Case 23

Red to brown papules on the trunk

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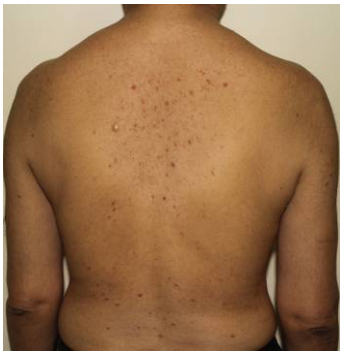
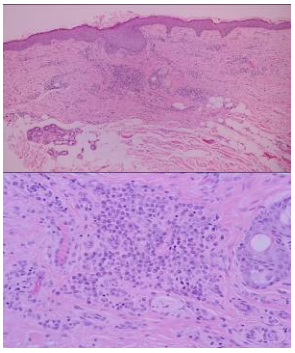
Patient: A 58-year-old Thai man from Nakhon Si Thammarat

Chief complaint:
Multiple brownish macules and papules on trunk for 1 year

Present illness:
The patient had initially developed multiple bilateral cervical, axillary and inguinal lymphadenopathy for 2 years. Later, he was referred to our dermatology department with multiple asymptomatic erythematous to brownish rashes on chest and back of a year's duration. The patient reported low-grade fever, fatigue and significant weight loss of 7 kilograms over a 1-year period.

Past history:
- Hypertension





Physical examination:
V/S: PR 99 bpm, RR 16 times/min, BP 123/83 mmHg
GA: A Thai man, good consciousness
HEENT: Mild pale conjunctiva, anicteric sclera
LN: Symmetrical multiple bilateral soft-consistency cervical, axillary and inguinal lymph nodes enlargement sized 1-3 cm
CVS and lung: Normal
Abdomen: Soft, not tender, no hepatosplenomegaly

Skin examination:
- Multiple discrete, non-tender, erythematous to brownish macules and papules with no scale on chest and back

Histopathology:
Skin (S13-31645, chest)
- Superficial and deep perivascular inflammatory-cell infiltration of numerous plasma cells admixed with a few lymphocytes in the dermis

Lymph node (cervical lymph node)

- Extensive plasma cell infiltration
- Immunostains:
  - Marked positive IgG4-positive plasma cell
  - No HHV-8 positive cell
  - Polyclonal type

Bone marrow

- Adequate cellular marrow, showing slightly decreased erythroid series and adequate myeloid and megakaryocytic series
- Plasma cells are increased

Investigation:

Laboratory tests

CBC: WBC 13,200 (N82%, L10%, Mono 7%, Eo1%), Hct 27.5%
MCV 82.8fL, Platelet 346,000/mm^3

LFT: AST/ALT 17/24 U/L ALP/GGT 77/56 U/L
TB/DB 0.3/0.2 mg/dL TP/Alb 110/17 g/L

SPEP: Blood Kappa: 305 mg/L (3.3-19), Lamda 221mg/L (5.7-26), Kappa/Lambda 1.446 (0.26-1.65)

Imaging study

CT scan of the chest and abdomen:

- Multiples enlarged lymph nodes at neck, axillary, mediastinum, intraabdominal and inguinal regions
- No hepatosplenomegaly
- No lung mass

Diagnosis: Multicentric Castleman's disease with cutaneous involvement

Treatment: Chemotherapy with COP regimen (cyclophosphamide, vincristine, prednisolone)

Discussion:

Castleman's Disease (CD) is an uncommon B-cell lymphoproliferative disorder characterized by lymph node hyperplasia with vascular proliferation. It was first described by Dr.Benjamin Castleman in 1956.[1] CD most often presents as an isolated mediastinal mass (unicentric), although multiple lymphadenopathies (multicentric) form of the disease also exists. Lesions can be nodal and/or extranodal. Histologic variants of CD are composed of hyaline-vascular (90%), plasma cell (10%), and a mixed type. CD is divided by clinical and histological finding into 4 subtypes[1], which are

1. Hyaline-vascular CD: Classic hyaline-vascular form, usually present with unicentrically localized of thoracic lymphadenopathy.

2. Plasma Cell CD: More commonly in multicentric than unicentric, frequently has constitutional symptom. This form has been associated with POEMs syndrome.

3. HHV-8-associated CD: Occurs in immunosuppressive/HIV-positive patients, presents with generalized lymphadenopathy and constitutional symptoms. High risk of progression to large B-cell lymphoma has been observed. Generally, the prognosis is poor with the survival being for months.

4. Multicentric CD: Unrelated to HHV-8 infection, occurred in older patient (6^{th} decade). Clinical presentations are generalized lymphadenopathy and constitutional symptoms. The histological subtype is usually plasma cell type. This type of CD is associated with increased risk of lymphoma.

Cutaneous involvement in CD is rare. Most of cases are presented in multicentric form.[2]The cutaneous lesion shows multiple erythematous to brownish nodules and plaques predominately on back, but it can be found on the face, trunk and extremities.[3-6] The skin and lymph node biopsies usually show nodular aggregates of lymphocytes and polyclonal proliferation of plasma cells.[2]

Surgical excision is effective for unicentric CD.[7] However there is no consensus for the treatment of the multicentric type. There is a variety of approaches including surgery, chemotherapy (CHOP or AVBD regimen), radiation therapy, specific antibodies and anti-viral therapies.[2,7,8]

Our patient presented with generalized lymphadenopathy, polyclonal hypergammoglobulinemia and multiple cutaneous erythematous to brownish rashes. Histopathology from cutaneous eruption, cervical lymph node and bone marrow biopsy showed extensive plasma cells infiltration. With all findings, he was diagnosed with multicentric Castleman's disease with cutaneous involvement.

Our patients received 7 cycles of COP regimen; cutaneous lesions were improved as well as the generalized lymphadenopathy decreased in size and number without major adverse effect from chemotherapy.

References:

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