Case 21
A 34-year-old Thai man from Samut Prakan
Chief complaint: Progressive swelling of the rash on the face of 1-week duration

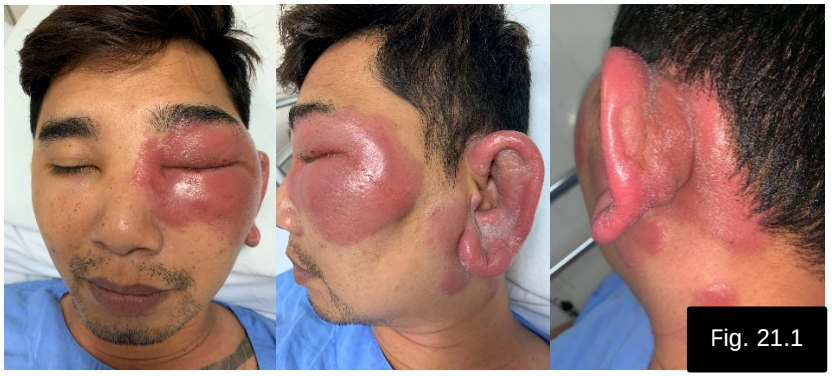
Fig. 21.1

Present illness:
He had developed numbness around the left ear for 1 year but no rash was initially observed. A month prior to this visit, reddish numb rash was first noticed on the left cheek and then gradually involved the left ear and the left periorbital area. He was neither bothered by pain nor itch. He then sought medical attention and had been diagnosed with orbital cellulitis. Although a course of appropriate intravenous antibiotic was given, he reported that the rash became swollen and progressive of 1-week duration. He had no history of fever or weight loss. He was therefore transferred to our hospital due to worsening of the rash.

Past history: He has no underlying diseases and denied a history of leprosy contact.

Family history: There was no family history of similar lesions.

Physical examination:
- Vital signs: normal
- Eyes: conjunctival injection and chemosis of the left eye, pupils 3 mm RTLBE, negative RAPD, full EOM, VA 20/20 both eyes
- Lymph nodes: left cervical lymphadenopathy
- Neurologic system: decreased light touch and pinprick sensation on the lesional skin as well as on the areas supplied by left supraorbital and left great auricular nerve. Thickening and tenderness of the left supratrochlear and left great auricular nerve.
- Other systemic examinations were otherwise unremarkable.

Dermatological examination: (Fig. 21.1)
Few discrete well-defined infiltrated edematous erythematous plaques on the left periorbital area, left ear, pre- and post- auricular area, and on the left side of the neck. Few erythematous papules asymmetrically scattered on the trunk and abdomen.

Histopathology: (S19-020464, neck)

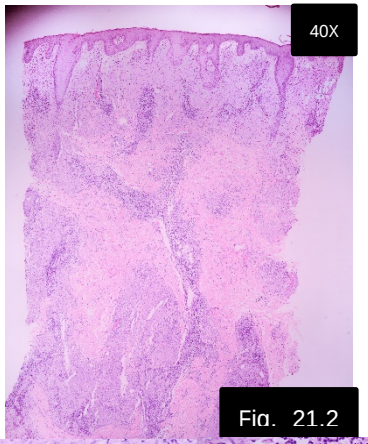

Fig. 21.2

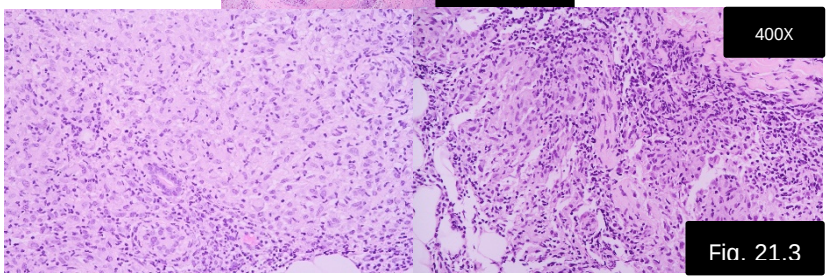

Fig. 21.3

- Nodular infiltrate of histiocytes, some giant tuberculoid cell histiocytes admixed with lymphocytes and plasma cells forming granulomatous dermatitis. (Fig. 21.2)
- Perineural involvement and marked papillary dermal edema are noted. (Fig. 21.3)
- Negative AFB and Fite staining.

Laboratory investigations:

- Slit-skin smear: right ear 2+, left ear 3+, neck (lesion) 3+, trunk (lesion) negative
- CBC: Hb 15 g/dL, Hct 45%, Plt 328,000 /mm^3, WBC 6,410 /mm^3 (N 69%, L 24, M 5%, E 0%, Band 2%)
- AST/ALT: 19/52 U/L
- BUN/Cr: 18/1.0 mg/dl
- Anti-HIV: negative
- CT Brain and orbits: preseptal cellulitis of the left orbit with associated soft tissue inflammation is likely but no definite evidence of postseptal involvement or subperiosteal abscess.

