症例報告

# 家族性正常圧水頭症が疑われた一家系

稲山 靖弘[1]　渡辺 浩年[1]

## A case of familial normal pressure hydrocephalus

Yasuhiro　Inayama　Hirotoshi Watanabe[1]

「抄録」40歳後半に水頭症を発症し、発端者の父、姉、伯母も同様の症状がみられ、家族性正常圧水頭症と考えられた症例を経験したので報告する。症例：50才後半男性、家族歴、父が60歳時正常圧水頭症にて脳室ー腹腔（VP）シャント術を受けた。父方の伯母が50歳時水頭症発症したが、シャント術受けずにその後死亡。発端者の姉が50歳時正常圧水頭症を発症し腰椎ー腹腔（LP）シャント術を受けるも認知症が進行し50歳後半に死亡。患者はX-10年、緩徐に進行する歩行時のふらつき、失禁、易興奮の出現により、脳外科受診し水頭症を指摘されLPシャント術を受けた。シャント術後、歩行障害、失禁は消失するも、X年，記憶力低下、失禁、意欲低下、歩行時ふらつきが再度出現し当院外来受診となった。改訂長谷川式簡易知能評価スケール（HDS-R）は26/30、Time up and go test（TUG）テストは10秒。指鼻テストは正常、筋強縮もなかった。頭部MRI検査では、著明な脳室拡大、高位円蓋部脳溝は狭小、$^{123}$I-IMP脳血流シンチ（SPECT）検査では、両側頭頂葉、後部帯状回に血流低下を認めた。受診後、一ヶ月間でHDS-Rが4点低下し、尿失禁の回数も増加し、歩行時のふらつきも悪化したため、再度脳外科へ紹介した。結語：家族性正常圧水頭症の報告は極めて少なく、本邦では2家系目である。常染色体優性遺伝形式で発症したと考えられた症例を報告した。

「**Abstract**」We report a case of a male patient who presented a familial normal pressure hydrocephalus (NPH) in his late 40's, and similar symptoms were seen in his father, his older sister and his aunt. **Case:** A male patient in his late 50's was herein presented. The family history showed that his father received ventriculo-peritoneal (VP) shunt because of normal pressure hydrocephalus in 60 years. The aunt of his father died without shunt in 50 years. His elder sister in the 50 years had normal pressure hydrocephalus and took an lumbo- peritoneal (LP) shunt, but she developed dementia and died later. The patient developed an incontinence, a gait disturbance and easy excitability in a X-10 year. Then he was diagnosed a hydrocephalus and had an LP shunt operation. After the treatment, the gait disturbance and the incontinence disappeared. In the first visiting our hospital, he showed memory disturbance, incontinence, gait disturbance. The revised Hasegawa's dementia rating scale (HDS-R) was 26/ 30. Time up and go test was 10 sec. Finger-nose test and the muscle rigidity in his upper extremities were normal. Brain MRI showed an enlargement of the ventricles with a narrowing of the subarachnoid space and cortical sulci at the high convexity of the cerebrum. $^{123}$I-IMP cerebral blood flow scintigraphy (SPECT) showed a hypoperfusion in the bilateral parietal lobe and the posterior cingulate gyrus. He was admitted to the department of neurosurgery again because of rapid deterioration in HDS-R, his incontinence and gait disturbance in a month. **Conclusions:** There are extremely few reports of the familial normal pressure hydrocephalus. We reported the second case of NPH to be inherited in an autosomal dominant fashion.

**Key Words**：家族性，正常圧水頭症，iNPH，シャント術

1］聖志会　渡辺病院　Medical corporation Seishkai , Watanabe Hosipital