractory epilepsy. Furthermore, more than 82.5% of the patients studied were described as having some form of psychiatric or behavioral disturbance.

As would be expected in children and adults who have an HH syndrome,[3] the diagnostic impression of mental re-

Table 1. Demographic and Clinical Characteristics of HH Patients by Neuropsychological Type

Variable	Type 1 (n = 15) mean (SD)	Type 2 (n = 12) mean (SD)	Type 3a (n = 21) mean (SD)	Type 3b (n = 9) mean (SD)	F/χ^2	*p*
Demographic Variables						
Age when tested (yrs)	17.60 (11.10)	14.33 (7.27)	17.64 (10.09)	12.13 (7.62)	0.51	0.61
Gender (% males)	46.7 (-)	58.3 (-)	68.2 (-)	87.5 (-)	5.77	0.22
Handedness (% right)	73.3 (-)	91.7 (-)	81.8 (-)	100 (-)	3.43	0.33
Seizure History						
Age at onset of any form of seizure	16.20 (26.42)	6.73 (8.88)	11.27 (14.59)	16.75 (28.28)	0.87	0.43
Epilepsy onset before 1 mo of age (n)	7	5	10	2	2.52	0.64
Epilepsy onset before 1 mo age (%)	46.7	45.5	45.5	25.0		
Duration of epilepsy (mos)	206.36 (128.11)	143.55 (62.61)	200.81 (130.56)	133.25 (91.85)	1.08	0.35
Seizure Type						
Gelastic only (n)	2	1	2	2	10.16	0.25
Gelastic only (%)	13.3	8.3	9.1	25.0		
Mixed (n)	12	10	20	6		
Mixed (%)	80.0	83.3	90.9	75.0		
Precocious puberty						
n	1	3	13	4	12.16	0.02
% present	6.7	27.3	59.1	50.0		
Pallister-Hall syndrome						
n	1	1	1	1	8.76	0.72
% present	6.7	8.3	4.5	12.5		
Hypothalamic hamartoma						
Presurgical size (cm)	1.29 (1.76)	1.31 (1.16)	5.72 (6.55)	4.41 (4.77)	5.69	0.01
Attachment						
Right						
n	6	5	6	3	6.11	0.64
% present	40.0	41.7	27.3	37.5		
Left						
n	7	3	7	2		
% present	46.7	25.0	31.8	25.0		
Bilateral						
n	1	1	1	1		
% present	13.3	33.3	40.9	37.5		

Table 2. Verbatim Descriptions of the Psychiatric and Behavioral Disorders of HH Patients with Refractory Epilepsy Found in Patients' Records as a Function of Neuropsychology Type

Neuropsychology Type 1	Neuropsychology Type 2†	Neuropsychology Type 3a†	Neuropsychology Type 3b†
Adjustment reaction with anxious mood	Anger problems	Adjustment disorder with depressed mood	Attention deficit hyperactivity disorder
Anorexia	Asperger's disorder	Attention deficit hyperactivity disorder	Autistic disorder
Asperger's disorder	Attention deficit hyperactivity disorder	Autistic spectrum disorder, probably Asperger's disorder	Behavioral problems
Attention deficit hyperactivity disorder	Attention deficit hyperactivity disorder NOS	Behavioral dyscontrol	Developmental delay
Autistic disorder	Behavioral dyscontrol	Borderline intellectual functioning	Episodic dyscontrol syndrome
Borderline traits	Borderline intellectual functioning	Cognitive disorder	Intermittent explosive disorder
Cognitive disorder NOS	Cognitive deficits	Cognitive disorder secondary to Asperger's	Learning disability
Cognitive dysfunction	Cognitive disorder due to medical condition	Cognitive dysfunction	Mental retardation
Depression NOS	Cognitive disorder NOS	Dementia secondary to seizure disorder	Obsessive compulsive disorder (features)
Developmental delay	Developmental delay	Developmental retardation	Oppositional behaviors
Dysthymic disorder	Emotional/behavioral disturbance	Developmental delay	Pervasive developmental disorder NOS
Major depressive disorder	Feeding problems	Emotional/behavioral disturbance	
Mood disorder	Impulse control disorder due to medical condition	Learning disability NOS	
Mood disorder secondary to HH	Learning disability	Major depressive disorder	
Oppositional defiant disorder	Learning disability NOS	Mental retardation	
Psychosis	Major depressive disorder	Obsessive compulsive disorder	
Self-injurious behavior	Mental retardation	Oppositional defiant disorder	
Temper outbursts	Mood instability with anxiety and depression	Pervasive developmental disorder	
Unspecified emotional and behavioral problems	Oppositional behavior	Psychosis	
	Paranoid disorder	Stuttering	
	Pervasive developmental disorder due to medical condition	Tourette's disorder	
	Poor impulse control		
	Sensory integration disorder		
	Schizoid personality traits		
	Specific language delays		

HH = hypothalamic harmatoma, NOS = not otherwise specified; †Five Type 1 patients, 2 Type 2 patients, and 3 Type 3a patients had no psychiatric diagnosis or behavioral disorder listed in their medical record.

