ELECTIVE ROTATION IN NEUROMUSCULAR PATHOLOGY

Graduate Medical Education

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1. GENERAL DESCRIPTION

The elective rotation is a combination of didactic presentations and neuromuscular cases reviewing that intends to provide an up-to-date and comprehensive overview of muscle and nerve pathology with instructions on subtleties of advanced muscle and nerve biopsies diagnosis. Clinical, biochemical and molecular features that are relevant to pathology and/or neurology residents/fellows will also be included in order to provide a coherent understanding and background into various neuromuscular diseases.

Rotations are typically two weeks in duration with 2-3 hours of one-to-one instruction per day. Timeframes for each rotation are decided at least 30 days prior to the beginning of the rotation. Under special circumstances, elective rotations may be held online. Prior to the rotation, the resident will be asked to take an online quiz that aims at assessing his/her very basic anatomical/histological knowledge of the neuromuscular system.

2. LEARNING GOALS & OBJECTIVES

The main goal of the neuromuscular pathology elective rotation is to promote an increased understanding of the science and practice of neuromuscular pathology in the medical setting and its utility in relation to the residents' specialties.

The Neuromuscular pathology elective rotation is also intended to provide the residents with hands on experience in evaluating and identifying various histopathological features related to neuromuscular disorders using nerve or muscle biopsies. By the completion of the rotation, it is expected that participants will:

- ✓ Complete a basic review of the fundamental concepts in neuromuscular pathology and histology through a study of diseases processes in current and archived patient material

- ✓ Demonstrate a satisfactory level of diagnostic competence characterized by the ability to recognize most common histopathological features associated with neuromuscular disorders
- ✓ Gain skills enabling them to order appropriate stains outside routine neuromuscular related panels for the completion of their diagnostic studies
- ✓ Develop autonomy in requesting appropriate molecular studies when needed
- ✓ Gain experience in processing fresh nerve and muscle biopsies

3. TOPICS COVERED

MUSCLE	
1	Anatomy, Development, & Histology of Skeletal Muscle
2	Metabolic and Immune Features of Skeletal Muscle
3	Dystrophies vs Myopathies
4	Other dystrophies and allied diseases
5	Metabolic Myopathies
6	Inflammatory Myopathies
7	Neurogenic Myopathies
8	Channelopathies and Neuromuscular Junction Disorders
9	Toxic and other myopathies

NERVE	
1	Development, Anatomy & Histology of Peripheral nerves
2	Pathological Patterns of Peripheral Nerves
3	Histopathological Features of Peripheral Nerves (1)
4	Histopathological Features of Peripheral Nerves (2)

4. DETAILED OUTLINE

A. MUSCLE

Session1. Macroscopic & Microscopic Anatomy and Development of Skeletal Muscle

- ✓ Skeletal muscle development and anatomy
- ✓ Muscle biopsy procedure and sample processing
- ✓ Normal histological & ultrastructural features of skeletal muscle
  Routine histological studies and special panels
- ✓ Steps into diagnostic process 1

Session2. Metabolic and Immune Features of Skeletal Muscle

- ✓ Normal "histometabolic" features and fiber typing of skeletal muscle

Mitochondrial bioenergetics, fatty acid oxidation, glycogen metabolism, motor units, fiber typing, *etc*

- ✓ Normal "Immune" features of skeletal muscle
  Immune Privileges of the Skeletal Muscle: Major histocompatibility complex, resident immune cells, immunoglobulins, terminal complement complex, *etc*
- ✓ Steps into diagnostic process 2

Session3. Dystrophies vs Myopathies

- ✓ Clinical & histopathological features
  Congenital myopathies vs Congenital dystrophies

Session4. Other dystrophies and allied diseases

- ✓ Clinical & Histopathological features
  DMD/BMD, Limb-Girdle muscular dystrophies, Fascioscapulohumeral, myotonic, and oculopharyngeal muscular dystrophies, and Emery-Dreifuss muscular dystrophy

Session5. Metabolic Myopathies

- ✓ Clinical & Histopathological features
  Mitochondrial disorders, lipid myopathies, and glycogen storage diseases

Session6. Inflammatory Myopathies

- ✓ Clinical and Histopathological features
  Dermatomyositis, Immune-mediated necrotizing myopathies, anti-synthetase myositis, inclusion body myositis, macrophagic myofasciitis, and other rare forms of myositis *e.g.* sarcoidosis, focal myositis, proliferative myositis, *etc.*

Session7. Neurogenic Myopathies

- ✓ Clinical and Histopathological features
  Motor neuron diseases vs peripheral neuropathies

Session8. Myofibrillar Myopathies and Other Myopathies with Rimmed Vacuoles

- ✓ Clinical and Histopathological features
  Myofibrillar myopathies, other myopathies with autophagic vacuoles and protein aggregates

Session9. Channelopathies and Neuromuscular Junction Disorders

- ✓ Clinical and Histopathological features

  Neuromuscular junction disorders and channelopathies

Session10. Toxic and other myopathies

- ✓ Clinical and histopathological features
  - Toxic and drug induced myopathies
  - Endocrine disorders and myopathies
  - Muscle involvement in connective tissue diseases
  - Muscle disorders associated with infections

NOTE: Adapted neuromuscular cases are included in all sessions for reviewing and interpretation.

B. NERVE

Session1. Macroscopic & Microscopic Anatomy and Development of Peripheral nerves

- ✓ Peripheral nervous system developmental and anatomical features
- ✓ Nerve biopsy procedure and processing
- ✓ Normal histological & ultrastructural features of peripheral nerves

  Routine histological studies and special stains: Nerve fibers diameter-frequency, myelination, cellular reactions, proteinaceous deposits, vasculature…

Session2. Pathological Patterns of Peripheral Nerves

- ✓ Histological and physiological features
  - Wallerian degeneration
  - Axonal degeneration
  - Segmental demyelination
- ✓ Steps into diagnostic approach

Session3-4. Histopathological Features of Peripheral Nerves

- ✓ Histopathological features observed within:
  - Epineurium
  - Perineurium
  - Endoneurium
- ✓ Practical approach to nerve specimen examination

NOTE: Adapted neuromuscular cases are included in all sessions for reviewing and interpretation.

5. RESOURCES

Suggested textbooks

- ✓ ***Muscle Biopsy (4th and 5th edition): A practical approach.*** Victor Dubowitz, Caroline Sewry, and Anders Oldfors. ISBN: 9780702074714

- ✓ **Biopsy Diagnosis of Peripheral Neuropathy.** Juan M. BILBAO, and Robert E. Schmidt. ISBN 978-3-319-07311-8
- ✓ **Muscle Disease: Pathology and Genetics, 2nd Edition.** Hans H. Goebel, Caroline A. Sewry, Roy O. Weller. ISBN: 978-0-470-67205-1
- ✓ **Neuromuscular disorders of infancy, childhood, and adolescence. A Clinical Approach. Second edition. 2014.**. Basil T. Darras, H. Royden Jones, Jr, Monique M. Ryan, and Darryl C. De Vivo. ISBN 978-0-12-417044-5

Online resources

- ✓ **https://neuromuscular.wustl.edu/**

6. ASSESSMENT OF LEARNING OUTCOMES

Throughout the elective rotation, residents will be continuously evaluated by reviewing and assessing a comprehensive body of pathological lesions of the peripheral nervous and muscular systems using current and archived neuromuscular cases.

Moreover, a summative assessment at or near the end of the course will be performed against some pre-set, possibly external standard, in order to evaluate the residents' learning outcomes.