



UNIVERSITY OF TARTU

Case report

ESIM 2014

Winterschool in Saas-Fee

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Estonia



Presentation

- Male, 27 years old
- Allergic to fish
- Non-smoker
- Previously diagnosed autoimmune thyroiditis in June, 2012
- Takes L-Thyroxine 100 μg daily
- No operations done



August, 2012

- Complaints:
 - weight gain
 - oedema of the legs
 - lower back pains
 - low urine output
- Admitted to the department of nephrology



Objective status

- Pitting oedema on the legs to the knees
- On the left side of the neck – 2 cm diameter lymph node (painless, appeared 3-4 months ago)
- RR 151/86 mmHg, HR 80 x/min, SpO2 98%
- Diuresis 1000ml/die
- Otherwise normal



Workup

- Urine: U-Prot 4+, U-Ery 2+
- Proteinuria **35,1** g/die (negative)
- Blood: Hgb 160 g/l (130-180 g/l)
Ery $5,79 \times 10^{12}/l$ ($4,5-6,0 \times 10^{12}/l$)
Leu $3,98 \times 10^9/l$ ($4-9 \times 10^9/l$)
Plt $213 \times 10^9/l$ ($150-400 \times 10^9/l$)
Eosinophils **$3,67 \times 10^9/l$** ($0,1-0,4 \times 10^9/l$)
30,5% (1-4%)
- Biochemistry: Cholesterol **11,6 mmol/l** (<5 mmol/l)
Albumin **15,8 g/l** (31-46 g/l)
Protein **36,9 g/l** (66-87 g/l)
Creatinine 76 $\mu\text{mol/L}$ (62-109 $\mu\text{mol/L}$)
TSH **15,9** mIU/L (0,4-4,0 mIU/L)



Nephrotic syndrome

- Proteinuria, hypoproteinaemia, hypercholesterolaemia, oedema

+

- Marked eosinophilia!



Additional analysis

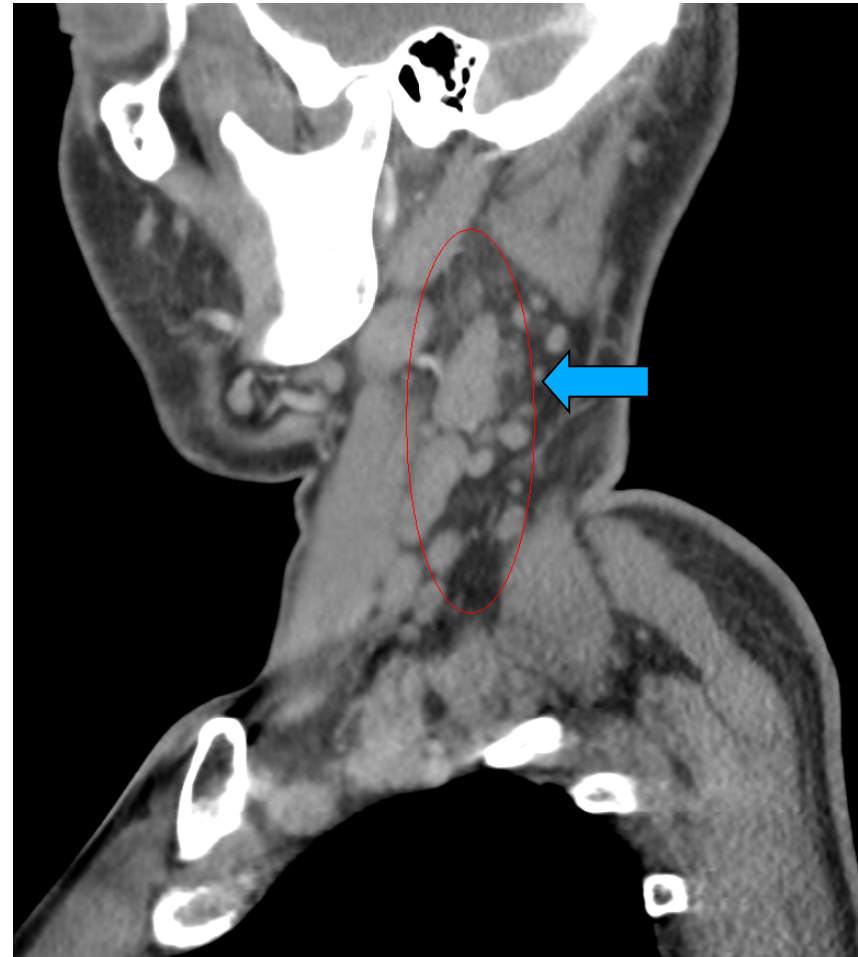
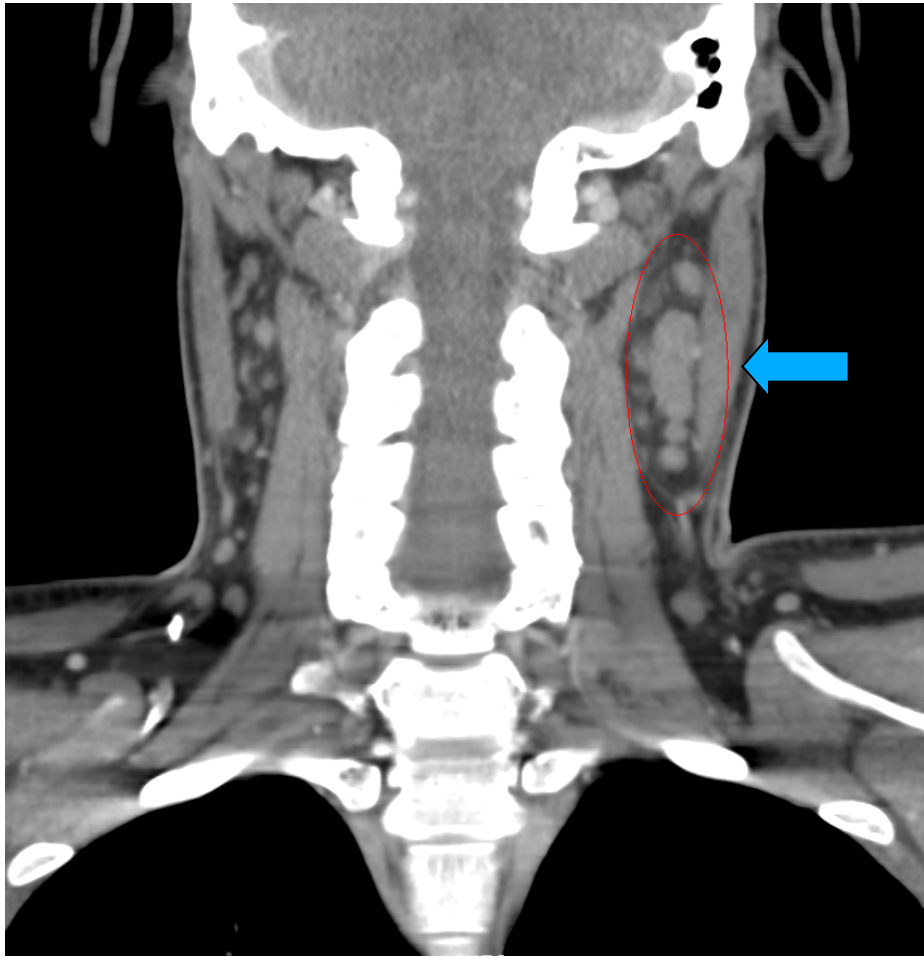
- C3, C4, AOS-titer normal
- HIV, EBV, CMV, hepatitis B, C serology negative
- ANA, ANCA, anti-GBM antibodies negative
- IgE **2751 kU/l** (<100 kU/l)
- IgE to various allergens - negative
- Giardia lamblia, Cryptosporidium parvum, Entamoeba histolytica in stools - negative