Table 3. Summary of Psychiatric and Behavioral Disorders of HH Patients with Refractory Epilepsy as a Function of Neuropsychology Type

Psychiatric and Behavioral Disorder	Type 1 (n = 15)	Type 2 (n = 12)	Type 3a (n = 21)	Type 3b (n = 9)	Total (n = 57)	% of all diagnostic terms applied	% of patients having the term applied
Attention deficit/hyperactivity disorder	2	4	1	2	9	7.83	15.79
Oppositional defiant disorder and problems with behavioral dyscontrol/anger management					21	18.26	36.84
Oppositional defiant disorder	1	0	1	0	2	1.74	3.51
Temper outbursts	1	0	1	0	2	1.74	3.51
Anger problems	0	1	0	0	1	0.09	1.75
Impulse control problem	0	2	0	0	2	1.74	3.51
Behavioral dyscontrol/labile	1	1	3	3	8	6.96	14.04
Episodic dyscontrol	0	0	0	2	2	1.74	3.51
Oppositional behavior	0	1	0	1	2	1.74	3.51
Intermittent explosive disorder	0	0	1	1	2	1.74	3.51
Autistic spectrum disorder and other pervasive developmental disorders (PDD)					8	6.96	14.04
Asperger's disorder	1	1	1	0	3	2.61	5.26
Autistic disorder	1	0	0	1	2	1.74	3.51
PDD NOS	0	1	1	1	3	2.61	5.26
Psychosis, schizoid, or borderline traits	2	2	1	0	5	4.35	8.77
Mood disorder (major and minor)	5	3	3	0	11	9.57	19.30
Mental retardation/developmental delay/borderline intellectual functioning	1	4	14	7	26	22.61	45.61
Cognitive disorder/dysfunction	2	3	3	0	8	6.96	14.04
Learning disorder	0	3	1	2	6	5.22	10.53
Adjustment disorders	1	0	1	0	2	1.74	5.26
Other	4	4	7	4	19	16.52	33.33

HH = hypothalamic hamartoma, NOS = not otherwise specified

tardation was the most common term encountered. In the present study, however, fewer than 50% of the patients were described as having oppositional-defiant disorders or attentional deficit disorders. These findings contrast with those of Weissenberger et al.[10] who systematically studied 12 children with psychiatric comorbidities associated with HH and seizure disorder. In their small sample of patients, 83.3% were diagnosed as having an oppositional-defiant disorder, 75% were diagnosed as having attentional deficit/hyperactivity disorder, 33.3% were described as having a conduct disorder, and 16.7% were described as having an anxiety or mood disorder.

Our clinical impression is that a higher percentage of our patients showed oppositional-defiant disorders than was captured in the medical records. However, the percentage was less than 50% of our sample. Moreover, several children showed features indicative of an autistic spectrum disorder. Many behavioral terms were applied to these children, including problems in relating to others emotionally, having difficulties with social relationships, and early problems with feeding and with relating to early caregivers emotionally. We were impressed that most of the HH patients examined had subtle-to-obvious difficulties with social interaction, even if their intelligence was judged to be within the normal range.

Of the 10 HH patients who did not receive a psychiatric diagnosis or who were not described as having behavioral difficulties, 5 were estimated to have normal intelligence. This finding suggests

that there may be a relationship between normal intelligence and the absence of psychiatric/behavioral problems. Once a psychiatric and/or behavioral problem is present, estimates of IQ per se do not seem to relate to the diagnosis. However, other underlying neuropsychological deficits that do relate to the psychiatric diagnosis may be present. For example, neuropsychological impairments suggestive of a disorder of the frontal lobe system may be related to diagnoses such as attentional deficit/hyperactivity disorder or autistic spectrum disorder. This issue should be explored in future studies.

The present findings do not provide definitive estimates of the incidence of different types of psychiatric disorders observed in HH patients with refractory epilepsy. We also did not systematically assess the neurocognitive impairments underlying these disorders. However, our findings do suggest that numerous psychiatric problems may emerge in relationship to HH and untreatable seizure disorders. It is unlikely that a single variable (e. g., size of HH, mode of attachment, history of seizure disorder, cognitive strengths and weaknesses) can explain the varied psychiatric disturbances observed in this patient population. It is probable that the psychiatric and behavioral difficulties of HH patients represent complex interactions among their underlying brain disorder, seizure history, and their cognitive strengths and limitations (Prigatano GP, unpublished data, 2006). Furthermore, the role of medications and environmental factors contributing to the psychiatric difficulties of these patients cannot be excluded. Further studies should systematically assess these and other variables to help explain the various psychiatric disturbances seen in HH patients with refractory epilepsy.

