Editorial

Dissections

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Dissections of cervical and cerebral arteries, either spontaneous or associated with trivial neck torsion, are not rare. Most seasoned neurologists have had experience with a case or two. Dissections are being increasingly recognized and reported in recent years, in part due to the widespread availability of high-quality angiography and in part due to the increasing recognition of the characteristic clinical and radiographic features. Early case reports emphasized major stroke as a consequence of dissection, but recent experience suggests that the outcome is relatively benign in most patients. While cervical carotid dissections have been more frequently reported in the medical literature, it is our anecdotal impression that vertebral artery dissections are at least as common.

The prevalence of dissection of the arteries supplying the brain is uncertain. While dissections underlie less than 1% of unselected brain ischemic events, they may account for 5% or more of ischemic stroke in young adults. One to three cases per year are reported from large, academic-affiliated hospitals. Several hundred cases have been reported in the medical literature in the past decade. Dissections afflict all age groups, but particularly young adults. The mean age is between 25 and 45 years depending upon the artery involved.

Dissection occurs when blood extrudes into the arterial wall, often narrowing the true lumen and sometimes causing focal or diffuse dilatation. "Dissecting aneurysm" is sometimes indiscriminately applied to this phenomenon, but has little descriptive value. Subintimal dissections often do not cause arterial dilatation. When arterial dilatation does occur, pseudoaneurysm is the more accurate term. In most cases, it is unclear whether a primary intimal tear allows dissection of blood from the lumen into the arterial wall, or whether a primary intramural hematoma secondarily ruptures into the true lumen. In a minority of instances, no communication between the dissection cavity and the true lumen can be demonstrated pathologically.

Dissections usually occur in otherwise healthy people. The potential relationship of arterial dissections to strenuous activity or everyday, trivial neck rotation/torsion is difficult to quantitatively define, but is similarly difficult to ignore. Such casual potential trauma as violent coughing, trampoline exercises, neck turning while leading a parade, playing basketball, "head-banging" during dancing, and minor falls have immediately antedated the initial symptoms of dissection.

Cervical rotation and extension can compress the carotid and vertebral arteries against bony structures, possibly precipitating dissection in predisposed persons. In many other patients with dissections, no unusual activity is recognized to precede the initial symptoms. However, the onset of the dissection may precede the initial symptoms by several hours or days, further clouding the relationship between dissection and the presence or absence of trivial neck injury or torsion. The distinction that is made between "spontaneous" dissections and those associated with minor trauma appears to be arbitrary in literature reports. Caplan et al. in this issue report a "spontaneous" vertebral artery dissection which apparently followed a primary respiratory arrest and intubation (with the potential for unusual neck torsion), while others have categorized vertebral dissections related to backing-up an automobile or to swinging a baseball bat as "traumatic." The pathogenetic association of both cervical carotid and vertebral dissections with neck torsion that is inconsequential in most people remains a complex and ill-defined issue.

While the diagnosis of dissection is largely dependent on the characteristic angiographic or pathologic features, dissections cause recognizable clinical syndromes in many afflicted patients. The clinical features, epidemiology, pathology, and perhaps pathogenesis of dissection differ depending on the artery involved.

Cervical carotid dissections usually cause cerebral ischemic symptoms associated with prominent pain in the ipsilateral neck, face or head. A partial Horner's syndrome (oculomotoric paresis) is a frequent finding, and, when associated with undue pain, is often the initial clue suggesting dissection. Pulsatile tinnitus is sometimes present. The mean age of onset is 45 years. Cervical carotid dissection usually involves the distal portion of carotid artery in the neck, terminating at the entrance of the artery into the petrous bone. Angiography reveals a string-sign or tapered occlusion beginning distal to the cervical bifurcation, a scalloped stenosis, a pseudoaneurysm, or a double-
4.1 Intramedial hematoma causing pseudoaneurysm is the rule. Underlying fibromuscular dysplasia is reported in about 20% of cases. At present, it is not possible to determine the relative merits of the various therapies for cervical carotid dissection. Clinical reports of consecutive patients show the outcome to be surprisingly benign. At least 80% of patients fully recover or experience only minor sequelae. Brain ischemic symptoms often evolve over several days. We currently anticoagulate these patients for approximately four months while healing occurs. We base this treatment on the hypothesis that distal embolism of thrombotic fragments at the site of dissection frequently causes or contributes to brain ischemia.

Intracranial carotid dissections and those involving the middle cerebral artery have distinct clinical features warranting separation from their extracranial counterpart. The age profile is distinctly younger (mean age of 25 years), the site of dissection is usually subintimal, and massive stroke with death appears to be a more frequent outcome. However, because the angiographic features of intracranial dissections are less specific, autopsied patients dominate the literature. Milder cases may go unreported or unrecognized. Intracranial dissections have been associated with intense physical exertion in many patients. Severe headache and major stroke are the usual presenting manifestations. Pathologic examination often does not demonstrate morphologic abnormalities underlying these dissections, although fibromuscular dysplasia and fibroelastic intimal abnormalities have been reported. Subarachnoid hemorrhage can complicate these dissections. Optimal management of intracranial dissections is uncertain.

Cervical vertebral dissections usually present with prominent headache, typically unilateral and localized to the upper cervical region, with features of the lateral medullary syndrome. Vertebral artery dissection is a common cause of the lateral medullary syndrome occurring in young adults, especially if associated with unusual neck torsion (e.g., minor falls, cervical chiropractic manipulation). Of all sites of dissection, vertebral artery dissections are most consistently associated with unusual, though often minor, neck torsion. Symptoms of brain ischemia commonly evolve over several hours to a few days. The mean age
is about 40 years. Angiography demonstrates scalloped, irregular narrowing with or without a pseudoaneurysm and a double-lumen. These abnormalities are usually maximal in the distal vertebral artery, but occasionally they involve the entire length of the vessel and extend into the basilar artery (Figure 1). Bilateral, simultaneous vertebral dissections are often associated with fibromuscular dysplasia. The outcome is benign in most patients, reflecting the adequacy of the collateral circulation in these relatively young patients. Dissections which extend intradurally can result in subarachnoid hemorrhage. Dissection in a recent patient (Figure 1) extended to completely occlude the proximal basilar artery. Nevertheless, the patient recovered uneventfully. Management recommendations are largely based on clinical anecdotes and remain empiric. We currently anticoagulate these patients if examination of the spinal fluid shows no evidence of subarachnoid hemorrhage.

The pathogenesis of dissection in most patients remains an enigma. Fortunately, only a small fraction of patients come to portmortem examination. The potential role of minor neck torsion in precipitating dissection does not explain why most people tolerate such trivial trauma without arterial injury. Yet fibromuscular dysplasia and cystic medial degeneration are recognized to underlie only a minority of dissections. Striking cases of multiple arterial dissections associated with fibromuscular dysplasia have been published. The pathogenesis may well be multifactorial. In cervical vertebral dissections, the association with unusual, if trivial, neck torsion is particularly prominent. Any hypothesis for the cause of dissection must account for the tendency to occur in middle adult years, an epidemiologic feature shared with saccular aneurysms. It seems reasonable to speculate that as yet ill-defined, focal morphologic abnormalities may often be the culprits. These abnormalities could be formefrustes of fibromuscular dysplasia in the extracranial arteries or of fibroelastic intimal abnormalities in intracranial dissections. Additional, very thorough, pathologic studies of a larger number of patients will be needed to clarify the pathogenesis of dissections.

References

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