

CASE REPORT

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Estonia

Case report

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- **Internal Medicine Department in August 2009**
- 63 year old male, retired butcher
- Symptoms: fever ($>39^{\circ}\text{C}$), malaise, fatigue, itching skin rash, decreased appetite, weight loss ($\sim 10\text{kg}$ within past few months)
- Duration of symptoms: intermittently throughout past 7 years

Case report: *past medical history (1)*

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□ **2002 Gastroenterology Department**

- Fever, itching skin rash, lymphadenopathy
- Severe anemia (Hgb 55g/l; microcytic), hypergammaglobulinemia (polyclonal)
- Diagnostic hypothesis: myeloma-> referred to hematologist.
 - Investigations (including bone marrow biopsy) did not confirm the diagnosis
 - Lymph node biopsy – patient refused

□ **2006 Pulmonary tuberculosis (culture positive)**

- Treated 6 months, recovered

Case report: *past medical history* (2)

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□ **2009 April, Gastroenterology Department**

- Abdominal pain, constipation, fever, lymphadenopathy
- Anemia (Hgb 81 g/l, microcytic), hypergammaglobulinemia
- Gastroscopy, colonoscopy, capsule endoscopy: normal findings

□ **2009 May, Hematology, outpatient**

- Lymph node biopsy: plasmacytic infiltration reactive to chronic inflammation

Case report:

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- **Internal Medicine Department in August 2009**
- Examination:
 - Cachectic man
 - Bilateral axillar and inguinal non-tender lymphadenopathy (Ø 3cm)
 - No palpable organomegaly
 - On the face, neck, trunk, extremities - erythematous maculopapular rash
 - Clubbing fingers
 - Temperature 39.2°C

Skin finding and clubbing fingers

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Case report

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□ Analyses

- Anemia: Hgb 66 (134-170g/l), microcytic
- ESR 116 (<14mm/H); CRP 124 (<5 mg/l)
- Albumin: 25 g/l (34-48 g/l)
- IgG 35.4 (7.0-16.0 g/l); IgM 12.58 (0.40-2.30 g/l); IgA 3,07 (0.70-4.00 g/l)
- Beta2- microglobuline 6004 (609-2366 µg/L)
- Liver and kidney tests without changes
- Urine analysis normal
- **CT** – widespread lymphadenopathy (neck, axillae, chest/mediastinum, abdomen, pelvis), without organomegaly

Case report: *differential diagnosis (1)*

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□ Myeloma?

- No lytic lesions
- No M-spike (polyclonal gammopathy)
- Bone marrow aspiration and biopsy: mild plasmacytic infiltration (<10%)

□ Lymphoma?

- No histological proof

□ Sarcoidosis?

- iCa 1.18 (1.17-1.29 mmol/l); ACE 52 (8-52U/l)
- Bronchoscopy (done 2006): bronchoalveolar lavage negative
- No histological proof

Case report: *differential diagnosis* (2)

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- **Infection (bacterial, viral)?**
 - Urine, blood cultures negative
 - Several antibiotics without effect
 - HIV, CMV, EBV, Toxoplasma, Tbc negative
- **Atypical tuberculosis?**
 - Negative blood, urine cultures
 - Does not explain hypergammaglobulinemia
 - Skin and lymphnode histology: no granulomas
- **Autoimmune disease?**
 - Autoantibodies negative

Case report

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▣ Further investigations:

- Re-examination of excised lymph node: histology matches with **Castleman's disease (multicentric plasmacytic form)**

Castleman's disease

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- Lymphoproliferative disorder
 - Rare disease, precise incidence is not known
 - Histologically: angiofollicular lymph node hypertrophy
- **2 main forms:**
 - Unicentric: asymptomatic, localized, cured by surgical resection
 - Multicentric: aggressive, systemic symptoms, diffuse lymphadenopathy, association with HHV-8, HIV
 - Asthenia, weight loss, fever, polyadenopathy, hepatosplenomegaly
 - Anemia, thrombocytopenia, hypoalbuminemia, polyclonal hypergammaglobulinemia, increased CRP and ESR

Castleman's disease

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- Etiology is poorly understood
 - Immunoregulatory deficit -> B-cell lymphoproliferation -> outgrowth of polyclonal B-cell populations
 - IL-6 overproduction
 - Viral infection: HIV; HHV-8
- Treatment
 - Glucocorticoids
 - Chemotherapy
 - Anti IL-6 antibodies
- 5 year survival rate in multicentric form is 82%
 - Prognosis is better than with malignant lymphomas

How our patient is doing today?

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- Treatment: CVP- Cyclophosphamid 1.3g i/v;
Vincristine 1mg i/v; Prednisolone 100mg p/o
- After first treatment - feeling good, no fever
- CRP decreased 108mg -> 34 mg/l