Acknowledgment

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Diffuse Idiopathic Skeletal Hyperostosis versus Ankylosing Spondylitis: Brief Case Review

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Diffuse idiopathic skeletal hyperostosis is sometimes confused with ankylosing spondylitis. We present a case of diffuse idiopathic skeletal hyperostosis and describe its clinical management and the characteristics that may help differentiate this entity from ankylosing spondylitis. Clinicians must be aware of overlapping characteristics between these two disease processes and be able to distinguish between the two because the course of their treatment is considerably different. Key criteria for excluding ankylosing spondylitis and diagnosing diffuse idiopathic skeletal hyperostosis are the absence of sacroiliac fusion, erosion, or sclerosis, which can be determined by obtaining a plain x-ray of the pelvis.

Key Words: ankylosing spondylitis, cervical ankylosis, diffuse idiopathic skeletal hyperostosis

Abbreviations Used: CT, computed tomography; HLA, human leukocyte antigen; MR, magnetic resonance; OPLL, ossification of the posterior longitudinal ligament; OVAL, ossification of the vertebral arch ligaments

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Diffuse idiopathic skeletal hyperostosis is an idiopathic form of degenerative arthritis. It is usually characterized by exuberant bony growth along the anterior longitudinal ligament and typically affects males over 60 years of age. Diffuse idiopathic skeletal hyperostosis, previously known as Forestier's disease, is the most common enthesopathy. In 1950 it was originally described by Forestier and Rotes-Querol as a senile ankylosing hyperostosis of the spine.[2] This condition is associated with extraaxial involvement (i.e., ossification of the nuchal ligament or tendons in the extremities). Patients with this diagnosis can suffer from heel spurs or ligamentous calcification at the elbow or knee.

The other spinal enthesopathies are OPLL and OVAL. Diffuse idiopathic skeletal hyperostosis and ankylosing spondylitis are cervical ankyloses that can be difficult to distinguish from one another. Pathologically, diffuse idiopathic skeletal hyperostosis differs from ankylosing spondylitis. Clinically, however, both are similar. Diffuse idiopathic skeletal hyperostosis has even been referred to as a "senile ankylosing spondylitis."

Ankylosing spondylitis, a chronic inflammatory rheumatic disease, tends to affect relatively young white males. Classically, it is associated with sacroiliac and/or apophyseal fusion or sclerosis. Patients with ankylosing spondylitis are symptomatic and suffer from a myriad of associated conditions such as iritis, uveitis, or ulcerative colitis.

In contrast, patients with diffuse idiopathic skeletal hyperostosis tend to be asymptomatic. However, findings such as dysphagia from esophageal impingement, spinal cord compression, and pe-

ripheral nerve entrapment have been attributed to this form of cervical ankylosis. In practice, most patients are diagnosed after trauma and associated severe neurological deficits, such as quadraparesis or quadriplegia. Even trivial trauma, such as a minor fall or a syncopal episode, may be associated with severe neurological deficits in these individuals.[3]

Inevitably, the diagnosis of diffuse idiopathic skeletal hyperostosis is confused with a ankylosing spondylitis. We present the case of a patient with diffuse idiopathic skeletal hyperostosis and focus on the features that help differentiate it from ankylosing spondylitis.

Illustrative Case

A 69-year-old African-American man with a history of hypertension, diabetes mellitus, and peripheral vascular disease fell down a flight of stairs. He did not lose consciousness. On admission to our Emergency Department, he was significantly quadriparetic and had lost bowel control. A neurosurgical consultation was obtained.

On examination the patient was oriented to person, place, time, and situation. He opened his eyes spontaneously. His speech was fluent and his repetition was intact. He was able to follow commands. His Glasgow Coma Scale score was 15 (eye opening, 4; verbal response, 5; motor response, 6 following commands). His pupils were equally round and reactive to light. Grossly, his extraocular muscles and cranial nerve functions were intact. However, he had a sensory deficit at the level of the C4 dermatome, below which his sensation to light touch and pin prick was significantly impaired. On motor examination he was found to have a central cord syndrome. His arms were weaker than his legs.

Bilaterally, the strength of his deltoids was 1/5 to 2/5. Bilaterally, the strength of his biceps and triceps was 1/5, and the strength of his grip was 0/5. The cast on his lower right leg precluded a formal motor examination of this limb. However, strength in his left iliopsoas was 3/5 and 2/5 in his left quadriceps. Both his left plantar strength and dorsiflexion strength were 2/5. A digital rectal examination revealed mildly diminished rectal tone. The patient was started on the methylprednisolone spinal cord injury protocol.

