A Toddler with Prolonged Fever

Robert Listernick, MD

This two-and-a-half year old girl was evaluated for 15 days of fever. She was well until 2 weeks prior to the visit when she developed temperature of 101°F and vomiting at night. Although the vomiting subsided over the next day or two, she continued to have fever. At times, she would say that her “butt hurts”, although this was not a particularly consistent complaint. She continued to walk normally. In addition, there was no history of further vomiting, diarrhea, rhinitis, cough, rash, arthritis, red lips, red eyes, or other symptoms.

Her past history was remarkable for a similar episode 3 months earlier that started with nighttime vomiting and proceeded to similar fever that lasted 3 weeks. There were no other past medical problems. Her development was normal. She was the 7-lb product of a 36-week gestation to a 31-year-old G2P2. The pregnancy was complicated by maternal migraines. She was born by cesarean section due to breech presentation. There were no perinatal problems.

There were no exposures to pets or unusual food such as unpasteurized milk or cheese or undercooked meat. The only travel history was to Ohio several months earlier. However, the grandparents are missionaries who returned from Papua, New Guinea shortly before the onset of the illness. The father is of Dutch descent and the mother does not know her genetic background.

On exam, she was somewhat fearful, but otherwise healthy-appearing. Her weight was in the 25th percentile, height in the 90th percentile, and head circumference in the 90th percentile. BP was 92/50. There were no rashes. She was normocephalic. HEENT exam was normal. There was no significant adenopathy. Lungs were clear. Cardiac exam was normal. Abdominal exam was normal without hepatosplenomegaly. She had Tanner 1 genitalia. Her neurologic examination was unremarkable in detail. Her gait was normal. At the base of her spine, there was a 9 mm round flat vascular lesion within which there was a dimple with a central protuberance.

Representative laboratory data: CBC with normal hemoglobin, white blood cell count 14,000/mm3 with 59% neutrophils, 36% lymphs, platelet count 300,000/mm3; ESR 8 mm/hour, CRP 0.8 mg/dL. Hepatic function panel, urinalysis, and chest x-ray were all normal. Urinalysis and urine culture were normal.

Robert Listernick, MD, moderator:
Comments?

Anthony Mancini, MD, pediatric dermatologist: The lesion on her back is a capillary malformation (port wine stain, salmon patch) with a little papule in the middle. Its location high up from the superior margin of the anal verge makes it a low-risk lesion for an intraspinal component. However, the risk of intraspinal extension increases when there truly is an appendageal remnant or skin tag within the lesion.

Dr. Listernick: Are there data that support your ability to predict which cutaneous lesions have intraspinal components?

Dr. Mancini: There are few prospective studies of lumbosacral lesions. However, there are several statements that I feel comfortable making. Our recent hemangioma research group data suggest that as many as 25% of lumbosacral hemangiomas have associated dysraphism or spinal cord abnormalities. Thick coarse hair, skin tags or appendages, deeper palpable lipoma-like nodules, or larger congenital melanocytic nevi overlying the lumbosacral spine are all lesions that should undergo neuroimaging. In general, without true supporting prospective data, pediatric dermatologists feel that the presence of two or more findings in this area (eg, capillary malformation plus dimple or sinus) warrants neuroimaging.

Dr. Listernick: What about isolated sacral dimples?

Dr. Mancini: There was a prospective descriptive study that looked at this question in a normal newborn nursery. All babies with any sacral dimples un-
derwent neuroimaging. Pits that were larger than 5 mm in diameter or further than 2.5 cm from the anus were more likely to be associated with intraspinal extension, occult spinal dysraphism, or cord defects.

**Dr. Listernick:** If the physician decides that imaging is warranted, what diagnostic study should be performed?

**Delilah Burrowes, MD, pediatric neuroradiologist:** Ultrasonography is a reasonable screening test in infants less than 3 months of age. If negative (no masses or tethered cord), no further testing is generally necessary. Abnormal scans need to be followed up by magnetic resonance imaging (MRI).

**Dr. Mancini:** I just want to point out that in our study I mentioned, several babies had normal ultrasounds only to have subsequent MRI scans which demonstrated intraspinal abnormality (tethered spinal cord, lipoma, hemangioma). The reliability of ultrasound in this setting is very dependent on the experience of the radiologist.

**Dr. Listernick:** For whatever reason, she never had neuroimaging or any evaluation of this lesion. So what do you make of her history of repeated fever with perfectly normal inflammatory markers?

**Stanford T. Shulman, MD, pediatric infectious disease physician:** It’s hard to say because recurrent minor febrile illnesses are common. However, listening carefully, it sounds as though the duration of these episodes is too long to be attributable to a simple viral infection. Although one might consider a periodic fever syndrome, she has none of the common symptoms such as rash, oral ulcers, arthritis or abdominal pain. In addition, the inflammatory markers should be quite elevated during a “flare”.

**Dr. Listernick:** Does she have a fever of unknown origin (FUO)?

**Dr. Shulman:** That’s a difficult call. Classically, there are two defining features of FUO. First, the fever has to have lasted for a prolonged period of time. There’s no generally accepted duration in pediatrics; many specialists suggest a minimum febrile period of 7 to 10 days. Second, the physician has to be actively looking for the source of the fever. This child is a bit unusual, as she’s had recurrent periods of low-grade fever and it’s difficult to know what to make of the vomiting.

