An 11-Year-Old Boy With Ear Pain and Facial Palsy

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Case Report

An 11-year-old boy presented to primary care with intermittent self-limited peripheral left facial nerve palsy and left ear pain with no evidence of discharge. He was treated with a 7-day course of empiric oral antibiotics for a speculative diagnosis of left otitis media with partial improvement of his otalgia. Two months prior, he had reported a brief period of fever, chills, sweating, diffuse musculoskeletal pain, and fatigue. Otherwise, he had been previously well.

One month after his antibiotic therapy, the patient developed a subacute debilitating arthralgia, starting in his left hip joint and progressing to involve the left shoulder and knee. His joint pain increased with activity, woke him from sleep, and resulted in a 2-week absence from school. The pain showed limited response to nonsteroidal anti-inflammatory drugs and acetaminophen. Concomitantly, he had low-grade fever, night sweats, and progressive left ear pain and discharge as well as persistence of the left facial nerve palsy. There was no history of appetite change or weight loss. He was retreated empirically with 14 days of oral high-dose amoxicillin. In addition, a 7-day course of oral corticosteroids (prednisone 80 mg/day) was added as an adjunct therapy for presumed Bell's palsy.

Due to persistence of his symptoms, the patient was referred to our hospital for further assessment. On presentation, he appeared well and had normal vital signs. Otoscopic examination revealed left external auditory canal swelling and debris, and the tympanic membrane was difficult to visualize. The soft tissue over the left mastoid bone was swollen and tender, and he demonstrated ipsilateral infra-nuclear facial nerve palsy. Musculoskeletal exam was significant for limited internal rotation of the left hip and point tenderness of the left sacroiliac joint. The remainder of his physical exam was unremarkable.

Given the persistent left otomastoiditis, left facial nerve palsy, and musculoskeletal findings, the differential diagnosis was widened to include atypical bacterial infections (tuberculosis, Lyme disease), systemic rheumatologic conditions (chronic recurrent multifocal osteomyelitis, human leucocyte antigen [HLA] B27 related arthropathies), and malignancies such as leukemia.

Final Diagnosis

Acute lymphoblastic leukemia

Hospital Course

Initial blood work on presentation to our hospital, including infectious serology, was remarkable only for an elevated C-reactive protein, erythrocyte sedimentation rate, and lactate dehydrogenase (Table 1). Petrous bone computed tomography scan without contrast showed opacification of the left mastoid air cells and middle ear cavity, with no definite evidence of bone destruction. X-ray of the right hip showed sclerosis of the acetabular roof. Whole body magnetic resonance imaging revealed multiple areas of abnormal ill-defined foci of signal within the bone marrow consistent with a disseminated process (Figure 1).

Auditory canal and left mastoid bone biopsies were performed in conjunction with a bone marrow examination. Both biopsies showed a dense infiltrate of lymphoblasts and necrotic tissue. Neoplastic cells had positive nuclear staining for terminal deoxynucleotidyl transferase, and strong cytoplasmic and membrane staining for

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Table 1. Comprehensive Hematological and Infectious Workup.

Investigation	Result	Normal Range
Hematology and chemistry		
White blood cell count	12.6 × 10^9/L (high)	4-10 × 10^9/L
Hemoglobin	137 g/L	120-160 g/L
Platelet count	299 × 10^9/L	150-400 × 10^9/L
Neutrophil count	5.23 × 10^9/L (high)	2-7.5 × 10^9/L
Bands	1.55 × 10^9/L (high)	0.0-0.01 × 10^9/L
Monocytes	1.4 × 10^9/L (high)	0.05-0.8 × 10^9/L
Atypical lymphocytes	1.97	
Metamyelocytes	0.14	
Myelocytes	0.28	
Blood film	No blasts	
Erythrocyte sedimentation rate	72 mm/h (high)	1-10 mm/h
C-reactive protein	36.7 mg/L (high)	0-0.8 mg/L
C-3 complement	2.32 g/L (high)	0.77-1.43 g/L
C-4 complement	0.43 g/L (high)	0.07-0.4 g/L
IgG	8.7 g/L	7.0-15.5 g/L
IgA	1.9 g/L	0.5-3.6 g/L
IgM	1.6 g/L	0.4-2.9 g/L
Creatine phosphokinase	40 U/L	60-330 U/L
Lactate dehydrogenase	1087 U/L (high)	432-700 U/L
Microbiology and serology		
Cytomegalovirus IgG by EIA	Negative	
Cytomegalovirus IgM by EIA	Negative	
Epstein-Barr virus EA IgG by EIA	Negative	
Epstein-Barr virus EBNA IgG by EIA	Negative	
Epstein-Barr virus VCA IgG by EIA	Negative	
Herpes simplex virus IgG by EIA	Negative	
Varicella zoster virus IgG by EIA	Negative	
Lyme disease antibodies	Negative	
Aerobic blood culture	No growth	