His metabolic profile was as follows: sodium, 138 mEq/L; potassium, 4.5 mEq/L; chloride, 104 mEq/L; bicarbonate, 25 mEq/L; blood-urea-nitrogen level, 21 mg/dL; creatinine, 1.2 mg/dL; and glucose, 231 mg/dL. His white blood cell count was 11 x 10^3/mL. His hemoglobin was 11 gm/dL, and his hematocrit was 33%. His platelet level was 255 x 10^3/mL. His coagulation panel was normal.

CT of the head showed no intracranial hemorrhage, skull fracture, or extraaxial collections. The size and configuration of the ventricles were normal. As interpreted by the radiologist, CT of the cervical, thoracic, and lumbar spine showed "changes consistent with ankylosing spondylitis" but no acute fractures.

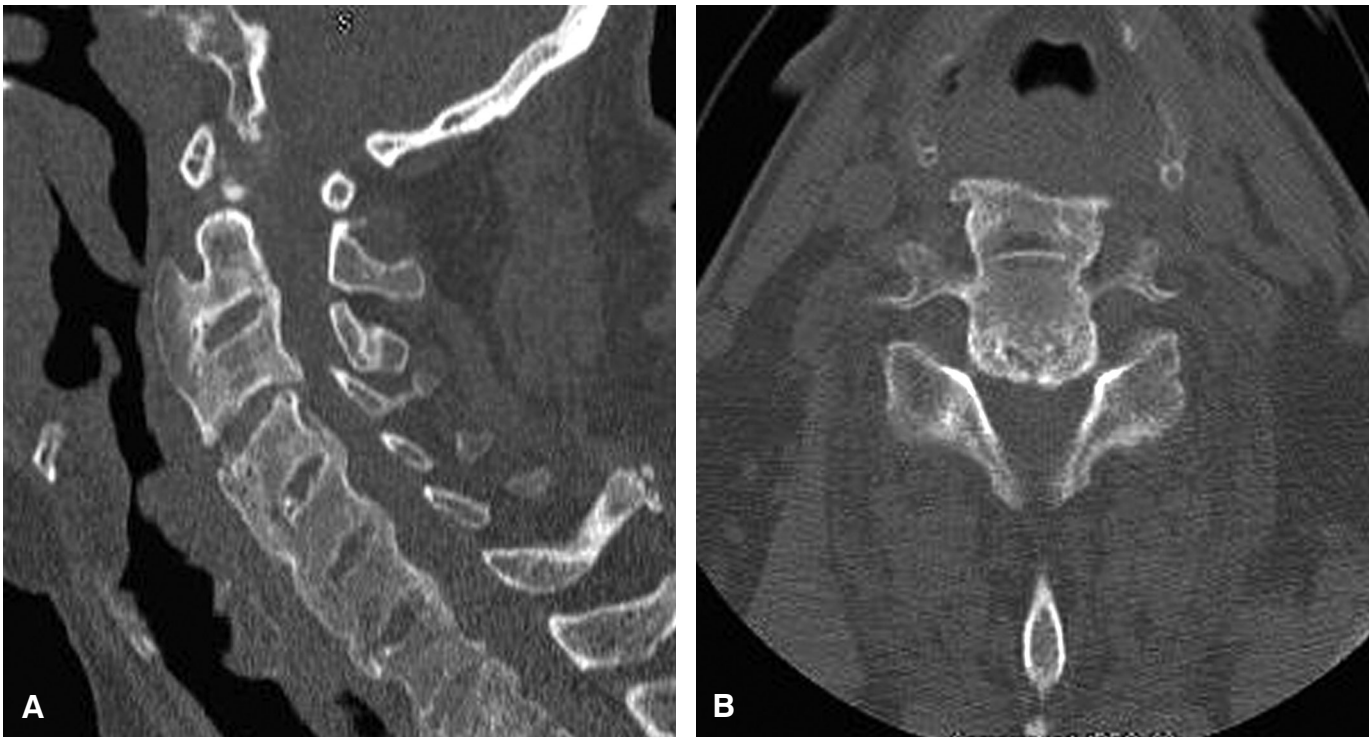

Figure 1. (*A*) Sagittal and (*B*) axial CT scans of the cervical spine show either diffuse skeletal hyperostosis or ankylosing spondylitis; both conditions have a similar appearance on CT studies. Ossification of the anterior and posterior longitudinal ligaments is associated with severe osseous ridging along the spine. Hyperostotic changes contribute to the severe stenosis of the spinal canal.

