Leprosy is caused by the acid-fast bacillus Mycobacterium leprae, affecting mostly the skin, peripheral nerves, and mucus membranes. Clinical forms of Hansen disease represent a spectrum of cellular mediated immune responses to Mycobacterium leprae. Leprosy reactions are an immunological phenomena that occur before, during, or after completion of multi-drug therapy (MDT) for leprosy. This case describes a type 2 or erythema nodosum leprosum (ENL) reaction.

**LEARNING OBJECTIVES**

1. Recognize a presentation of ENL in leprosy
2. Know appropriate treatment to prevent damaging effects of ENL

**INITIAL PRESENTATION**

**Presentation**

Pt is a 26yo Filipino male with known h/o Hansen’s disease that was diagnosed on skin biopsy in 2007 presents with 4 days of progressive arthralgias in knees, ankles, wrists and elbows, accompanied by diffuse painful nodular rash, subjective fevers, night sweats, right testicular pain and swelling, right eye pain and blurry vision. Pt has chronic numbness in his left arm and bilateral feet, but notes worsening anesthesia in 4th and 5th digits of both hands. As for the patient’s Hansen’s disease, pt is followed and treated by a leprosy clinic and had been recently tapered off his clofazimine, dapsone, and prednisone three weeks earlier after being asymptomatic for several months.

**Exam**

Vital signs: T of 102.6 F HR 130s BP 120s/80s

Skin: several raised, hot, tender, red, tender nodules on extensors of bilateral upper, lower extremities, and palms of hands

Eyes: R eye: conjunctival injection, cloudy, white opacity in anterior chamber, L eye: normal, EOMI

GU: Right tender, warm, red scrotal swelling

Neuro: bilateral UE and LE sensory deficits with decreased sensation in ulnar nerve distribution of left hand, 1st-3rd digits of b/l feet, weak left hand grip

**Data**

WBC of 27; Blood Cultures NGTD; Scrotal US: R Epididymal orchitis

Punch Skin Biopsy of Left Arm 4/07: Fite/AFB stain showed+ AFB stain, some within nerve fibers, confirming diagnosis of leprosy. Features consistent with lepromatous rather than tuberculoid spectrum. Presence of neutrophils and e/o vasculitis suggest erythema nodosum leprosum. +“foamy” histiocytes with perineural involvement.

**CLINICAL COURSE**

At this time, given dramatic inflammatory presentation, erythema nodosum leprosum was diagnosed clinically, and patient was given solomedrol in the ED. Patient’s symptoms responded immediately with overnight resolution of his rash and new motor neuropathy. He was started on Prednisone 60mg poqam, Clofazimine, and Dapsone. Ophthalmology was consulted for his uveitis and was started on prednisolone eye drops without improvement in his vision, requiring subconjunctival decadron injection. Thereafter, patient’s vision and eye pain improved.

**DISCUSSION**

Erythema nodosum leprosum can be diagnosed clinically in patients with lepromatous leprosy (LL) and is usually associated with multi-drug therapy. It can occur in as much as 50% of pts with LL. Usually occurs within first year of therapy, but can occur after therapy is discontinued. It is thought to be an immune complex disorder associated with elevated levels of TNF alpha.

This case illustrates the importance of diagnosing an immune reaction in the setting of leprosy. Given pt’s sepsis presentation, first-line therapy was corticosteroids, not antibiotics. An urgent ophthalmology consult was also warranted to prevent vision loss in setting of uveitis with blurry vision. Recognition of this syndrome is critical to instituting appropriate therapy and preventing progression of motor and sensory loss, skin changes, and vision loss.

**HISTORICAL PEARLS**

- B.C.: Leprosy was feared, misunderstood, thought to be a punishment from God.
- 17th century: Leprosy was a significant medical problem in Norway. Dr. Gerard Hansen of Norway discovered Mycobacterium leprae, which became the first meaningful cure for leprosy, an alternative name, Hansen’s disease, has been advocated by healthcare groups.
- In early 1900s, USPHS funded a leper colony in Carville, La, from which came the first meaningful cure for leprosy, dapsone.

**REFERENCES**