Extended Abstract Journal of Neuroinfectious Diseases 2020
Vol.11 No.5

Clinical, Histopathological and Neurophysiological Differential Patterns in Sporadic Inclusion-Body Myositis and Hereditary Inclusion Body Myopathy

Giovanni Antioco Putzu
Casa di Cura Sant'Elena, Italy

Abstract
Sporadic Inclusion Body myositis (s-IBM) represents a form of chronic polymyositis unresponsive to towards the corticosteroids, affecting patients over of 50 members. In contrast the hereditary Inclusion-Body Myopathy (h-IBM) strikes younger patients. Clinical hallmark of both forms are distal muscle involvement whereas the salient histopathological features were characterised by inflammatory exudates (only in s-IBM), rimmed vacuoles, eosinophilic cytoplasmic inclusions, 16 to 18 nm tubulofilamentous inclusions in both cytoplasm and as well as in nucleus. Small amyloid deposits near or within the vacuoles and within the nucleus as well as the nuclear membrane abnormalities and nuclear breakdown and other findings such as angulated and round fibres, necrotic-regenerative fibres and even ragged red fibres. None of these hallmarks were specific of IBM and can also be found in a great number of muscle and even the nerve pathologies such as Oculopharyngeal muscular Distrophy (OPMD), Desminopathies, Glycogenosis, Ceroid lipofuscinosis, ALS, Post-polio syndrome, Paraneoplastic neuropathies, and many others that we will be illustrated and discussed in the presentation. Neurophysiological findings of s-IBM and h-IBM are not specific and include mixed myogenic and neurogenic features. In conclusion the s-IBM and h-IBM both are an interesting group of muscle pathology with a complicated differential diagnostic process. It also represents a challenge to both Clinicians and Researchers involved in the neuromuscular disorders field.

Introduction:
An Inflammatory myopathies represent the main group of acquired myopathies in the clinical practice with a well-established clinical, laboratorial, imaging and also therapeutical aspects related to the idiopathic and paraneoplastic polymyositis and dermatomyositis. Despite of it's the designation and classification as an inflammatory myopathy by most of authors in the past. The Inclusion body myopathy or myositis has been progressively recognized as a chronic degenerative muscle disease. The frequent changes regarding the pathophysiological processes were the absence of the clinical response to a definite specific therapeutic approach and the recognition of new sporadic and hereditary clinical presentations are disclosing several heterogeneous facets about IBM complexity. Sporadic IBM (sIBM) represents the main acquired myopathy in patients over the age of about 50 years. Being frequently underdiagnosed, especially in the early stages of clinical compromise in which there is a clear predominance of selective weakness in long flexors of the fingers and forearms and quadriceps femoris before progression to scapular girdle, lower limb distal groups and bulbar regions. There are well-established clinic pathological diagnostic criteria described by the European Neuromuscular Centre which are currently used to diagnose sIBM with clinical and research purposes.

An Inflammatory myopathies represent the main group of acquired myopathies in the clinical practice with a well-established clinical, laboratorial, imaging and also therapeutical aspects related to the idiopathic and paraneoplastic polymyositis and dermatomyositis. Despite of it's the designation and classification as an inflammatory myopathy by most of authors in the past. The Inclusion body myopathy or myositis has been progressively recognized as a chronic degenerative muscle disease. The frequent changes regarding the pathophysiological processes were the absence of the clinical response to a definite specific therapeutic approach and the recognition of new sporadic and hereditary clinical presentations are disclosing several heterogeneous facets about IBM complexity. Sporadic IBM (sIBM) represents the main acquired myopathy in patients over the age of about 50 years. Being frequently underdiagnosed, especially in the early stages of clinical compromise in which there is a clear predominance of selective weakness in long flexors of the fingers and forearms and quadriceps femoris before progression to scapular girdle, lower limb distal groups and bulbar regions. There are well-established clinic pathological diagnostic criteria described by the European Neuromuscular Centre which are currently used to diagnose sIBM with clinical and research purposes. An Inflammatory myopathies represent the main group of acquired myopathies in the clinical practice with a well-established clinical, laboratorial, imaging and also therapeutical aspects related to the idiopathic and paraneoplastic polymyositis and dermatomyositis. Despite of it's the designation and classification as an inflammatory myopathy by most of authors in the past. The Inclusion body myopathy or myositis has been progressively recognized as a chronic degenerative muscle disease. The frequent changes regarding the pathophysiological processes were the absence of the clinical response to a definite specific therapeutic approach and the recognition of new sporadic and hereditary clinical presentations are disclosing several heterogeneous facets about IBM complexity. Sporadic IBM (sIBM) represents the main acquired myopathy in patients over the age of about 50 years. Being frequently underdiagnosed, especially in the early stages of clinical compromise in which there is a clear predominance of

This work is partly presented at 30th International Conference on Neurology & Cognitive Neuroscience, February 24-25, 2020 at London, UK

selective weakness in long flexors of the fingers and forearms and quadriceps femoris before progression to scapular girdle, lower limb distal groups and bulbar regions. There are well-established clinic pathological diagnostic criteria described by the European Neuromuscular Centre which are currently used to diagnose sIBM with clinical and research purposes. An Inflammatory myopathies represent the main group of acquired myopathies in the clinical practice with a well-established clinical, laboratorial, imaging and also therapeutical aspects related to the idiopathic and paraneoplastic polymyositis and dermatomyositis. Despite of it's the designation and classification as an inflammatory myopathy by most of authors in the past. The Inclusion body myopathy or myositis has been progressively recognized as a chronic degenerative muscle disease. The frequent changes regarding the pathophysiological processes were the absence of the clinical response to a definite specific therapeutic approach and the recognition of new sporadic and hereditary clinical presentations are disclosing several heterogeneous facets about IBM complexity. Sporadic IBM (sIBM) represents the main acquired myopathy in patients over the age of about 50 years. Being frequently underdiagnosed, especially in the early stages of clinical compromise in which there is a clear predominance of selective weakness in long flexors of the fingers and forearms and quadriceps femoris before progression to scapular girdle, lower limb distal groups and bulbar regions. There are well-established clinic pathological diagnostic criteria described by the European Neuromuscular Centre which are currently used to diagnose sIBM with clinical and research purposes.

Conclusion:

HIBM represents a rare and heterogeneous hereditary myopathy group. Certainly underdiagnosed, and associated with the complex neurogenetic and pathophysiological dysfunctions with proper clinical and pathological hallmarks depending on the metabolic or the degenerative basis involved. Previously we considered merely an extension of sporadic IBM, hIBM is distinct in different aspects and represents a widening and noteworthy group of hereditary vacuolar myopathies which makes them difficult to classify as a group. But also acts as a prototype of broadening neurogenetic spectrum of clinical conditions and offers a great opportunity to provide proper development of the targeted therapies based in molecular and genetic approaches.

This work is partly presented at 30th International Conference on Neurology & Cognitive Neuroscience, February 24-25, 2020 at London, UK