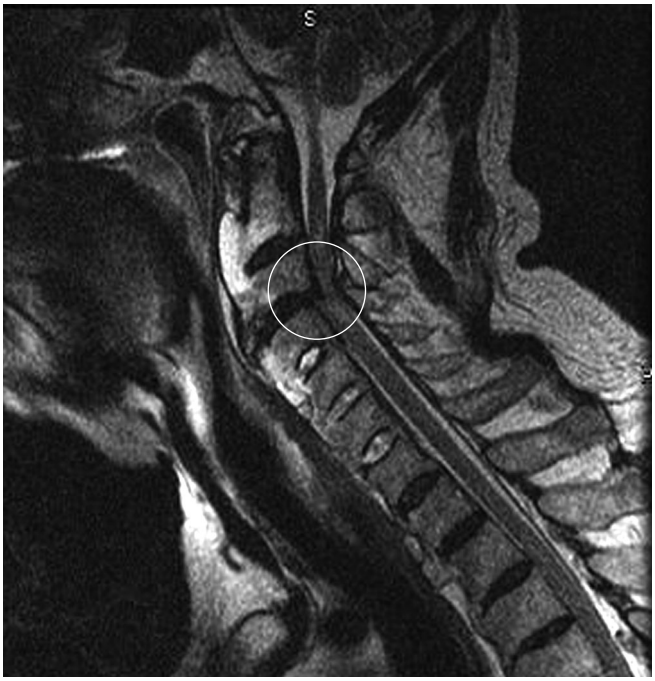

Figure 2. Sagittal T2-weighted MR image of the cervical spine shows a herniated nucleus pulposus at C3-C4 causing a change in the signal intensity of the spinal cord, stenosis of the central canal, and effacement of the ventral cerebrospinal fluid space (*circle*).

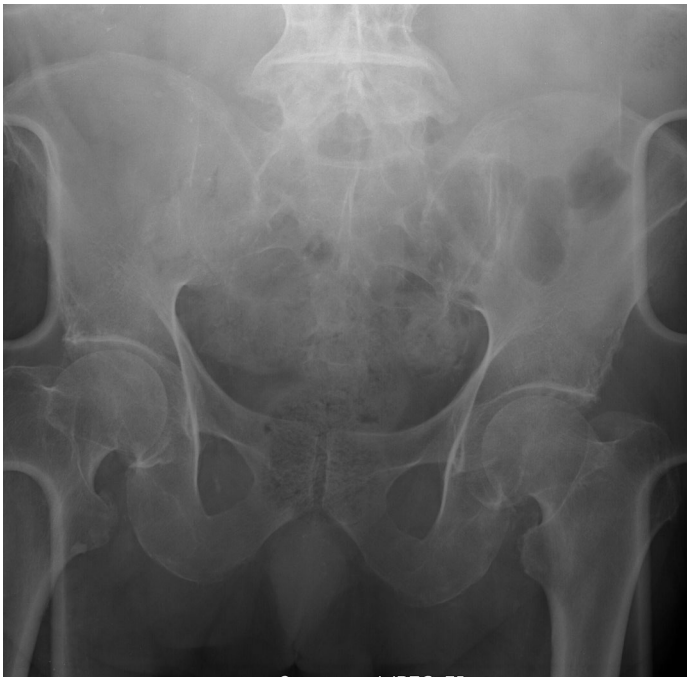

Figure 3. Anteroposterior x-ray of the pelvis shows no evidence of fusion, erosion, or sclerosis of the sacroiliac joint.

Specifically, CT of the cervical spine showed about 3 mm of retrolisthesis of C3 on C4 (Fig. 1). CT of the thoracic spine showed multilevel fusion and satisfactory alignment of the spine. There was evidence of mild-to-moderate degenerative changes associated with osseous ridging. At the midthoracic levels, the central canal was mildly to moderately narrowed. CT of the lumbar spine showed ossification anterior to the vertebral bodies and satisfactory alignment. Small osteophytes were present at L3-4 and L4-5, as was moderate narrowing of the central canal. Calcifications were also present in the descending aorta and proximal iliac arteries. A tiny calcific density involved the right kidney.

MR imaging of the cervical spine showed possible C3 and C4 fractures. Significant T2-weighted changes in the spinal cord at these levels were consistent with edema associated with severe stenosis of the central canal and a herniated nucleus pulposis at C3-4 (Fig. 2). No fusion of the sacroiliac was detected on pelvic radiographs (Fig. 3).

On admission to the intensive care unit for spinal cord trauma, the methylprednisolone protocol was continued. Subsequently, the patient was fixated in a halo brace in neutral cervical position. Based on the findings on his MR images, we performed an anterior cervical discectomy with allograft fusion to decompress the spinal canal at C3-C4 (Fig. 4).

