Widening Myopathological Spectrum associated with Plasma Cell Dyscrasia

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History prior to referral

• 74 year-old male
• 1½ years of problems with lifting his arms, hand-grip good
• No relevant family history
• Biopsy: Inflammatory and dystrophic features
• Treatment attempts without improvement:
  – Off Atorvastatin, previously on for 14 years
  – 3 months of Prednisolone and Methotrexate
• Fascio-scapulo-humeral dystrophy?
Neurological and laboratory examination

- CN normal
- Arms
  - *Proximal weakness, wasting of deltoids: marked and symmetric*, periscapular preservation
  - Mild weakness of biceps and triceps
- Legs
  - *Mild hip flexion weakness*, distal strength normal
- Hyperlordotic abdomen with myopathic gait
- CK 950 IU/L
- Myositis-specific autoantibodies negative
- D4Z4 repeat analysis normal (double digest)
CD 138
Kappa light chain
Lambda light chain
Kappa light chain

Congo red
Follow up investigations

- Immunofixation: IgG kappa paraprotein 8 g/L
- No indication of typical end organ damage
- Patient not available for further investigations
Case summary

- Symmetric proximal weakness with pronounced wasting of deltoids
- End-stage inflammatory myopathy
- Monoclonal, kappa-restricted plasma cell infiltrates
- Light chain deposition
- Diagnosis: Plasma cell dyscrasia
- Manifestations of multiple myeloma in skeletal muscle?
Skeletal muscle involvement in multiple myeloma

• Autopsy studies: 15% of patients
  – Buerger et al. 1966: 15% (1/7) of multiple myeloma patients; multiple muscles affected; no gross pathological abnormalities
  – Acinas Garcia et al. 1984: 20 % of skeletal muscle metastatic disease due to lymphoma (34 of 723 cancer patients had muscle metastases)

• Radiological studies: 2% with diffuse swelling or focal mass
  – Frequency: 6/270 = 2 % of multiple myeloma patients with muscle involvement (Surov et al., 2010; 10 year review)
  – Diffuse muscle swelling or intramuscular mass (Surov et al. 2014)
  – Selection bias for masses
Paraproteinemia and protein deposition disease in skeletal muscle

- Amyloid

- Light chain deposition (Zarkrzewski et al., Ophthalmologica 224, 2010; Ostrow et al., Muscle Nerve, 45(5), 2012)

- Disulfide bonds and multimerisation (Kaplan et al., Br J Haematol 144(5), 2009)
Multiple myeloma and inflammatory myopathy
Evidence from the literature

• Kappa light chain deposition and IgD deposition
  (Kiprov et al., Lancet 8413(2), 1984; Colombo et al., Neurol Res Int 2010)

• Non-secretory multiple myeloma (Vallabhajosyula et al.,

• Paraneoplastic DM/PM (0.3-0.6% of patients with underlying multiple myeloma; Marie et al.,
  Autoimmun Rev 11(9), 2012; multicentre review, 10% with haematologic malignancy)
Muscle pathology associated with plasma cell dyscrasia

- Infiltration
- Protein deposition
- Inflammation

Clinical presentations of muscle involvement in plasma cell dyscrasia

- Diffuse muscle swelling
- Focal mass
- Symmetric pseudohypertrophy
- Symmetric wasting
- Symmetric weakness
Conclusion

Multiple myeloma can be in the differential of an inflammatory myopathy.
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