Multiple Cerebral Aneurysms and Subarachnoid Hemorrhage in a Patient with Alagille Syndrome

Hans-Georg Schlosser, Thoralf Kerner, Christian Woiwiekowsky and Goetz Benndorf

Summary: Although intracranial hemorrhage has frequently been found responsible for mortality in adult patients with Alagille syndrome (AGS), no specific underlying cause has been identified. We describe the case of severe subarachnoid hemorrhage in a 30-year-old woman harboring five intracranial aneurysms and multiple peripheral vascular anomalies. To evaluate a possible higher incidence of intracranial aneurysms, a study of the cerebral vasculature in all AGS patients by using noninvasive imaging techniques should be considered.

Case Report

A 30-year-old woman with AGS was found comatose after 3 days of severe headache. At the emergency department, she presented with a Glasgow coma scale of 6 (Hunt-Hess grade 5), and CT showed a subarachnoid hemorrhage (SAH; Fisher grade 4) and a right parietal subdural hematoma causing a midline shift of 0.5 cm.

Cerebral angiography depicted a ruptured 8-mm aneurysm of the right suprachoroidal internal carotid artery (Fig 1A). In addition, a small right extradural internal carotid artery aneurysm, two left internal carotid artery aneurysms (Fig 1B), and a basilar tip aneurysm (Fig 1C) were depicted. Furthermore, the abdominal angiogram showed aneurysms of the right renal artery, the celiac (trunk, the splenic artery, and a coarctation of the thoracic aorta (Fig 1D–F) without related clinical symptoms.

After surgical clipping of the right internal carotid artery, pathohistologic analysis of the arterial wall showed the typical structure of an aneurysm with an internal elastic lamina reduced to fragments and hypertrophied margins. The postoperative course was uneventful, and after rehabilitation the patient returned to normal life.

Received October 23, 2003; accepted after revision December 24.
From the Departments of Neurosurgery (H.-G.S., C.W.) and Anesthesiology and Critical Care Medicine (T.K.), Charité, Virchow-Clinic, Humboldt-University of Berlin, Berlin, Germany; and the Department of Radiology, Baylor College of Medicine, the Methodist Hospital (G.B.), Houston, TX.
Address correspondence to Goetz Benndorf, MD, PhD, Department of Radiology, Baylor College of Medicine, One Baylor Plaza, Houston, TX 77030.

© American Society of Neuroradiology
Whether the relatively high mortality of patients with AGS resulting from ICH is possibly related to previously unidentified SAHs caused by ruptured intracranial aneurysms remains questionable and can be answered only by more supporting data. For evaluation of a possible increased incidence of cerebral aneurysms in patients with AGS compared with that of the normal population, CT or MR angiography studies should be considered. The concurrence of multiple intracranial aneurysms and multiple extracranial vascular malformations in AGS has not yet been reported. Multiple peripheral vascular abnormalities, as found in our patient, are known and in agreement with other reports (9–11). Berard (10) found in his series a total obstruction or stenosis of one or both renal arteries in four patients that caused renovascular hypertension and a stenosis of the celiac trunk in one patient; however, an aneurysm of the splenic artery has not yet been described. Similarly, aortic coarctations have been reported in AGS (9, 10) and can be responsible for hypertension, intermittent claudication, or abdominal pain.

**Conclusion**

We describe a case of AGS in a patient with severe SAH in whom multiple and various cerebral and peripheral vascular lesion were detected. This case helps expand the existing knowledge of vascular abnormalities in AGS, emphasizing the predisposition toward vasculopathy in this genetic disorder. The limited data on the histologic nature of these vascular lesions in the literature, as well as the clinical importance of a ruptured berry aneurysm potentially causing fatal SAH, prompt more detailed evaluation of the cerebral vasculature in these patients by use of noninvasive imaging techniques such as CT or MR angiography.

**References**