TOPIC: Degenerative CNS Diseases

MAIN CONCEPT: Correlation of pathology of degenerative diseases of the central nervous system with pathogenesis and clinical consequences

Review the following patient history and slides with your laboratory instructor. Use the clinical and pathologic information that you are given along with pertinent lecture notes and text sections from Robbins Basic Pathology, 8th Ed., Chap 23 pp. 891-897 to answer the questions listed below.

Case #1: Slides # 391 & 460
The chief financial officer of a small manufacturing company with a past medical history of hypertension controlled with amlodipine begins having difficulty remembering recent events at age 69. His social habits include regular exercise and 2-3 ounces of alcohol daily. At age 72, the patient is brought to his physician by the daughter because of increasing confusion and forgetfulness. The patient has neglected to pay his usual bills and he became lost while returning from the neighborhood grocery store several times over the past year, which has prompted concern for his safety. The daughter states that her father seems depressed and is increasingly uncommunicative. Neurologic exam reveals a confused patient, oriented only to person. He can recall three items immediately after naming them, but can not remember them five minutes later. He can name a pencil and a watch, but is unable to spell world backwards. Neurologic exam shows no focal abnormalities. Lab studies: serum calcium, vitamin B12, thyroid hormone levels, syphilis serology and CSF exam are all normal; apolipoprotein genotyping shows one Apo _4 allele. A CT scan shows generalized cerebral cortical atrophy and hydrocephalus ex vacuo.

Questions & discussion:

1. For the case, identify the major clinical problem, develop a differential diagnosis, describe the likely etiology, any associated risk factors, and the pathogenesis of the disorder. Based on your examination of slides # 391 & 460, what is the diagnosis?

2. Describe the pathologic features of the condition and explain how the lesions are related to the clinical features of the disorder.

   For Slides #391 & 460: how are the pathologic features different from normal brain aging? What are paired helical filaments? What is the source of the beta amyloid in the center of neuritic plaques and CNS blood vessel walls?

3. What are the laboratory and other tests, if any, used in the diagnosis of degenerative CNS disorders? How is the disorder treated?
4. What are other major CNS degenerative disorders and what pathologic lesions and clinical features do they produce?

Learning Objectives

1. Be familiar with the common degenerative diseases of the central nervous system. Understand the epidemiology, etiology, pathogenesis and pathophysiology of these disorders.

2. Be able to describe and recognize the pathologic lesions and clinical features of major CNS degenerative diseases.