

09 - Motor neurone disease (MND) type

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© SPMM Course 3. Frontotemporal Dementia (FTD)

FTD is associated with three types of underlying pathology: Frontal lobe degeneration type □ Most common type □ Spongiform degeneration or microvacuolation of the superficial neuropil is seen chiefly in layers III and V of the cortex. □ Loss of large cortical nerve cells with minimal gliosis
Pick's type □ Pick's disease is characterized by a preponderance of atrophy in the frontotemporal regions. □ These regions also have a loss of large cortical nerve cells, abundant gliosis, and neuronal Pick's bodies, which are masses of cytoskeletal elements. □ Abnormal swollen oval-shaped neuronal cells with loss of Nissl's substance and peripherally displaced nucleus are called Pick cells □ Pick's bodies are seen in some postmortem specimens but are not necessary for the diagnosis. These are argentophilic, tau and ubiquitin reactive filamentous inclusions. □ Hirano bodies may also be seen albeit with a lesser frequency than in Alzheimer's. Motor neurone disease (MND) type □ Cerebral atrophy is less marked; limbic areas are largely preserved □ Loss of large cortical nerve cells, microvacuolation, and mild gliosis. □ Ubiquitinated but not tau-immunoreactive inclusions are present within the frontal cortex and hippocampus □ MND pathology is also seen in anterior horn cells.

HUNTINGTON'S DEMENTIA

Pathologically there is severe loss of small neurons in the caudate and putamen with subsequent astrogliosis.

Characteristic protein deposits form nuclear inclusions in neurons of HD patients.

With the loss of cells, the head of the caudate becomes shrunken and there is "ex vacuo" dilatation of the anterior horns of the lateral ventricles.