Over the next few days after surgery, the patient developed respiratory distress. Auscultation throughout the lung fields indicated the presence of scattered rhonchi. After several days of aggressive pulmonary toilet and successful weaning from the ventilator, the patient was extubated. Initially, he tolerated the extubation, but the next day he desaturated and became further obtunded. After a respiratory code, he was fiberoptically reintubated emergently. Soon thereafter, an elective tracheostomy was placed.

Over the next few days, the patient's neurological examination fluctuated. Overall, his arms were weaker. However, strength throughout his left lower extremity was slightly improved to 3/5. Due to the worsening of his upper extremity and to the mild compromise of his cervical spinal canal associated with spinal cord compression on postoperative imaging, we decided to proceed with a decompressive laminectomy of C3 and C4 (Fig. 4). Postoperatively, the patient's strength was improved mildly as follows: bilateral biceps, 1/5 to 2/5; triceps, 3/5; and right and left grips, 1/5 to 2/5, respectively. Compared to his preoperative baseline, his lower extremity motor examination was unchanged, except for slight improvement of his left plantar flexion to 4/5 strength. At this point, the patient was transferred to neurorehabilitation.

Discussion

Diagnostic criteria for diffuse idiopathic skeletal hyperostosis include calcification or ossification of the anterolateral aspect of at least four contiguous vertebral bodies with or without osteophytosis; preservation of disk height without profound degenerative disk dis-

ease; and absence of ankylosing spondylitis, which includes fusion, erosion, or sclerosis of the sacroiliac.[5,10]

Pathologic features of diffuse idiopathic skeletal hyperostosis include focal and diffuse calcification and ossification of the anterior longitudinal ligament, paraspinal connective tissue, and annulus fibrosis. Other features include degeneration of the peripheral annulus fibrosis fibers; L-, T-, and Y-shaped anterolateral extensions of fibrous tissue; hypervascularity; chronic inflammatory cellular infiltration; and periosteal formation on the anterior surface of the vertebral bodies.[7]

Lower thoracic spine involvement is typical of diffuse idiopathic skeletal hyperostosis, but the rest of the spine can also be affected. Interestingly, the left side of the spine appears to be spared of changes relative to the right side. This observation has been attributed to pulsations from the nearby aorta.

One study described the vertebral involvement of 215 cadaveric spines and 100 patients with diffuse idiopathic skeletal hyperostosis. Radiographic features noted were linear osteogenesis along the anterolateral aspect of the thoracic spine, a bumpy contour, subjacent radiolucency, and irregular and pointed bony excrescences at the superior and inferior vertebral margins in the cervical and lumbar regions.[7]

As mentioned, most patients with diffuse idiopathic skeletal hyperostosis are asymptomatic unless they experience a precipitating trauma. However, some individuals with diffuse idiopathic skeletal hyperostosis complain of back stiffness, dysphagia (presumably because of the exuberant anterior spinal column and anterior longitudinal ligament ossification and osteophytosis impinging on the esophagus), and/or symptoms of myelopathy related to spinal canal compromise such as gait disturbance. Finally, chronic pneumonia has been reported due to bronchial obstruction and compression of the inferior vena cava.

Both diffuse idiopathic skeletal hyperostosis and ankylosing spondylitis are most common in Caucasians. In African-Americans ankylosing spondylitis is rare while diffuse idiopathic skeletal hyperostosis is common. Clinicians must be aware that the features of these two disease processes overlap and must be able to differentiate the two conditions because the course of their treatment is considerably different. For example, the treatment of ankylosing spondylitis may include the use of indomethacin and other nonsteroidal anti-inflammatory drugs, immunosuppressive treatment with monoclonal antibody therapy, and physical therapy.[1]

