Intracranial hemorrhage from cerebral cavernous malformation

Cavernous malformations have an annual bleeding incidence of 0.2-2.3% [1) 2)] Most hemorrhages attributable to cavernous malformations are characterized by microhemorrhages and are seldom catastrophic or fatal. Most notably, supratentorial cavernous malformations leading to massive, life-threatening hemorrhages are rare entities [3)]

Gross et al reviewed hospital databases to identify children with CMs who had not been treated surgically and who had clinical and radiological follow-up. Annual Intracranial hemorrhage from cerebral cavernous malformation rates were calculated in lesion-years, and risk factors were assessed using the Cox regression.

In a cohort of 167 patients with 222 CMs, the mean patient age at the time of diagnosis was 10.1 years old (SD 6.0). Ninety patients (54%) were male. One hundred four patients (62%) presented with hemorrhage from at least 1 CM, 58 (35%) with seizures with or without CM hemorrhage, and 43 (26%) with incidental lesions. Twenty-five patients (15%) had multiple CMs, 17 (10%) had a family history of CMs, and 33 (20%) had radiologically apparent developmental venous anomaly (DVAs). The overall annual hemorrhage rate was 3.3%. Permanent neurological morbidity was 29% per hemorrhage, increasing to 45% for brainstem, thalamic, or basal ganglia CM and decreasing to 15% for supratentorial lobe or cerebellar lesions. The annual hemorrhage rate for incidental CMs was 0.5%; for hemorrhagic CMs, it was 11.3%, increasing to 18.2% within the first 3 years. Hemorrhage clustering within 3 years was statistically significant (HR 6.1, 95% CI 1.72-21.7, p = 0.005). On multivariate analysis, hemorrhagic presentation (HR 4.63, 95% CI 1.53-14.1, p = 0.007), brainstem location (HR 4.42, 95% CI 1.57-12.4, p = 0.005), and an associated radiologically apparent DVA (HR 2.91, 95% CI 1.04-8.09, p = 0.04) emerged as significant risk factors for hemorrhage, whereas age, sex, CM multiplicity, and CM family history did not.

The 5-year annual and cumulative symptomatic hemorrhagic risk in our pediatric FCCM cohort equals the overall risk described in children and adults with all types of CCM. Imaging features at first brain MRI may help to predict potential symptomatic hemorrhage at 5-year follow-up [4)].

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