lowed by toxic mitochondrialopathy. These results suggest that a test for HIV is advised in case of polymyositis.

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P5.27
Evidence for the Implication of Th-1 and Treg cells but not Th-17 in sporadic Inclusion Body Myositis

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Sporadic Inclusion Body Myositis (sIBM) is an inflammatory myopathy characterized by CD8+ cytotoxic infiltrates and amyloid deposits. Regulatory T cells (Treg) are key regulators of immune response. We analysed 54 parameters of the immune response, in attempt to develop new therapeutic approaches. We included 22 definite sIBM patients (mean age: 70.1) and 22 controls (matched on age and sex). No sIBM patients nor controls received immunosuppressants. Peripheral blood mononuclear cells (PBMC) were analyzed using flow cytometry. Using a multiplex assay, the concentrations of 25 cytokines and chemokines was determined. Muscle biopsies of 8 patients were tested by immunohistochemistry for the presence of Treg. In blood, mean percentage of activated CD4+ T cells (16.2 ± 13% vs 8.7 ± 4.6%; p = 0.03) and terminally differentiated CD8+ CD28-T cells (61 ± 23.9% vs 44 ± 20%; p = 0.023) was increased in sIBM patients compared to controls. The mean percentage of CD3+ CD8+ IFNγamma+ was higher in sIBM patients (60.8 ± 18% vs 45.8 ± 16%; p = 0.01) whereas the mean percentage and CD3+ CD4+ IL-17+ was not statistically different in both groups. We observed an increase of IL-12 concentration in the sera of sIBM patients (301.36 ± 142.08 pg/ml vs 154.25 ± 28.41 pg/ml, p = 0.0002) and of the Th-1 chemokines such as CXCL-9 (186 ± 12 pg/ml vs 13 ± 7 pg/ml, p < 0.0001), and CXCL-10 (187 ± 62 pg/ml vs 13 ± 6 pg/ml, p < 0.0001).

The percentage of Treg among CD4+ T cells was lower in sIBM patients (5.5 ± 0.3% vs 6.6 ± 0.4%, p = 0.043). Treg cells were detected in muscle biopsies and represented 18.7 ± 7.4% of CD4+ cells in the muscle. Together these results suggest that CD4+ and CD8+ T cells are more activated and engaged in a Th1 lineage in sIBM. Treg cells are decreased in blood whereas they are present in muscle inflammatory infiltrates, where they seem unable to control effector cells.

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P5.28
Culture supernatants from HTLV-1 infected T cells modify adhesion molecule-related gene signature in differentiated human muscle

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Human T-cell lymphotropic virus type 1 (HTLV-1) is a human retrovirus and the etiologic agent for a progressive neurological disease called HTLV-1-associated myelopathy/tropical spastic paraparesis (HAM/TSP), which primarily affects the thoracic spinal cord and is associated with inflammatory muscle diseases such as polymyositis. However, HTLV-1 virus is detected in infiltrating CD4+ lymphocytes but not in muscle fibers. Evidence in the literature supports the hypothesis that myositis is mediated by the release of cytokines and/or the viral Tax transactivator by HTLV-1 infected mononuclear cells that infiltrate the muscle. We investigated the gene expression of cultured human muscle cells in response to soluble factors secreted by infected HTLV-1 T cells, and if this modulation could induce T cell recruiting to the inflammatory muscle sites. Cultured myotubes, resulting from in vitro differentiation of human myoblasts, were incubated for one hour with: (1) PBS; (2) the 30–50 KDa supernatant protein fraction from a HTLV-1 infected T cell line derived from a HAM/TSP patient; or, (3) the same protein fraction from Jurkat cells, a human non-infected leukemia T cell line. After this incubation we washed the myotubes and did the RNA extraction for real-time RT-PCR. We found an upregulation of various integrin, laminin and other adhesion molecule-related genes in myotubes stimulated by the HTLV-1 infected T cell-derived culture supernatant. These findings reveal that myotubes respond to HTLV-1 T cell secreted factors, which modify the adhesive microenvironment, which in turn may be involved in the increased recruiting of T cells to inflammatory muscle sites.

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P5.29
Quadriceps extensor strength is a sensitive marker for disease progression in sIBM patients

