Skull and Soft Tissue Syphilis: 15-year-old with Scalp and Skin Lesions

THADDEUS WRIGHT, MD; OSAMA IBRAHIM, MD; ELENA PUSCASIU, MD; GLENN A. TUNG, MD, FACR; SABINA D. HOLLAND, MD

CASE REPORT

A 15-years-old female without significant past medical history presented to her primary care provider after noticing a firm lump on her forehead that had been growing for one month. She was referred to pediatric neurosurgery and oncology for evaluation of a calvarial mass. Physical examination (PE) revealed a firm, nontender 5-centimeter mass without overlying skin changes and scattered 0.5–1 centimeter violaceous, brown papules on the scalp, trunk, and extremities. Laboratory evaluation was unrevealing (complete metabolic panel, complete blood count with differential, uric acid, c-reactive protein, and coagulation studies). Magnetic resonance imaging (MRI) with contrast (**Figure 1**) demonstrated an ill-defined, enhancing expansile frontal calvarial lesion with cortical dehiscence and extension through the outer table into the subgaleal scalp. Given the unusual presentation, the differential diagnosis was broad including eosinophilic granuloma, a localized form of Langerhans cell histiocytosis (LCH), xanthoma disseminatum, cutaneous lymphoma, blastic plasmacytoid dendritic cell neoplasm, pott's puffy tumor, and eruptive dermatofibroma.

The patient was referred to pediatric infectious disease after biopsy revealed spirochetes and perivascular lymphoplasmacytic infiltrates; immunochemistry was consistent with spirochetes (**Figure 2**). Additional history revealed the patient was born in the United States without history of travel. She had two lifetime male sexual partners. There was no prior history of sexual transmitted infections (STI). PE revealed exophytic nontender lesions on the labia and a lesion on the hard palate consistent with condyloma lata, a manifestation of secondary syphilis. The remainder of her PE, including neurological and musculoskeletal exams, was normal. Additional studies included Treponema IgG/IgM antibodies >8.0 [negative ≤0.8], rapid plasma regain (RPR) reactive, titer 1:256. Cerebrospinal fluid (CSF) analysis was without abnormality except reactive fluorescent treponemal antibody absorption (FTA-ABS); CSF venereal disease research laboratory (VDRL) testing nonreactive. STI testing including HIV was negative.

DISCUSSION

Clinical presentations of syphilis are widely variable such that it is often referred to as "The Great Imitator."[1] Since reaching a historic low in the year 2000, the incidence of syphilis has risen dramatically, notably by 28.6% between 2020 and 2021.[2] Primary syphilis characteristically presents with a solitary chancre at the initial site of inoculation.[3] Secondary syphilis manifests with more systemic symptoms: fever, rash (including palms/soles) and lymphadenopathy. Condyloma lata and alopecia occur in <10% of patients. Widespread spirochete dissemination contributes

Figure 1. Sagittal and axial MRI: [**A**] Sag T1 demonstrating expansile frontal calvarial lesion with cortical dehiscence and extension through the outer table into the subgaleal space. [**B**] Sag T1 post-contrast with enhancement and [**C**] Axial T2 fat suppressed showing osteitis.

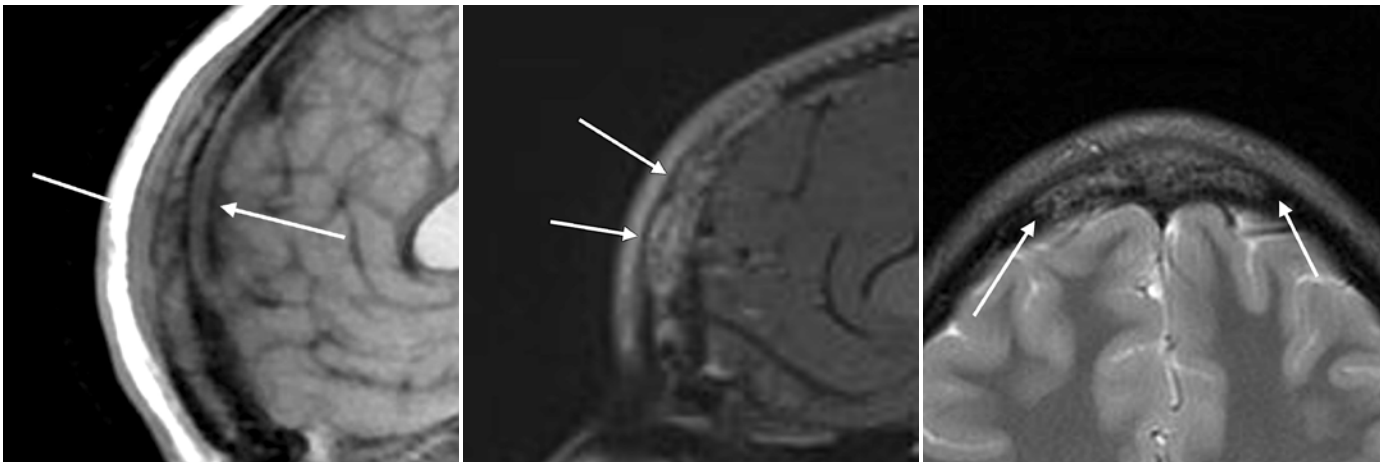


Figure 2. Steiner stain revealing scattered spirochetes.

