Superior Sagittal Sinus Thrombosis
10 Years After Surgery For Ulcerative Colitis

MARK S. YERBY, M.D. AND GEORGE M. BAILEY, M.D.

SUMMARY Cerebral vein thrombosis is a known complication of active ulcerative colitis. It is generally believed that panproctocolectomy ameliorates the thromboembolic and other systemic complications of ulcerative colitis. We report an unusual patient, 10 years post-panproctocolectomy for ulcerative colitis, who developed a cerebral sinus thrombosis. Physicians should be aware of the possibility of thromboembolic complications of ulcerative colitis long after definitive surgery has been performed.

ONE OF THE MORE SERIOUS COMPLICATIONS of active ulcerative colitis is thromboembolic phenomena. Thrombotic complications generally occur within the deep veins of the lower extremities and some of the larger veins in the abdomen. Recently Mayeux and Fahn reviewed the subject and described seven patients with cerebral vein thrombosis in association with exacerbation of their disease. It is generally believed that thromboembolism and most other systemic complications of ulcerative colitis, with the possible exceptions of pyoderma gangrenosum and liver disease, can be ameliorated by panproctocolectomy. The purpose of this paper is to report a young man, ten years post-panproctocolectomy for ulcerative colitis, who developed cerebral vein thrombosis and to alert physicians to the possibility of thromboembolic complications in ulcerative colitis long after definitive bowel surgery has been performed.

Report

A 28-year-old white man was hospitalized because of status epilepticus. Four days previously, he noted the onset of a bilateral throbbing fronto-temporal headache which was continuous and of increasing intensity. He continued his daily activities and denied he had visual changes or weakness. On the day of admission, he had one episode of vomiting and began having focal motor seizures which started in the left face and became generalized. There was no prior history of head trauma, loss of consciousness or headaches of this type, and he was not taking medication.

At the age of nine he developed ulcerative colitis which was documented by sigmoidoscopy, barium enema and biopsy specimens. At age 18, he suffered a right ileofemoral thrombosis. A diagnosis by karyotype of Kleinfelter’s syndrome was made at this time. Three months later a right renal vein thrombosis was documented and anticoagulation therapy with coumadin began. After four months of this treatment, a panproctocolectomy was performed (1968). Venograms in July 1970 showed patency of the right deep femoral system indicating resolution of the previous venous obstruction. Fourteen months later extensive bilateral venous thrombosis involving the distal, popliteal and femoral veins occurred. Coumadin was again given but was subsequently discontinued because of poor compliance. He remained asymptomatic until the present admission. All clotting studies in past admissions were normal, as were liver function tests. There was no family history of clotting disorders.

On admission, physical examination revealed a well hydrated, tall, obese male with an ileostomy in the right lower quadrant of his abdomen. The lower extremities were edematous with evidence of chronic stasis dermatitis below the knee. There was no papilledema, fever, or nuchal rigidity and the remainder of the general physical exam was normal. The patient was having almost continuous, generalized convulsions. During the interictal periods, the neurological exam was within normal limits. Seizures were treated with phenytoin and phenobarbital.

Laboratory studies on admission revealed a platelet count of 114,000. All other laboratory tests, including erythrocyte sedimentation rate, pro-thrombin time, partial thromboplastin time, fibrinogen, plasma protamine para-coagulation, ethanol gel, anti-thrombin III and platelet aggregating factors were normal. Isotope brain scan showed increased isotope uptake over the frontal-temporal region. CT scan was interpreted as normal. Cerebral angiography in the venous phase revealed non-filling of the superior sagittal sinus from its most anterior extent posteriorly to the coronal sutures. Oblique views (figure) revealed thrombosis of the anterior portion of the superior sagittal sinus. Exploratory bowel surgery revealed no evidence of bowel disease. The patient responded well to anticonvulsant therapy and was discharged home asymptomatic.

