

Bulging Anterior Fontanel Associated with Apparent Benign Intracranial Hypertension

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Few clinical findings provoke more concern from the clinician than a bulging anterior fontanel in an infant or young child. A bulging fontanel signifies increased intracranial pressure and can be a sign of serious central nervous system disease.¹⁻³ Because of the potential for serious morbidity and mortality, the immediate concern is diagnosing and treating bacterial meningitis. If bacterial meningitis is ruled out, other pathologic causes may be considered, including encephalitis, intracranial abscess or hemorrhage, hydrocephalus, lead poisoning, sinus thrombosis, or tumor.¹⁻⁴ Alternatively, a bulging fontanel can result from a number of more common, nonpathologic disorders resulting in benign intracranial hypertension.¹

Case Reports

Case 1

A 5-month-old boy had a 12-hour history of fever, irritability, and bulging anterior fontanel. On physical examination he was an alert, interactive infant with a red throat, fluid behind the tympanic membranes, and a tense bulging anterior fontanel. A complete blood count revealed a white cell count of 8200/ μ L with a differential of 30 percent segmented neutrophils, 2 percent band cells, 65 percent lymphocytes, and 3 percent monocytes. A spinal tap specimen was grossly bloody and the cerebrospinal fluid was submitted for Gram stain and culture only. The Gram stain revealed no organisms. Blood cultures were obtained, and the infant received symptomatic treatment with in-hospital observation for 24 hours. During this time the fontanel became flat, and rhinitis and cough became prominent. The blood and cerebrospinal fluid cultures were ster-

ile, and the child recovered uneventfully. He has continued to do well and develop normally for the following 18 months.

Case 2

A 4-month-old girl had an 8- to 10-hour history of a fever of 104 °F, irritability, and bulging anterior fontanel. Findings on her physical examination were remarkable for a nonfebrile, irritable, but consolable, infant with a tense and pulsatile fontanel. A complete blood count revealed a white cell count of 25,700/ μ L with a differential of 60 percent segmented neutrophils, 9 percent band cells, 24 percent lymphocytes, 6 percent monocytes, and 1 percent basophils. Blood cultures were obtained. A spinal tap was performed, but no organisms were seen on Gram stain; protein was 60 mg/dL, glucose was 50 mg/dL, and the mononuclear cell count was 3/ μ L. The child was treated symptomatically at home. Blood and cerebrospinal fluid cultures were subsequently sterile. Her condition was reevaluated a few days later and showed resolving symptoms. During a 2.5-year follow-up period she has continued to do well.

Case 3

A 6-month-old boy had a 24-hour history of a bulging anterior fontanel and irritability. His parents reported no fever. During the physical examination the infant was alert and playful with no abnormalities except for a tense, bulging anterior fontanel. A complete blood count showed a white cell count of 8300/ μ L with 19 percent neutrophils, 66 percent lymphocytes, 2 percent basophils, 3 percent eosinophils, 9 percent monocytes, and 1 percent atypical lymphocytes. Because the infant was afebrile and did not appear ill, the possibility of bacterial meningitis was excluded, and a spinal tap was not performed. The cause of the bulging fontanel was still unknown following the initial examination. Subsequently a noncontrast computed tomographic scan of the head was performed and interpreted as normal. The child was observed without specific medical intervention.

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Within 2 days the fontanel was flat. A second episode of bulging anterior fontanel occurred 6 weeks later and was associated with a mild viral illness with rhinitis and cough. Again, the child was observed with no specific medical intervention. He is presently 2 years old and growing and developing normally.

Case 4

A 6-week-old boy was reported to have a fever of 101 °F and mottled skin, and feeding poorly. Examination showed an irritable but consolable infant with a mildly injected throat and mottled skin. The anterior fontanel was tense and bulging. A complete blood count showed a white cell count of 6100/ μ L with 14 percent neutrophils, 2 percent band cells, 83 percent lymphocytes, and 1 percent monocytes. The platelet count was 735,000/ μ L. A spinal tap produced bloody cerebrospinal fluid, which was submitted for Gram stain and culture only. The Gram stain showed no organisms. Because of the possibility of sepsis, the infant was hospitalized, given intravenous fluids, and prescribed ceftriaxone 100 mg/kg/d and gentamicin 7.5 mg/kg/d after blood and urine were collected for culture. In view of the bloody cerebrospinal fluid and the infant's appearing ill, an intracranial hemorrhage was suspected. A noncontrast computed tomographic scan of the head was performed and reported as normal. By the second hospital day the infant was afebrile. Blood, spinal fluid, and urine cultures were sterile at 72 hours. The infant was discharged home after 72 hours on no medication and continues to do well at 15 months of age.