**Dr. Listernick:** I gave you the answers to most of the specific questions you would explore when evaluating a child with FUO.

**Dr. Shulman:** You did. We want to explore the possibility of unusual exposures – animals, travel, visitors who have traveled, undercooked meat or unpasteurized milk, and possible exposures to tuberculosis.

**Dr. Listernick:** What would each of you have done now?

**Dr. Shulman:** I would have performed blood and urine cultures and considered imaging her abdomen and, perhaps, her lower spine.

**Dr. Listernick:** That’s a good start. I gave you the answers to most of the specific questions you would explore when evaluating a child with FUO.

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Stanford T. Shulman, MD
Editor-in-Chief, PEDIATRIC ANNALS
placed. She was sent home to perform a fever diary and await the results of the cultures. However, 2 nights later she awoke screaming in pain, although the parents could not identify its source. She was admitted to the hospital and the plan was to obtain a computerized tomography scan of the abdomen. However, she required general anesthesia to perform the scan and we were forced to wait 36 hours. While waiting for the scan to be performed, it was noted that she had recurrent fever to 104°F. In addition, she was twice noted to have a fleeting macular salmon-colored rash on her back, thighs and buttocks. The rash appeared at the height of the fever and disappeared when she defervesced. So, have we just made the diagnosis of juvenile idiopathic arthritis (JIA)?

Marisa Klein-Gitelman, MD, pediatric rheumatologist: Hardly. Most importantly, she doesn’t have the “A,” arthritis. You cannot make a diagnosis of JIA without arthritis. She needs to have a swollen joint or loss of the normal range of motion with pain in order to make that diagnosis.

Dr. Listernick: That makes sense. Do you have a strong reason to suspect it in this child?

Dr. Klein-Gitelman: Not really. It’s true that you may not see arthritis for months after the onset of inflammatory symptoms in JIA. However, she does not have leukocytosis or thrombocytosis nor does she have morning stiffness. I think you’ll have to broaden the differential diagnosis.

Dr. Listernick: Under the guise of “I’d rather be lucky than right,” she had a computerized tomography scan of the abdomen looking for an intraabdominal abscess or tumor.

Dr. Burrowes: Imaging of the abdomen and pelvis was unremarkable. However, it was noted that there was abnormal signal with questionable enhancement within the central canal of the spinal cord. In addition, there appeared to be a cerebrospinal fluid track (CSF) track leading from the cord to the surface of the back. Following this, MRI of the spine was performed. On the T2 images, you can appreciate that there’s extensive enhancement along the ventral and dorsal aspects of the distal cord and cauda equina. The nerve roots appear clumped together; we don’t see distinct nerves of the cauda equina floating in CSF.

Dr. Listernick: And your interpretation of these findings?

Dr. Burrowes: There’s a dermal sinus tract leading to arachnoiditis and meningitis.

Dr. Nordli: I was quite serious about the need for a rectal exam earlier. Abnormal anal tone or the presence of sacral anesthesia might have led to the diagnosis sooner.

Dr. Shulman: I presume this is a dermoid cyst. Although these lesions may present with frank meningitis with either enteric organisms or skin flora, they may also cause recurrent episodes of smoldering chemical meningitis due to rupture of the dermoid contents into the cord.

Aiblash Haridas, MD, pediatric neurosurgeon: She was taken to the operating room. We found a very large lumbar cyst filled with a cheesy material quite characteristic of a dermoid. There was evidence of severe arachnoiditis; the nerve roots of the cauda equina, rather than being free-floating, were all clumped together. We gingerly tried to separate these nerve roots and remove the cyst itself. This was accomplished with the aid of electrophysiologic monitoring of the anal region. Everything went well and we’re hopeful for a completely normal neurologic outcome. Even if she had neurologic residua, we would expect improvement over the ensuing 6 months.

Dr. Listernick: Was the CSF infected?

Julie Stamos, MD, pediatric infectious disease physician: The culture grew Klebsiella oxytoca. Although a cell count inadvertently was not obtained, the fluid looked clear. In the end, we decided to treat her with 3 weeks of intravenous ceftriaxone, although I was never truly convinced that she had bacterial meningitis.

Dr. Haridas: I agree. My sense is that she more likely had recurrent chemical meningitis from intermittent rupture of the dermoid cyst.

Dr. Listernick: Thank you, everybody.

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**Key Learning Points**

1. Thick coarse hair, skin tags or appendages, palpable lipomatous nodules, prominent sacral dimples, large or complicated vascular lesions, or congenital melanocytic nevi overlying the lumbosacral spine should suggest consideration for neuroimaging in order to evaluate for spinal dysraphism, intraspinal extension, or spinal cord defects.

2. In experienced hands, ultrasonography is a reasonable screening test for intraspinal abnormalities in infants less than 3 months of age. If negative (no masses or tethered cord), no further testing is generally necessary. Abnormal scans need to be followed up by magnetic resonance imaging (MRI).

3. Abnormal anal tone or the presence of sacral anesthesia are important clues to the presence of a lumbosacral spinal cord abnormality such as a tethered cord or tumor.

4. Dermal sinus tracts with intraspinal extension may present with frank bacterial meningitis. Alternatively, they may lead to recurrent episodes of smoldering chemical meningitis due to rupture of the dermoid contents into the cord.