

Introduction

To evaluate the incidence of recurrent intracranial bleeding and mortality in patients with hemorrhagic Moyamoya disease(MMD), and to identify risk factors and the time course of these events.

Methods

A total of 128 patients with complete follow-up treated conservatively were included.The occurrence of recurrent stroke over long-term follow-up was documented. Annual and cumulative incidence rate of rebleeding was generated via Kaplan–Meier survival analysis, and risk factors for rebleeding were analyzed through Cox proportional hazards regression models.

Results

The median follow-up time was 10.1 (range:1–27) years. During 1300.7 patient-years,47(36.7%) patients experienced fifty-nine episodes of rebleeding, rendering an average annual incidence of 4.5%. Among them, 9 patients (19.1%) died from rebleeding and 12 patients had serious disability (modified Ranking Scale=3 ). The cumulative risk of rebleeding was 7.8% at 5 years, 22.6% at 10 years, and 35.9% at 15 years. Only 4 (3.1%) patients experienced ischemic stroke, yield an average annual incidence of 0.3%. Decreased of regional cerebral blood flow (rCBF) (HR, 2.23; P=0.03),smoking and drinking history (HR, 2.43; P=0.04) were independent predictors of rebleeding. Rebleeding (HR,10.76; P=0.02) and hypertension (HR,5.29; P=0.02) were associated with increased mortality. Age, types of first bleeding, DSA stage, family history, and coexistent with cerebral aneurysms were not associated with any increased risk of rebleeding .

Conclusions

Rebleeding events were common in patients with hemorrhagic MMD and the risk of rebleeding increased within a very long-term follow-up. Rebleeding was strongly associated with increased mortality. Decreased of rCBF, smoking and drinking history seem to be risk factors for rebleedin

Learning Objectives

To evaluate the natural course of hemorrhagic MMD in China.

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