Dandy-Walker Malformation: A Case Study of an Infant With an Increasing Head Circumference and Delayed Developmental Milestones

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KEY WORDS
Dandy-Walker malformation, developmental delays, increasing head circumference

CASE PRESENTATION

A 9-month-old African American girl presented for a pediatric well-child visit after having missed the 2-month and 6-month visits. The child's developmental progress, therefore, had not been followed from a primary care standpoint.

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Conflicts of interest: None to report.

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Chief Complaint

The mother's chief complaint was that the infant's head seemed to be very large and that the infant may have some muscle weakness, as demonstrated by her inability to sit without losing her balance. The mother also expressed concerns regarding other aspects of the infant's development including motor, social, and language skills when she filled out the Parents' Evaluation of Developmental Status form.

History of Present Illness

The infant was delivered via a spontaneous vaginal birth after cesarean at 39 weeks' gestation. Her birth weight, length, and head circumference were 2.438 kg (2.9th percentile), 45.72 cm (3.6th percentile), and 32.5 cm (11.5th percentile), respectively. At discharge, her weight was 2.296 kg (1.1 percentile). Her hearing screening and newborn screening results were normal. The mother reported that the child has been in good health with varied diet, no known allergies, and no medications.

Family and Social History

The infant was born to parents who are of African American descent and enrolled in Medicaid. The infant lives with her mother and two older siblings, both of whom had normal development according to the mother. The infant's mother has legal custody and is the primary caregiver. The infant is enrolled in daycare while the mother works as a hostess at a restaurant.

Past Medical History

Head circumference was not recorded at the 1-month visit, so baseline percentile was not known. The mother did not have any developmental concerns at that time. The infant missed her 2-month visit. At the 4-month visit the head circumference was 41.91 cm (75th percentile), and the mother did not have any developmental concerns. Because of interruption in the child's expected health maintenance schedule, this child was not up to date on immunizations and therefore required a catch-up schedule as recommended by the Centers for Disease Control and Prevention (2016). At the 4-month visit, the infant received her 2-month immunizations, including the diphtheria, tetanus, and pertussis vaccine, the Haemophilus influenzae type b vaccine, the pneumococcal conjugate vaccine, and the inactivated polio vaccine. Although the plan was to continue the immunization catch-up schedule, the mother failed to follow up, so the child missed the 6-month health maintenance visit.

With regard to motor skills, the mother noted that the infant sits without support but loses her balance easily. The mother reported that the infant does not creep, crawl, or pull to stand, stand holding on, or cruise on her own. According to the mother, the infant is unable to feed herself finger foods or reach for objects with a pincer grasp. Social and language skills are reported to include the ability to use her voice to express joy and pleasure and to respond to her name. The mother reported that, thus far, the infant has not demonstrated the ability to imitate speech, play social games, or understand a few words. The mother also reported that the infant does not show stranger or separation anxiety, show understanding of object permanence, or point out objects. The mother stated that the infant appears to hear well and was mostly worried about the infant's lack of crawling, scooting, or pulling up to stand.

Review of Systems

The patient's mother reported that the child has been well and that she has no concerns regarding eyes, ears, nose, or throat. The mother denied any intercurrent illnesses or history of respiratory, cardiovascular, or gastrointestinal concerns. The mother reported that the infant slept 7 to 8 hours at night without waking and took one 2-hour nap during the day. She reported that the infant had approximately eight to nine wet diapers per day with one bowel movement per day. The mother reported that the child is delayed compared with her siblings' development in sitting and inability to crawl or pull up. The mother noticed that the child's head looked larger than normal during the month before the 9-month visit and that the infant may have some muscle weakness. The infant's mother reported that the infant can roll herself from supine to prone, but gets stuck with rolling prone to supine.

Physical Examination

At this visit, the infant weighed 6.87 kg (2.35th percentile), with a length of 71 cm (63.53th percentile) and head circumference of 47 cm (98.68th percentile). Her vital signs were normal. The anterior fontanelle was flat and open without bulging or separated sutures. The infant was alert, content, and interactive with her mother. When placed in the prone position she was able to prop briefly on her elbows with her arms abducted and lift her head briefly. While in the supine position she brought her hands bilaterally to midline and to her feet. She maintained her head in the midline with pull to sit, but demonstrated head lag over the first 45°. The infant sat without support, but lost balance easily. The infant's reflexes were integrated appropriately for her age. All other results of the physical examination were normal.

Diagnostic Studies and Referrals

Because of suspicion of global developmental delay, an outpatient physical therapy evaluation was completed. The clinical summary stated that the infant demonstrated low muscle tone throughout her trunk and extremities with significantly delayed motor skills. She scored significantly less than the 5th percentile on the Alberta Infant Motor Scale, which is a performance-based, norm-referenced test that is used to measure motor maturation of infants from term to the age of independent walking. She was observed to sit without support, but her balance was not reliable. She demonstrated prone skills at the 3- to 4-month level, with poor tolerance for this position. She required assistance to maintain standing. She rolled from supine to prone independently, but did not consistently roll from prone to supine. She did not otherwise transition between positions.

Because of the gross motor delays described by the patient's mother and seen on examination, a hip x-ray examination was ordered to rule out or confirm developmental dysplasia of the hip. The hip x-ray imaging result was negative. To further explore the increased head circumference, head ultrasonography (US) was ordered. The US showed a cystic structure at the dorsal aspect of the third ventricle, ventriculomegaly, and prominent retrocerebellar cerebral spinal fluid (CSF) collection in the posterior fossa. The US was followed by magnetic resonance imaging (MRI) of the brain with and without contrast including flow studies. A referral was also made to a neurosurgeon at the local children's hospital.