A Unique Presentation of Chronic Primary Sternal Osteomyelitis With Mediastinal Abscess

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CASE REPORT

A previously healthy 9-month-old boy presented to the emergency department with 4 days of daily fevers as high as 39.1°C rectally. Parents reported that he was increasingly difficult to console for the previous several days. Although not initially reported, after the diagnosis was made, the parents reported occasional tactile fever and difficulty sleeping, particularly when lying prone, over the preceding few weeks.

On examination, the child was febrile (39.2°C rectally) and tachycardic (pulse 140 beats/min). He was alert but crying inconsolably, seemingly in significant discomfort. Complete examinations by multiple physicians failed to demonstrate any apparent source of his pain and fever. Notably, bony tenderness was not appreciated, and he had no erythema or swelling over his sternum.

Initial laboratory data demonstrated a white blood cell (WBC) count of 15 900/uL with 53% neutrophils and 5% bands. Erythrocyte sedimentation rate was 20 mm/h and C-reactive protein (CRP) was 22 mg/dL.

On admission, cerebrospinal fluid examination, urinalysis, and computed tomography (CT) imaging of the head and abdomen were unremarkable. Chest x-ray demonstrated a retrocardiac left lower lobe infiltrate. Of note, the patient slept comfortably during the transthoracic echocardiogram. Chest CT demonstrated a large anterior mediastinal abscess (Figure 1). The abscess tracked around the sternum with marked bony erosion and fragmentation compatible with sternal osteomyelitis. The anterior mediastinal abscess extended from the level of the left brachiocephalic vein to the cardiac apex, and osseous fragments were lying within the abscess (Figure 2). Retrospective review of the chest x-ray revealed destruction of the sternal ossification centers that had not been recognized initially.

The cardiovascular surgeons incised and drained the mediastinal abscess, debrided his sternum, and placed a wound vacuum-assisted closure device. Contents of the abscess were under pressure, and 15 mL of purulent fluid with multiple free-floating bony fragments were drained. The wound was left open post-operatively. He had no recurrence of fever after surgery. Culture of the fluid also grew MSSA, and the antibiogram was identical to that of the MSSA isolate from the blood. Vancomycin was changed to intravenous cefazolin and oral rifampin, and in vitro synergy studies confirmed an additive effect. Over the next 3 weeks, he underwent 5 wound irrigation procedures. His CRP normalized within 2 weeks of his initial surgery. Final chest closure was performed 3 weeks...
after the original surgery, and he was discharged in good condition thereafter to receive intravenous cefazolin and oral rifampin.

Upon follow-up with the infectious diseases service 3 weeks after hospital discharge, he was doing well with a well-healed wound and normal examination. Erythrocyte sedimentation rate was 11 mm/h, and CRP was less than 0.5 mg/dL. Because of the severity and unusual location of the infection, an immune evaluation was performed. Neutrophil oxidative burst for chronic granulomatous disease was assessed by flow cytometry following dihydrorhodamine incubation of the patient’s WBCs. This test, along with quantitative immunoglobulins, vaccine antibody titers, and lymphocyte subset quantitation and functional assays, was normal. After receiving 6 weeks of intravenous cefazolin and oral rifampin, he was prescribed oral cephalexin to complete a 6-month antibiotic course.

DISCUSSION

Acute hematogenous osteomyelitis is the most common form of pediatric osteomyelitis, most frequently due to \textit{S. aureus} [1, 2]. Because of its high affinity for bone and soft tissues, \textit{S. aureus} bacteremia should prompt a careful investigation of these sites as a possible source of infection. Bacteremia with this organism without an obvious source should not be dismissed as a contaminant. \textit{Staphylococcus aureus} typically localizes to highly vascularized metaphyses, particularly of the long bones. \textit{Staphylococcus aureus} expresses a number of proteins on its surface, such as microbial surface components recognizing adhesive matrix molecules (MSCRAMMs), that facilitate adhesion to several components of the bone and soft tissue extracellular matrix. The organism then expresses a number of exotoxins and hydrolases that attack host cells and degrade the colonized tissue, respectively [2]. Primary sternal osteomyelitis is a very rare presentation of AHO in childhood, accounting for 0.2\% of cases of pediatric AHO in a large series [5].