Diagnosis: Borderline lepromatous leprosy with type 1 reaction
Treatment:

- Rifampicin (300 mg) 2 tabs monthly
- Clofazimine (300 mg) 1 tab monthly
- Clofazimine (50 mg) 1 tab daily
- Dapsone (100 mg) 1 tab daily
- Prednisolone (5 mg) 6 tabs twice daily

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Discussion:

Leprosy is a chronic granulomatous disease that is caused by the acid-fast bacillus Mycobacterium leprae. It involves mainly the skin and the peripheral nerves.[1] Although the route of transmission has not clearly understood, droplet infection through the nasal mucosa is assumed. Clinical manifestations vary considerably depending on the patient's genetically determined immune status in relation to pathogen and also the miroorganism's tropism for the skin and nerve tissue. Generally, leprosy is classified into 5 major types including tuberculoid (TT), borderline tuberculoid (BT), mid-borderline (BB) borderline lepromatous (BL), and lepromatous (LL) forms. Tuberculoid form, which is characterized by sharply demarcated erythematous patches, is seen when host immunity is good. In contrast, patients represent multiple red-brown nodular infiltrates in a symmetrical pattern, sometimes with leonine facies, if they have impaired T-cell immunity. However, the majority of infected individuals present with borderline or intermediate form showing variable clinical features of tuberculoid and lepromatous leprosy.[2]

Diagnosis can be made based on clinical features, slit-skin smear test, and histopathological findings. In addition, available serologic tests include a polymerase chain reaction for M.leprae and a phenolic glycolipid1 antigen (PGL-1) antibodies by means of enzyme-linked immunosorbent assay (ELISA) but the latter is beneficial only in cases with high bacterial load.[3]

Leprosy reactions are results of the host immune responses to M.leprae and can occur before, during, or after treatment. Two types including type 1 (reversal reaction) and type 2 (erythema nodosum leprosum or ENL) are described. Type 1 reaction is defined by the worsening of pre-existing skin lesions or the formation of new skin lesions, with edematous erythema.[4] Nerves can become thickened and painful, and cause signs of deterioration of neurological functions.[5] It is believed a consequence of the imbalance in pro-inflammatory cytokines (gamma interferon and interleukin-2) and anti-inflammatory cytokines (especially TGF beta and interleukin 10). Clinically, type 1 reaction can mimic erysipelas, cellulitis, drug eruptions, urticaria, psoriasis, sarcoidosis, lymphomas, sudden nerve paralysis, and even leprosy relapse itself.[6] Since it is sometimes misdiagnosed with facial or periorbital cellulitis, like in our case, the appropriate treatment may be delayed. This potentially results in neuritis and subsequent lagophthalmos.[7, 8] These carry a risk of

developing keratitis, ulcer, infection, and blindness.[1] Early recognition and prompt treatment is therefore crucial to prevent complications. Type 2 reaction is associated with excessive humoral immunity and caused by the formation of immune complex. It is typically seen in patients receiving treatment and represents symmetrically distributed subcutaneous inflammatory nodules or ENL, that is a type of small vessel vasculitis. General symptoms such as fever, malaise, myalgia, edema, arthralgia, or lymphadenopathy can occur.[1] Nonetheless, neuritis is less common than in type 1 reaction.[9]

In our case, the diagnosis of borderline lepromatous leprosy was established according to the compatible lesions which were multiple, asymmetric, infiltrative papules and plaques with diminished sensation together with thickening of the nerves and confirmed laboratory findings including slit skin smear and histopathologic study. The patient also suffered from type 1 reaction due to the worsening and edema of the skin lesions, neurologic alterations, as well as peripheral nerve tenderness.

Regarding treatment, WHO's recommendation is based on multidrug regimen including rifampicin, clofazimine, and dapsone for the first-line treatment of leprosy.[3] In multibacillary (MB) cases, as in this report, 2-year administration of the following agents should be given; rifampicin 600 mg monthly, clofazimine 300 mg monthly and 50 mg daily, and dapsone 100 mg daily.[10] In terms of type 1 reaction, especially with neural involvement, prednisolone is suggested in the dose of 1-2 mg/kg/day to suppress the cellular immune response. The dose can be adjusted following the clinical response and upon decreasing to the dose of 20 mg/day, then should be maintained for a long period of time until clinical resolution and complete recovery of neural functions are observed. General improvement is typically obtained within 3 or up to 6 months after the initiation of therapy. In our case, he developed lagophtalmos of the affected eye and then was sent for rehabilitation program. However, his rash and sensory function improved within 4 weeks after treatment.

References:

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