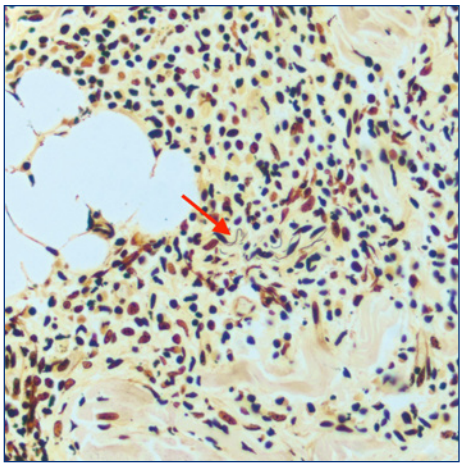
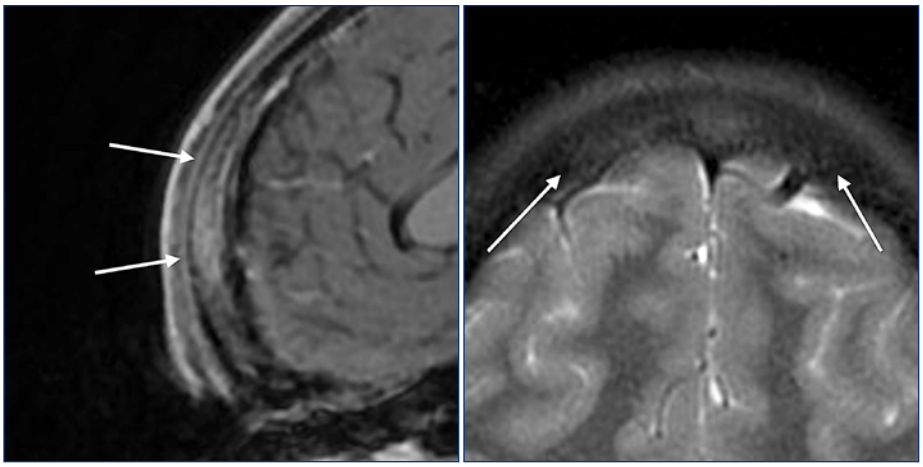


Figure 3. [**A**] Sagittal T1 post-contrast and [**B**] axial T2 fat suppressed MRI demonstrating decreased enhancement and osteitis six months later and post-treatment with antibiotics.



to the myriad of symptoms capable of involving most organ systems seen in secondary/tertiary syphilis.

Osseous lesions in syphilis occur in 0.15–8.7% of patients and present from six weeks after the initial chancre or up to 14 months after skin rash.[4-6] When syphilis presents as osseous lesions, long bones and skull are the most often affected.[6-8] Bone involvement in pediatric syphilis is well-established in congenital and tertiary syphilis but syphilitic osteitis in the early stages of pediatric-acquired syphilis is extremely rare.[9] There are two case reports: a 12-year-old with secondary syphilis with tibial periostitis, and a 13-year-old with upper- and lower-extremity periostitis.[10]

The differential diagnosis of adolescent skull lesions includes LCH, sarcoid, Gorham disease, intraosseous meningioma, intraosseous venous vascular malformation, and metastasis.[4,11,12] Given the increasing incidence, and wide spectrum of clinical presentations, clinicians must be aware of syphilis; left untreated, the long-term sequalae of syphilis are devastating.[3,13]

Treatment regimens are well-established;[14] there are no guidelines for treatment of syphilitic osteitis. Satisfactory results were reported using intramuscular benzathine penicillin G (BPG) for two to four weeks, intravenous aqueous penicillin G (APG) for two to six weeks, 10 days of intramuscular penicillin G procaine, or ceftriaxone.[15,16] Given the paucity of literature, she was treated for neurosyphilis with intravenous APG followed by intramuscular BPG for three weeks with reduction in RPR to 1:8, improvement in bone lesions (**Figure 3**), and resolution of rash and alopecia.

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Authors

Thaddeus Wright, MD, Department of Diagnostic Imaging, The Warren Alpert Medical School of Brown University, Providence, RI.

Osama Ibrahim, MD, Department of Pediatric Infectious Disease, The Warren Alpert Medical School of Brown University, Providence, RI.

Elena Puscasiu, MD, Assistant Professor of Pathology and Laboratory Medicine, The Warren Alpert Medical School of Brown University, Providence, RI.

Glenn A. Tung, MD, FACR, Professor of Diagnostic Imaging, The Warren Alpert Medical School of Brown University; Rhode Island Medical Imaging, Providence, RI.

Sabina Holland, MD, Associate Fellowship Program Director, Department of Pediatric Infectious Disease, The Warren Alpert Medical School of Brown University, Providence, RI.

Correspondence

Thaddeus Wright, MD
Department of Diagnostic Imaging
Rhode Island Hospital
593 Eddy Street, Main 327-A
Providence, RI 02903
thaddeus_wright@brown.edu