Discussion

A bulging anterior fontanel is frequently observed in infants who have few or no specific clinical signs. The workup of an infant with a bulging fontanel must be individualized. In all instances the initial responsibility of the clinician is to consider and reasonably exclude bacterial meningitis. In cases 1, 2, and 4, where either the infant appeared ill on examination or fever was reported, a spinal tap was performed, and blood cultures were obtained to rule out definitively bacterial meningitis. In those cases where the spinal tap was grossly bloody, the cerebrospinal fluid was submitted for Gram stain and culture. In case 3, bacterial meningitis was excluded following the

history and physical examination. Brain imaging was done in cases 3 and 4, in which either the cause of the bulging fontanel was unclear or an intracranial hemorrhage was suspected. In-hospital observation was initially required for cases 1 and 4. All of the infants had or subsequently developed signs of a presumptive viral illness. Each infant showed quick resolution of the acute illness and bulging fontanel. In selected cases, after bacterial meningitis was ruled out, no specific medical intervention was offered, and no signs of neurologic or developmental sequelae have appeared in any of the infants on follow-up care.

Each of the cases most likely represented the syndrome of benign intracranial hypertension. Features of this syndrome include (1) clinical evidence of increased intracranial pressure, (2) no history of acute insult or injury to the nervous system, (3) normal cerebrospinal fluid studies (except for increased pressure), (4) normal findings on neurologic examination and absence of focal neurologic signs, and (5) a normal ventricular system on imaging studies if performed.^{1,5} Benign intracranial hypertension occurs in children of all ages. Its only manifestation in infants might be a tense or bulging fontanel. The diagnosis is one of exclusion. Specific therapy is seldom required in the absence of a treatable precipitating event or identifying cause. The prognosis for recovery is excellent.⁵

Benign intracranial hypertension has been associated with a number of disorders including those associated with endocrine or metabolic disease (galactosemia, hypophosphatasia), nutrition (hypovitaminosis A), drugs (sulfamethoxazole, penicillin, gentamicin, nalidixic acid, hypervitaminosis A), impaired cerebrospinal fluid absorption, and viral infections (most notably roseola infantum).¹⁻³ The most common cause in infants is thought to be catch-up brain growth.⁵ In the cases reported here, benign intracranial hypertension as evidenced by a bulging anterior fontanel was presumed to be related to self-limited viral infections. The viral infections typically are minor illnesses unassociated with meningitis or other serious central nervous system disease.¹⁻³ In some cases a bulging fontanel can apparently be the first or only sign of a viral illness.

In the current climate of health care cost containment and medicolegal concerns, how extensive a clinical workup is warranted when a child

has a bulging anterior fontanel but lacks specific clinical findings? Elevations in intracranial pressure are easier to detect clinically in infants because of the accessibility of the anterior fontanel.⁶ Nevertheless, the anterior fontanel is a nonspecific clinical indicator of increased intracranial pressure. Differentiating between benign and pathologic elevations in intracranial pressure requires further clinical evaluation. In infants with patent fontanels, unfortunately, clinicians do not routinely trust their clinical assessment skills and judgment; rather, they are more likely to initiate extensive and expensive diagnostic procedures and therapeutic interventions for increased intracranial pressure as evidenced only by a tense or bulging anterior fontanel. This aggressive approach might not be warranted in many cases.

Any child with a tense or bulging anterior fontanel demands immediate attention because of the possible risks associated with increased intracranial pressure and bacterial meningitis, regardless of whether the presumed cause is benign. The history and physical examination should direct and focus the subsequent evaluation and management. If the infant is febrile, has changes in the state of alertness, or appears ill, or if there is a reasonable possibility of bacterial meningitis, a spinal tap is required for diagnostic purposes even in the presence of increased intracranial pressure.

Brain imaging (sonography, computed tomography, magnetic resonance imaging) is not routinely performed in cases of presumed or proven meningitis unless complications, such as subdural effusions or abscesses, are suspected. Brain imaging is indicated in infants with a bulging fontanel if (1) the clinical diagnosis is unclear, (2) neurologic deterioration or seizures occur, (3) focal or lateralizing neurologic deficits are present, (4) an intracranial hemorrhage or abscess is suspected, or (5) the patient is slow to recover, implying a possibly overlooked disorder.

If the history is unremarkable, the infant has nonspecific findings on physical examination, and a complete blood count is unremarkable or suggests a viral illness (normal white cell count with

lymphocytosis), expectant therapy with close clinical observation and follow-up would be the appropriate and preferred management. Brain scans, spinal taps, and other procedures should not be performed if they will provide little useful information or if it is extremely doubtful that the care of the patient will be altered.⁷ Repeated episodes of a bulging anterior fontanel in an infant regardless of clinical findings warrant a thorough workup to determine the cause.

A conservative management approach to a bulging anterior fontanel emphasizes close serial monitoring and assumes a low tolerance for any persistence or progression of illness or for the development of clinical signs suggesting a pathologic increase in intracranial pressure. If family compliance with recommended care might be an issue, the clinician is obligated to initiate a well-planned, intense evaluation with appropriate management.

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