Abbreviations: Ig, immunoglobulin; EIA, enzyme immunoassay; EBNA, Epstein-Barr nuclear antigen; VCA, viral capsid antigen.

colony differentiation factor 10, 3, and 20. Bone marrow aspirate and biopsy showed 40% blasts, confirming the diagnosis of pre-B acute lymphoblastic leukemia (ALL). Bacterial, mycobacterial, and fungal cultures were all negative.

The child was transferred to the oncology service for further management, where standard of care treatment for ALL was initiated. He responded well to treatment and continues to be in remission.

Discussion

Facial palsy in the context of otomastoiditis most commonly results from infection or inflammatory compression of the facial nerve as it traverses the petrous portion of the temporal bone.[1-3] When secondary to a hematologic malignancy, facial nerve involvement is often bilateral, accompanied by central nervous system (CNS) leukemic infiltrate.[4] When unilateral, facial nerve palsy in the setting of malignancy may be secondary to middle ear or mastoid bone infection, or tumor infiltrate of the mastoid with surrounding tissue edema. Rhabdomyosarcoma of the temporal bone has been reported to locally infiltrate the facial nerve territory and cause peripheral paralysis.[5]

Unilateral otomastoiditis and facial nerve palsy has been previously reported as a case of CNS relapse, without hematologic leukemic manifestations, in a 14 year-old boy previously diagnosed with ALL.[6] Furthermore, a retrospective study of children presenting with facial nerve palsy concluded that 90% were treated with corticosteroids, while 2% were ultimately diagnosed with acute myeloid leukemia.[7]

Our patient's cerebrospinal fluid analysis was negative for evidence of both malignancy and infection, supporting the theory that his unilateral facial paralysis was secondary to direct compression of the infiltrated left mastoid bone. Less likely is that our patient had

