Evaluation of the Child with Microcephaly

Case Presentation

An 18-month-old boy is brought to the neurology clinic by his parents. He is sent to you for a neurological consultation by his primary care doctor for microcephaly. The parents are concerned that the boy’s head seems small. They also note that he is not walking as well as his older brother did at this age. This boy was born at term. His APGAR scores were 7 and 8. His developmental milestones were notable for sitting at 11 months, crawling at 13 months, and walking at 18 months. He eats well and interacts well with other children his age. He is babbling and is able to say “mama” and “dada.” He has no significant past medical history. There were no obvious interuterine infections or chemical or toxic exposures, and his mother did not consume alcohol, smoke, or use drugs. He has had no surgeries. He has no allergies and is on no medications. Family history is noncontributory. Review of systems reveals no difficulties with sleep or digestion. There are no difficulties with cardiac, respiratory, or urinary function. There are no musculoskeletal, endocrine, or hematologic problems reported. There has been no clear seizure activity.

On physical exam the child is alert and responsive. Vital signs are BP 98/50, HR 88, and RR 18, and his temperature is 99. Head circumference is slightly below the 2nd percentile. Weight is in the 50th percentile. Height is in the 50th percentile. The child knows his name and correctly identifies his mother. He is able to follow simple commands to approach and raise arms. Cranial nerve examination to include a detailed funduscopic examination are normal. He responds well to visual stimulation in all quadrants. Pupils react normally, and extraocular movements are normal. Facial sensation and motor strength are normal bilaterally. Hearing is intact to finger rub. Palate, tongue, and uvula are midline. Motor examination is 5/5 throughout, with normal tone and no drift. Reflexes are 2/4 symmetrically bilaterally. Sensory is intact to light touch, pinprick, and vibration bilaterally. Coordination with finger to finger is intact. The child is able to walk, with a slightly wide-based gait. He loses balance when he turns. He is not able to walk on toes or heels. He is not able to balance on one foot. His mother’s head circumference is in the 50th percentile. His father’s head circumference is in the 80th percentile.

You discuss with the parents that the boy’s head circumference is low. You explain to the parents that there are a variety of reasons for microcephaly. You provide them with information on microcephaly. You discuss with them that, because he is having some difficulties with his walking, it may be useful to order blood work and imaging on him. You order a head CT, and review the complete blood count, chemistry panel, liver function tests, and thyroid levels that were performed one month prior by the primary care physician. You schedule a follow-up evaluation in 2 months. Greater than 50% of the 80 minute visit is spent counseling the parents on microcephaly, discussing the
possible causes, the differential diagnosis of microcephaly, natural history of the illness, management, and genetics.

Questions

1. When evaluating a patient with microcephaly, the following may be considered an early useful step:
   A. Neuroimaging with head CT or MRI to assess for structural causes
   B. PET scanning to evaluate for neurotransmitter abnormalities
   C. EEG to evaluate for epilepsy, which is always associated with microcephaly
   D. Cerebrospinal testing to evaluate for neurodegenerative changes
   E. Chest/abdominal and pelvic CT to evaluate for structural problems

   The correct answer is A. Neuroimaging may be considered useful in identifying structural causes in the evaluation of the child with microcephaly (Level C).  

2. Annually, approximately ______ infants in the United States will be diagnosed with microcephaly.
   A. 10,000
   B. 25,000
   C. 50,000
   D. 100,000
   E. 150,000

   The correct answer is B. Approximately 25,000 children are diagnosed with microcephaly every year. Microcephaly is an important neurological sign, but there is nonuniformity in its definition and evaluation. Microcephaly may result from any insult that disturbs early brain growth and can be seen in association with hundreds of genetic syndromes.

3. Children with severe microcephaly are more likely to have imaging abnormalities and more severe developmental impairments than those with milder microcephaly. Coexistent conditions include:
   A. movement disorders, epilepsy, and headaches
   B. headaches, gait difficulties, and epilepsy
   C. epilepsy, cerebral palsy, mental retardation, and ophthalmologic disorders
   D. ophthalmologic disorders, headaches, and back pain
   E. epilepsy, movement disorders, and headaches

   The correct answer is C.

   The guideline recommendations are:
   Neuroimaging may be considered useful in identifying structural causes in the evaluation of the child with microcephaly (Level C). Targeted and specific genetic testing may be considered in the evaluation of the child with microcephaly who has clinical or imaging abnormalities that suggest a specific diagnosis or who shows no evidence of an acquired
or environmental etiology (Level C). Screening for coexistent conditions such as cerebral palsy, epilepsy, and sensory deficits may also be considered (Level C). Further study is needed regarding the yield of diagnostic testing in children with microcephaly.

**ICD-9-CM© Discussion**

Microcephaly is stated as the diagnosis and is associated with a gait disturbance that is complex by description. The appropriate codes in this case are:

742.1 Microcephalus
781.2 Abnormality of gait

If a cause of the microcephaly is found, that diagnosis would become the first listed code.

**Evaluation and Management Coding Discussion**

Coding for the Evaluation and Management of this patient is relatively easy, as one can use the counseling and coordination-of-care method for coding. Since the patient is sent in consultation for an outpatient visit and more than 40 minutes of an 80-minute visit is spend in counseling and coordinating care, one would use code 99425 or a level 5 consultation code (80 minutes total spent with the patient and more than 50% expended in counseling or coordinating care).

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