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Sporadic inclusion body myositis (sIBM) is the most frequent myositis after the age of 50 years old. To date there is no validated treatment. Robust and reliable outcome measures are needed in attempt to develop new therapeutic approaches. Following a slowly progressive disorder as muscle weakness in sIBM patients involves sensitive and reproducible marker. The aim of this prospective study was to develop sensitive evaluation tools in sIBM patients. 22 patients with definite sIBM, not treated by any immunosuppressants, were assessed using manual muscle testing, non specific (Walton scale) and specific (weakness composite score for sIBM: WCSI, and Inclusion Body Myositis Functional Rating Scale: IBMFRS) functional scales, 6 min walk test (6WMT), strength dynamometry of hand grip and elbow, wrist, ankle and knee extension and flexion. All strength measurements were performed in isometric conditions. Data for strength and 6MWT were converted into percentages of normal. Among the 22 patients, 16 were reassessed 9 months later. Baseline line values showed a significant decrease of the muscular strength compared to normal values for all muscle groups tested (46 ± 15%). The weakest muscle functions were hand grip (36.7 ± 19.4%) at the upper limbs and knee extension at the lower limb (30.3 ± 26.8%). The patient strength was correlated with the disease duration only for knee extension (r = −0.7, p = 0.001). Moreover, knee extension strength was significantly correlated with all other measures (Walton scale, IBMFRS, WCSI and 6MWT). After 9 months, only knee extension strength showed a significant decrease (−7.2%, p = 0.003) whereas others measures including functional scales failed to detect any significant change. This study suggests that the quadriceps extensor strength is the most sensitive marker for disease progression in sIBM patients.

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P5.30
Inclusion body myositis: A diagnostic challenge

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The currently accepted diagnostic criteria for inclusion body myositis (IBM), first published in 1995, comprise pathological and clinical elements. The pathological features being endomyal inflammation with invasion of intact fibres, rimmed vacuoles and either amyloid or tubulofilamentous inclusions; all of these must be present for a definite diagnosis. However, these features are not specific to IBM and furthermore in many clinically typical cases one or more may be absent. Recently a variety of proteins more commonly associated with neurodegenerative diseases have been found to accumulate in muscle fibres in IBM. The benefit of immunohistochemical staining for these proteins in the diagnosis of IBM is unknown. We propose that these protein accumulations may assist in the diagnosis of IBM and help in differentiating it from other myopathies.

To identify the most sensitive immunohistochemical techniques available to UK diagnostic pathology laboratories of detecting such protein accumulations and to determine their sensitivity and specificity for the diagnosis of IBM. We identified 6 cases of pathologically definite IBM according to the current criteria and 6 normal control cases. All cases had been assessed clinically and had undergone a biopsy at the National Hospital for Neurology and Neurosurgery, Queen Square, London. A number of neurodegenerative proteins and inflammatory markers were identified that may be of diagnostic significance. Antibodies to these proteins were optimised using control tissue known to contain the protein of interest. Protein inclusions found in IBM were labelled with an antibody to p62, an adapter protein which binds ubiquitin and regulates signalling cascades through ubiquitination. These inclusions were not found in the normal controls. Further work to quantify the abnormalities in IBM and disease controls will be undertaken before any of these markers can be recommended for diagnostic use.


P5.31
Clinical, morphological and magnetic resonance imaging findings in sporadic inclusion body myositis

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In sporadic inclusion body myositis (sIBM), there is in most cases a characteristic pattern of muscle weakness. The primary cause of muscle destruction is debated. We have investigated 20 patients with definite sIBM in order to describe what disease causing mechanisms may be of importance in early (strong muscles) and late stages (weaker muscles). The tibialis anterior (T), quadriceps (Q) and biceps brachii (B) were chosen. Muscle strength was measured by a hand-held dynamometer and quantitative electromyography was done, analysing the duration, the amplitude, the area, and the number of phases and turns of the motor unit action potential (MUAP). We also evaluated spontaneous muscle fibre activity and the interference pattern. Data from all 18 cases showed that the duration and the area of the MUAP positively correlated at 0.01 level to strength for T and Q. In addition to that, the MUAP amplitude showed a positive correlation at 0.01 level to strength for T. No significance was found for B. Interference pattern was myopathic in majority of muscles and the amount of spontaneous activity showed no correlation to strength. Comparing means of the MUAP parameters between T and Q, in 15 cases in whom T was stronger than Q, showed that the amplitude was lower in the weaker Q, at the significance level 0.05. We had assumed that the weaker muscle should have shorter MUAP duration and less number of phases and turns, but these parameters showed no significant difference between the stronger and weaker muscle. In conclusion, our study of the MUAP parameters was not helpful to explain the mechanisms of importance for the evolvement of muscle weakness in sIBM. We discuss our findings in relation to previous studies.

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P5.33
Brachio-cervical inflammatory myopathy – A case report

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Myopathies can be classified by their etiology and pathophysiology, by their clinical phenomenology, or, by their underlying genetic defects. Brachio-cervical inflammatory myopathy (BCIM) is an acquired myopathy characterized by weakness in a brachial and posterior cervical distribution bilaterally, associated with a predominant active myopathy on biopsy: C5b-9 staining of endomyosium, focal perivascular and perimysial inflammation, often with a prominent B cell component, and endomysial dendritic cells. It remains to be determined whether BCIM syndromes constitute either a clinical phenotype or a distinct class of immunemediated myopathies. A 68-year-old woman was observed in the outpatient clinic with a history of progressive posterior neck, proximal arms and legs weakness producing difficulty in climbing stairs and walking, for a