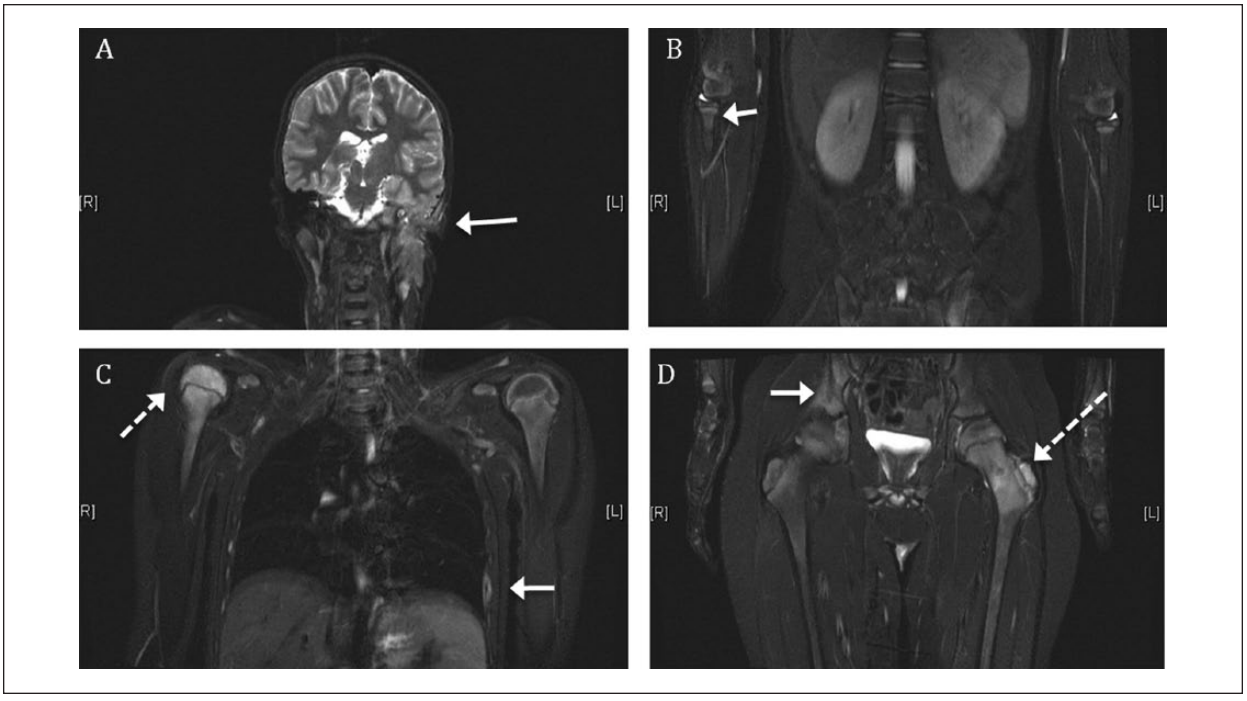

Figure 1. Magnetic resonance imaging of whole body showing foci of asymmetrical high signal within the region of (A) left mastoid (solid white arrowhead), (B) high signal is also noted within right radial head (solid white arrowhead), (C) right humeral head (dashed white arrowhead), left eighth and ninth ribs (solid white arrowhead), (D) right acetabulum with associated mild abnormality in surrounding soft tissue (solid arrowhead) and left femoral trochanter (dashed arrowhead).

evolving CNS leukemia, masked due to administration of corticosteroid therapy.

This case highlights important lessons for the clinician concerning the empiric use of corticosteroids. While the majority of patients with Bell's palsy are treated with corticosteroid therapy to reduce the potential risk of residual nerve paralysis, it is imperative to remember that Bell's palsy is idiopathic by definition, and that the absence of pain or findings on otoscopic evaluation is critical. In our case, the insidious onset of the palsy and associated systemic symptoms were red flags suggesting an alternate diagnosis. Furthermore, systematic review evidence has questioned the effectiveness of corticosteroids for Bell's palsy in the pediatric population.[8] In a well-designed retrospective study of patients who were diagnosed with leukemia, initial treatment with corticosteroids resulted in diagnostic delay, in addition to an increased likelihood of chemotherapy resistance and worse ultimate prognosis.[9]

Finally, the clinician will do well to recall the significance of musculoskeletal complaints in the pediatric population. In a multicenter retrospective study of children presenting to a pediatric rheumatology service with unexplained musculoskeletal complaints, Jones et al determined the 3 strongest predictive factors for ALL to be a history of nighttime awakening with pain, a low white blood cell count (less than 4×10^9/L), and a low-normal platelet count (150-250 $\times 10^9$/L). Together, these factors had a sensitivity of 100% and specificity of 85% for the diagnosis of ALL among patients with prolonged bone-joint disease.[10]

Conclusion

Otomastoiditis and concomitant facial palsy is an uncommon initial presentation of children with leukemia, a common childhood malignancy. Facial palsy should only be considered an idiopathic Bell's palsy if associated signs and symptoms such as pain or otologic findings are ruled out. Finally, musculoskeletal complaints in the pediatric population should always prompt consideration of malignancy.

Author Contributions

ARS: Contributed to conception and design; contributed to analysis; drafted the manuscript; critically revised the manuscript; gave final approval; agrees to be accountable for all aspects of work ensuring integrity and accuracy.

WB: Contributed to analysis; critically revised the manuscript; gave final approval; agrees to be accountable for all aspects of work ensuring integrity and accuracy.

BCP: Contributed to analysis; critically revised the manuscript; gave final approval; agrees to be accountable for all aspects of work ensuring integrity and accuracy.
CEB: Contributed to conception and design; contributed to analysis; critically revised the manuscript; gave final approval; agrees to be accountable for all aspects of work ensuring integrity and accuracy.

Declaration of Conflicting Interests

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