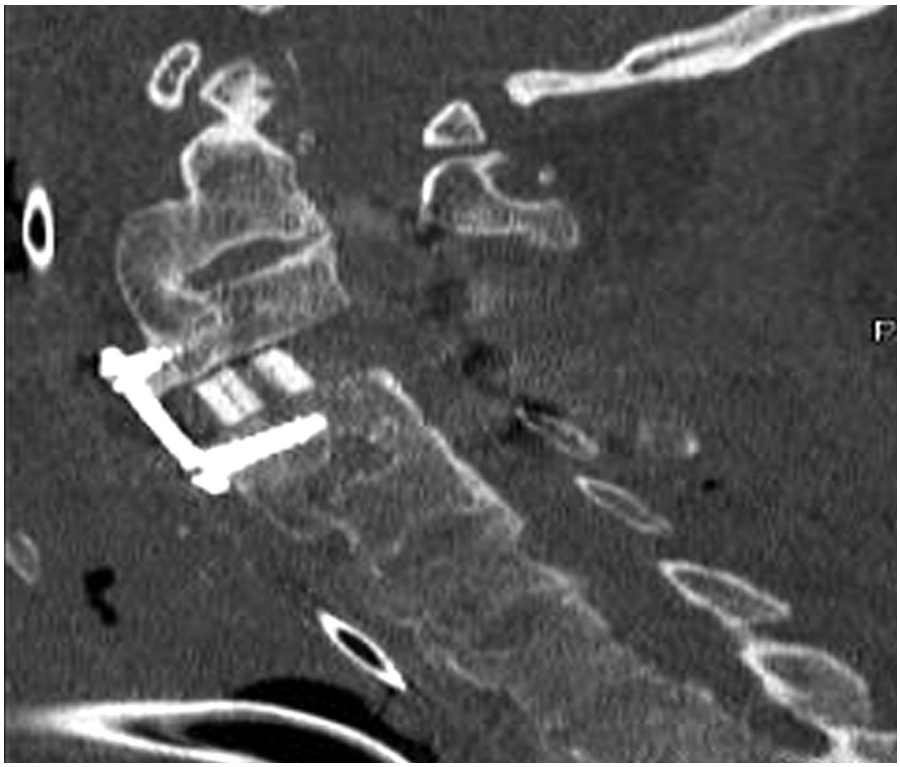

Figure 4. Postoperative sagittal CT scan of the cervical spine shows the decompressed central canal, anterior C3-C4 cervical discectomy with allograft and plate, and posterior decompressive laminectomy at C3 and C4.

The association between HLA-B27 and ankylosing spondylitis is strong.[4] Surprisingly, bacteria appear to play a role in the development of this disease.[8] In contrast, diffuse idiopathic skeletal hyperostosis usually occurs in middle-aged and older males and, as mentioned, is more common in African-Americans than ankylosing spondylitis. There is no ankylosis of the sacroiliac or apophyseal joints in patients with diffuse idiopathic skeletal hyperostosis (as evidenced by the normal pelvic radiograph in our patient). Finally, no strong genetic link has been identified in patients with diffuse idiopathic skeletal hyperostosis as has been found in patients with ankylosing spondylitis.[6]

The pathologic cause of diffuse idiopathic skeletal hyperostosis remains elusive. No genetic component appears to be present in most cases of chondrocalcinosis. However, a considerable number of cases seem to have a genetic predisposition for the condition, and an autosomal dominant pattern is evident. Linkage studies have shown the presence of a single locus in most cases of chondrocalcinosis. Based on the cases studied to establish a genetic background for disorders such as diffuse idiopathic skeletal hyperostosis or OPLL, chromosomes 5 and 8 appear to be most prominently involved, and two particular regions are involved on each chromosome. In one study, Tsukahara et al. showed a significant association between diffuse idiopathic skeletal hyperostosis in Japanese

patients and COL6A1 compared to Czech patients with the disease. Based on previous genomic linkage and linkage disequilibrium studies by the same authors and others, individuals with COL6A1 have been shown to be significantly susceptible to OPLL.[9]

Conclusion

Inevitably, the diagnosis of diffuse idiopathic skeletal hyperostosis is confused with ankylosing spondylitis. However, as described, identifying certain signs and symptoms leads to an accurate diagnosis. Diffuse idiopathic skeletal hyperostosis and ankylosing spondylitis are clinical entities with similar findings but different treatments. Accurate differentiation of these two conditions is vital to ensure that patients with severe cervical ankylosis receive the appropriate clinical management.

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De Novo Postoperative Simple Cerebellar Cyst

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Key Words: cerebellum, cyst, occipital sinus, pineal, supracerebellar infratentorial approach

Abbreviations Used: CSF, cerebrospinal fluid; CT, computed tomography; MR, magnetic resonance