Primary sternal osteomyelitis has been previously reported in 20 children [3]. Among the 15 children in whom a pathogen was recovered, \textit{S. aureus} was found in 7, with 2 of 7 being methicillin resistant. The next most common pathogens were \textit{Salmonella} species, isolated in 5 children with sickle cell disease. All but 3 of the 20 cases presented acutely within a week of symptoms, most often within 5 days of symptom onset. All 20 reported cases of PSO demonstrated obvious objective signs of sternal inflammation on physical examination, including swelling, erythema, and tenderness overlying the sternum. Our patient was unique in that examination by multiple physicians failed to demonstrate any objective signs of sternal inflammation, even after the diagnosis was confirmed radiologically. Furthermore, during his initial evaluation, he was observed to be sleeping comfortably during transthoracic echocardiogram. His extreme discomfort was most likely caused by pressure generated by the expanding mediastinal

Figure 1. Image from chest computed tomography with contrast demonstrating a large ring-enhancing anterior mediastinal fluid collection measuring 1.9 x 4.4 x 5.2 cm consistent with abscess.

Figure 2. Image from chest computed tomography with contrast demonstrating a ring-enhancing fluid collection surrounding the fragmented second sternal ossification center with bony fragments lying within the abscess.
abscess as it pushed anteriorly on his chest wall. This is supported by the large size of the abscess, its compression of the right ventricle, the contents of the abscess being clearly under pressure, and prompt resolution of his inconsolability postoperatively.

Management of AHO generally consists of targeted antibiotic therapy with surgical intervention as needed. Among the 19 cases of acute PSO reported during the antibiotic era since 1973, 5 required surgical drainage [3]. Surgical intervention both offers a potential therapeutic benefit, particularly when osteomyelitis is associated with abscess formation, and may improve diagnostic sensitivity by increasing the likelihood of identifying the pathogen. In a series of patients with acute PSO secondary to community-associated, methicillin-resistant S. aureus, culture from the surgical drainage was positive in all cases, whereas blood culture was positive in only 20% of the cases [3]. Bacteremia is even more unusual in chronic osteomyelitis, leading us to postulate that this patient was bacteremic secondary to the complicating mediastinal abscess.

Optimal duration of antibiotic therapy for PSO is unknown, although published reports suggest duration of 3–6 weeks for acute PSO [3–5], similar to that recommended for AHO of other anatomic sites [1, 2]. Optimal duration of therapy for chronic osteomyelitis is less clear. We generally treat chronic osteomyelitis with 6 weeks of intravenous therapy followed by oral therapy to complete a 6-month course [6]. Our patient was determined to have chronic PSO based upon the history of low-grade fevers and fussiness for several weeks preceding the onset of high fevers and inconsolability. This was supported by the radiologic finding of bony destruction of the sternum.

This child was evaluated for a possible immunodeficiency, and the work-up was negative. Although this child had an extensive immunologic evaluation, this severe invasive staphylococcal infection raised the highest suspicion for a defect in phagocytic function, especially chronic granulomatous disease (CGD) [7]. Invasive staphylococcal infections, as well as recurrent skin and soft tissue infections, are not uncommon in otherwise healthy children. Thus, the indications for pursuing evaluation of an immunodeficiency in these patients are not entirely clear, and the consideration of an immunodeficiency will vary somewhat among practitioners. However, there are certain scenarios in which CGD should be strongly considered. Chronic granulomatous disease, which is most commonly of X-linked inheritance, should be especially considered in young male patients with a particularly severe staphylococcal infection, particularly in the liver or lung. Recurrent invasive infections with S. aureus and/or other catalase-positive bacteria or fungi, such as Burkholderia cepacia, Nocardia species, Serratia marcescens, and Aspergillus species, should also prompt strong consideration for CGD.

This unique case challenges the notion that PSO always presents acutely with objective cutaneous signs of sternal inflammation. When PSO fails to elicit obvious external signs of infection, medical evaluation may be delayed, leading to development of chronic infection and contiguous spread into the mediastinum. Failing to recognize this clinical entity in its acute phase may significantly complicate the clinical course and increase the likelihood of requiring surgical intervention. Therefore, PSO should be considered in the broad differential diagnosis of a child with irritability and fever without a source.

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References
