Multiple Varices in the Unilateral Cerebral Venous System

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Case Report

Multiple Varices in the Unilateral Cerebral Venous System

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Summary: A case of multiple cerebral varices located in the superficial cerebral veins and ipsilateral internal jugular vein is reported.

Although a cerebral varix may frequently be associated with an arteriovenous malformation, a solitary intracranial varix is rare in the CNS (1–3). Several previous reports have described a cerebral varix composed of a single superficial or deep cerebral vein without other accompanying vascular malformations (4–7). We describe a case of unilateral varicosities in the superficial cerebral veins and ipsilateral neck.

Case Report

A 24-year-old woman was referred because of a palpable mass in the right side of the neck. Her history was not remarkable for childhood infection, congenital abnormality, or familial vascular disease. Physical examination revealed normal height and weight for age. The extremities were symmetrical, and there were no cutaneous lesions. Findings at neurologic examination were normal, and results of tests for bleeding time, prothrombin time, activated prothrombin time, fibrinogen, and fibrin/fibrinogen degradation products were all within normal limits. A contrast-enhanced cervical CT scan showed an enlarged right internal jugular vein as well as an opacified mass adjacent to the internal and external jugular vein (Fig 1A). There was no stenosis in the superior vena cava. A right internal carotid angiogram showed multiple varices and phlebectasis of the superficial cerebral veins (Fig 1B and C). The cervical varix was opacified via a right enlarged internal jugular vein as well as an external jugular vein (Fig 1D). A left internal carotid angiogram revealed no abnormality. The venous drainage was otherwise normal, including the superior sagittal sinus, lateral sinus, and sigmoid sinus. There was no arteriovenous shunting in the cerebral or cervical vascular system.

Surgical resection of the cervical varix was performed. Histologically, the specimen was composed of a saclike structure measuring about $8 \times 7 \times 7$ mm. Microscopic sections showed partial thickening of the intima. The media and adventitia were degenerated and contained atrophic smooth muscle and dense collagen fibers, consistent with the diagnosis of varix.

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Discussion

McCormick (2) categorized pure intracranial varix as a venous malformation. In a search of the literature, we found only eight cases of angiographically proved cerebral varix that were not associated with arteriovenous shunting (4–11). Cavernous or venous angioma was associated in three of eight cases (9–11); the other five cases were reported to have a varix composed of a solitary cerebral vein without other accompanying vascular abnormalities. Roda et al (5) reported an intraventricular varix that caused intraventricular and subarachnoid hemorrhage. Nishioka et al (6) described a varix of the insular vein that may have caused seizures. Shibata et al (7) reported a case of asymptomatic varix of the deep sylvian vein. To our knowledge, no previous report has described multiple varicose venous dilatations that involved unilateral superficial cerebral veins and that were accompanied by a varix of the internal jugular vein.

Cerebral varix consists of a relatively large, thin-walled vessel, usually lined by a single layer of endothelium and encircled by a relatively thin lamina of fibrous connective tissue (12). Congenital weakness of the vessel walls, previous inflammation, and trauma have been postulated as a cause of varix (12). In our case, the origin of the cervical varix remains unknown; it was surgically resected for cosmetic reasons. Although histopathologic analysis was not revealing, some connective tissue disorder may underlie the pathogenesis. The multiple varicose dilatations occurring in the ipsilateral head and neck might have been caused by a previous inflammation or by maldevelopment of the right cerebral venous systems. Klippel-Trenaunay-Weber syndrome is a phakomatosis accompanied by varicosities and phlebectasis of the superficial and deep venous systems as well as by cutaneous angioma and unilateral hypertrophic limbs (13, 14). However, the findings in our patient were not compatible with such a congenital abnormality.

Cerebral varix may cause intracerebral or intraventricular hemorrhage (5, 8). The prevalence of rupture is not known. Two of the eight reported cases that were diagnosed at angiography had a ce-

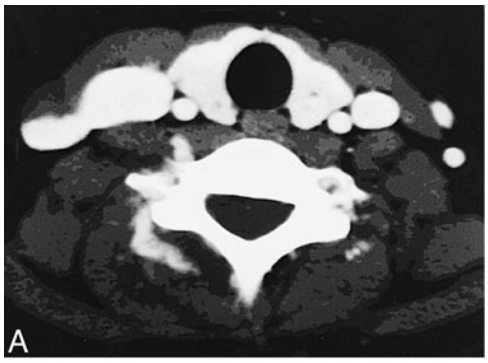
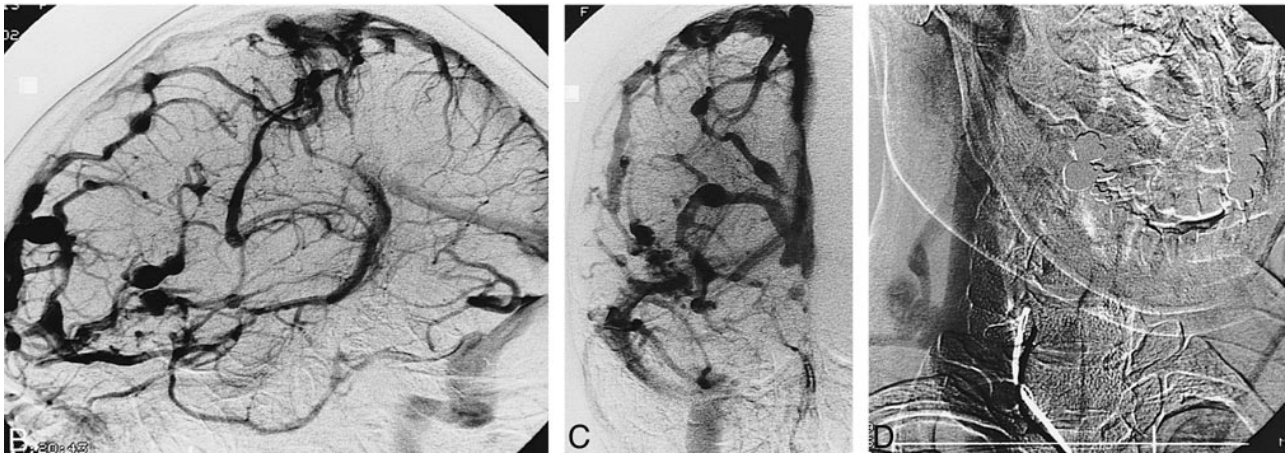


FIG 1. 24-year-old woman with a palpable mass in the right side of neck.

A, Contrast-enhanced cervical CT scan shows a varix adjacent to the right internal and external jugular vein. Asymmetry was evident in the internal jugular veins.

B–D, Right internal carotid angiogram shows multiple varices and enlargement in the superficial cerebral veins (*B, C*). Venous phase also revealed a cervical varix, which was opacified via internal and external jugular veins (*D*).



rebral varix resulting in intracranial hemorrhage (4–11). However, it is likely that, in most cases, the cerebral varix remains clinically silent. Thus, conservative follow-up should be recommended for incidental cases of cerebral varix. In particular, it is impossible to surgically excise the multiple intracranial varicose dilatations, as found in the present case. Although a cerebral varix is an apparently rare disease with neck tumors (15), further radiologic investigation may be warranted, particularly in the intracranial venous system.

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