Additional tests

- Abdominal US: thickened renal parenchyma, uneven echogeneity
 - US of the neck: several 5-9 mm diameter lymph nodes on the neck
 - Echocardiography: normal
 - CT: neck lymph nodes enlargement both sides, sin>dex
- Lymph nodes also enlarged in mediastinum
- Kidney biopsy: minimal change disease, no malignant changes

CT neck and thorax





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Hypothesis?



Differential diagnosis

Lymphoma?

- Lymph node excisional biopsy: reactive lymphoid hyperplasia, eosinophilia, eosinophilic abscesses
No T- or B-cell clonality
- Bone marrow biopsy: normal cellular erythropoiesis, serous degeneration in stroma, eosinophilia

Hypereosinophilic syndrome?

- Chromosomal and FISH: FIP1L1-PDGFR α (4q12) deletion – negative

Allergic reaction? Parasitic infection? Drugs?



Kimura's disease (eosinophilic follicular hyperplasia)

- Endemic in Asia, rare elsewhere
- First histological description 1948 Kimura et al.
- Young males (20-40 years)
- Etiology unclear (trauma, allergic reaction, autoimmune?)
- Clinical findings:
 - unilateral enlarged lymph nodes in neck area
 - may be salivary gland involvement
 - 20% renal involvement (minimal change disease, membranous or mesangioproliferative glomerulonephritis)
 - labs: eosinophilia, proteinuria, IgE ↑
 - IL-4, IL-5, IL-13 ↑
 - histology: eosinophilic infiltration in lymph nodes, eosinophilic abscesses



Treatment

- Steroids:
Methylprednisolone pulse 1g iv (3 days)
Oral prednisone 48 mg → 4 mg
- S. Albumin 20% 100,0 iv
- T. Telmisartan 40 mg x 1
- T. L-Thyroxin 150 µg x 1

- Surgical removal of lymph nodes (recurrence common)
- Histamine 1 receptor blockators?
- Radiation therapy?
- Cyclosporine, azathioprine?
- Anti-IgE antibodies (omalizumab)?



Take home message!

Kimura's disease

- Young male asians
- Rare disease
- Eosinophilia and IgE ↑
- Enlargement of cervical lymph nodes – differential diagnosis!





Thank you!

