Intracranial hypotension clinical features

The syndrome of spontaneous intracranial hypotension is characterized by the following in the absence of antecedent trauma or dural puncture:

1. low CSF pressure (generally < 6 cm H2O)

2. typically associated with orthostatic headache: dramatically worse when upright, improved in recumbency

3. diffuse pachymeningeal enhancement (cerebral and/or spinal) on MRI

For SIH, most patients have orthostatic headaches with sudden onset, but other headaches have been described such as thunderclap, non-positional, exertional headaches, headaches at the end of the day, and even paradoxical headaches with worsening upon lying.

Atypical patients have been described without headache, without pachymeningeal enhancement on MRI, with clinical signs of encephalopathy, cervical myelopathy or parkinsonism.

Since some patients may have normal intracranial pressure, the term “CSF hypovolemia” has been suggested.

The typical clinical manifestation - orthostatic headache - may be masqueraded by atypical clinical findings, including coma, frontotemporal dementia, leptomeningeal hemosiderosis-associated symptoms, and others.

There are three hypotheses to explain the occurrence of headache associated with low CSF fluid. The first is traction on pain-sensitive intracranial and meningeal structures; the second is CSF hypovolemia, and the third is the spinal loss of CSF resulting in increased compliance at the caudal end of the CSF space.

Other symptoms can include alterations in hearing, nausea, vomiting, neck stiffness, diplopia, visual field cuts and difficulty with concentration.

Cases with a rapid neurologic deterioration resulting in an altered mental status have been reported.