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Clinical, Histopathological and Neurophysiological differential patterns in Sporadic Inclusion-Body Myositis and Hereditary Inclusion body Myopathy

Sporadic Inclusion-Body myositis (s-IBM) represents a form of chronic polymyositis, unresponsive to corticosteroids, affecting patients over 50. In contrast, hereditary Inclusion-Body Myopathy (h-IBM) strikes younger patients. Clinical hallmark of both forms are distal muscle involvement, whereas salient histopathological features are characterised by inflammatory exudates (only in s-IBM), rimmed vacuoles, eosinophilic cytoplasmic inclusions, 16-18 nm tubulofilamentous inclusions in both cytoplasm and nucleus, small amyloid deposits near or within vacuoles and within the nucleus as well, 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 are specific of IBM and can be also found in a great number of muscle and even 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 (axonal) features.

In conclusion, s-IBM and h-IBM 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 field of neuromuscular disorders.

Biography

Giovanni Antioco Putzu is a Medical Doctor since 1992, with specialization in Paediatric Neurology. He achived his PhD in 1996. During PHD studies, He was a Research Fellow in Hammersmith Hospital of London, UK in 1992, then He moved in Marseille to work at INSERM (Genetics) and in Neurophathology. The Author has published more than 15 papers in the field of Neuromuscular Disorders.

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