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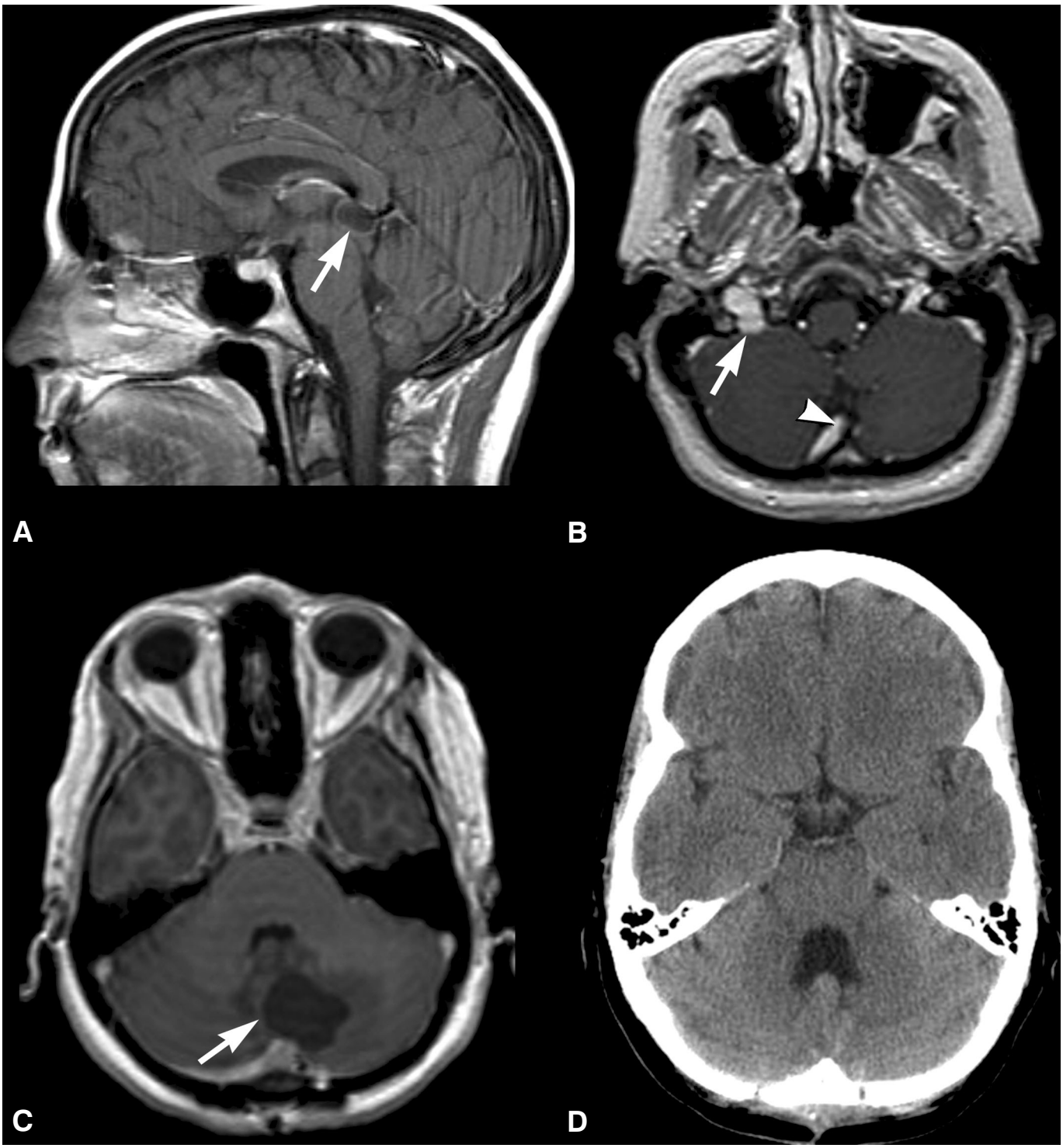
We could find no report in the literature similar to the following case, nor had any neurosurgeon in our institution previously encountered such a case.

A 45-year-old woman who sought treatment for symptoms of tectal compression and intermittent hydrocephalus underwent MR imaging. A sagittal contrast-enhanced T1-weighted MR image (*Panel A*) showed a pineal cyst (*arrow*) compressing the tectum. An axial contrast-enhanced T1-weighted MR image (*Panel B*) showed a dominant occipital sinus (*arrowhead*) draining into the jugular bulb (*arrow*).

During a supracerebellar infratentorial approach, bleeding from the dominant occipital sinus resulted in its sacrifice. A watertight dural closure was not performed. Postoperative MR imaging was unremarkable (*not shown*). The patient underwent placement of a lumboperitoneal shunt to manage a CSF leak. Six weeks after surgery her examination was normal. During the eighth week after surgery, she reported headaches, nausea, vomiting, dizziness, and unsteadiness. An axial contrast-enhanced T1-weighted MR image obtained at that time (*Panel C*) showed a nonenhancing, 2.8-cm cerebellar cyst (*arrow*) with no diffusion restriction associated with surrounding edema of the fourth ventricle. Laboratory examination was unremarkable. A presumptive atypical abscess was diagnosed.

Intraoperatively, we encountered a cyst with no evidence of infection. The cyst, which was under pressure, was fenestrated to the cisterna magna, and the lumboperitoneal shunt was externalized. Pathological examination revealed



degenerative white matter. Cultures remained negative. She never received antibiotics. Her externalized shunt was removed, and she was discharged home intact. Three months after surgery, she was normal. An axial nonenhanced CT scan (*Panel D*) confirmed resolution of the cerebellar cyst and fourth ventricular hydrocephalus ex vacuo.

We considered atypical abscess to be the most likely diagnosis. Ultimately, we are left with the diagnosis of a simple cerebellar cyst. We suspect that alteration in venous drainage related to ligation of the large occipital sinus, in concert with altered CSF flow from the lumboperitoneal shunting, may have played a role.