Discussion

The occurrence of cerebral venous thrombosis in association with ulcerative colitis was first reported by Graef et al. and Krayenbuhl in 1966. Subsequent reports by Harrison and Truelove in 1967, Borda et al. in 1973, and Mayeux and Fahn in 1978, have confirmed the increased risk of cerebral venous thrombosis among patients with ulcerative colitis. The subject has been recently reviewed by Mayeux and Fahn, who also reported 3 patients in whom a diagnosis of cerebral venous thrombosis appeared likely on clinical grounds although arteriography was not done.
Of the 7 patients with cerebral venous thrombosis in association with ulcerative colitis so far reported, a majority of them were suffering from an exacerbation of their disease when the thrombosis occurred. To our knowledge the only instances of cerebral venous thrombosis after successful panproctocolectomy have occurred during the immediate post-operative period. In our patient, sagittal sinus thrombosis occurred 10 years after surgery. Although there is no direct evidence to link the 2 problems in this patient, the history of previous venous thrombosis during exacerbation of his disease, and the absence of any other obvious etiological factor, suggest a more than coincidental association between the sagittal sinus thrombosis and the history of ulcerative colitis. A search of the literature revealed no association between venous thrombosis and Klinefelter's syndrome.

Venous thrombosis is known to occur with greater than random frequency in patients with ulcerative colitis. The incidence varies from 1.2% to 7.5% diagnosed clinically, to 39% diagnosed at autopsy. Several explanations for this phenomenon have been proposed: thrombocytosis, increase in thromboplastin generation, increase in coagulation factors III, V, VIII, increase in fibrinogen, decrease in antithrombin III, and corticosteroid therapy and its inhibition of reactive fibrinolysis. None of these explanations is entirely satisfying, and an explanation for the increased incidence of thrombosis in ulcerative colitis remains elusive. Abnormalities in coagulation are not consistently seen in patients with this disease. There is also no certainty that coagulation factors in themselves are thrombogenic. For example, high levels of several coagulation factors occur in hyperthyroidism in which there is no association with thromboembolic disease. Truelove and Witts, in a controlled trial in 1955, showed that corticosteroids do not increase risk of thrombosis.

In addition to the questions surrounding the etiology of venous thrombosis in ulcerative colitis, the question of the appropriate therapy has been raised. A recent report by Gettlefinger and Kokmen suggests that anticoagulants should not be used in patients with sagittal sinus thrombosis. This would seriously limit the ability to intervene in patients with diagnosed or suspected cerebral venous thrombosis. Since the proposed defects in coagulation in ulcerative colitis permit more rapid progression of the early phase of coagulation, perhaps these patients could be more safely managed prophylactically with antiplatelet agents, i.e., aspirin or dipyridamole, which are effective in reducing early phases of coagulation by limiting platelet aggregation.

We suggest that the clinician maintain a high index of suspicion for thromboembolic phenomena, including cerebral venous thrombosis, in patients with a history of ulcerative colitis and definitive bowel surgery, as well as those with active disease. Those with cerebral venous thrombosis may have an acute illness

<table>
<thead>
<tr>
<th>Year reported</th>
<th>Authors</th>
<th>No. of patients</th>
<th>Age</th>
<th>Sex</th>
<th>Confirmation of diagnosis</th>
<th>Status of colitis</th>
</tr>
</thead>
<tbody>
<tr>
<td>1966</td>
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<td>40's</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
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<tr>
<td>1967</td>
<td>Harrison &amp; Truelove</td>
<td>34</td>
<td>M</td>
<td>autopsy confirmed at autopsy</td>
<td>exacerbation</td>
<td></td>
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<tr>
<td>1973</td>
<td>Borda et al</td>
<td>1</td>
<td>23</td>
<td>M</td>
<td>angiography confirmed at autopsy</td>
<td>exacerbation</td>
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<tr>
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<td>Rosseau</td>
<td>1</td>
<td>18</td>
<td>M</td>
<td></td>
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<td>3</td>
<td>12</td>
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<td>28</td>
<td>M</td>
<td>angiography</td>
<td>cured by surgery</td>
</tr>
</tbody>
</table>

**TABLE. Cerebral Venous Thrombosis in Patients with Ulcerative Colitis**
with fever, elevated sedimentation rate, headache, papilledema, seizures and focal neurologic signs. The diagnosis is confirmed by cerebral arteriography or radionuclide scanning obtaining oblique views.24

While most patients with cerebral venous thrombosis are left with major neurological deficits, some recover with little or no residual. These latter patients may benefit from prophylactic antplatelet therapy.

References
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