Management of intracerebral hemorrhage in idiopathic thrombocytopenic purpura

Report of four cases

ROBIN P. HUMPHREYS, M.D., F.R.C.S. (C), ANTHONY D. HOCKLEY, M.B., F.R.C.S. (EDIN.), MELVIN H. FREEDMAN, M.D., F.R.C.P. (C), AND E. FRED. SAUNDERS, M.D., F.R.C.P. (C)

Divisions of Neurosurgery and Hematology, The Hospital for Sick Children, Toronto, Ontario, Canada

There has been little comment on the specific management of intracerebral bleeding occurring in patients suffering idiopathic thrombocytopenic purpura. The authors present the cases of four children with intracerebral hemorrhage due to this coagulation disturbance. A plan of management is described based on this experience; it includes immediate control of cerebral edema, emergency splenectomy, supportive care with platelet transfusions and corticosteroids, cerebral angiography, and a definitive neurosurgical procedure. If necessary, the radiological investigation and surgical therapy can be performed with a single general anesthetic. Three of the patients have survived without major neurological sequelae.

KEY WORDS idiopathic thrombocytopenic purpura platelets intracerebral hemorrhage coagulation disturbance

Case Reports

Case 1

This boy developed ITP at 8 years of age. No therapy was given. Following a fall 3 months after the onset of ITP, he developed headache, drowsiness, and vomiting over a 24-hour period.

Examination. The child was deeply comatose with a fixed, dilated right pupil and left hemiplegia. The platelet count was 5000/cu
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mm and there were adequate megakaryocytes in the bone marrow. A diagnosis of intracranial hemorrhage was made. Intravenous dexamethasone and mannitol (20%) brought about improvement in the level of consciousness.

Operation. An emergency splenectomy was performed, followed by transfusion of 12 units of platelet concentrate, resulting in a platelet count of 78,000/cu mm. Under the same anesthetic, a right carotid angiogram revealed an intratemporal space-occupying lesion. A right temporal craniectomy and cortical incision allowed direct evacuation of 40 to 50 ml of dark blood from within the moderately swollen temporal lobe.

Postoperative Course. There was no problem with hemostasis and immediately postoperatively the patient was much improved. Corticosteroid therapy was continued and 5 days after surgery, the platelet concentration was 1,163,000/cu mm. The patient was discharged 18 days after surgery with no therapy. Three weeks later he returned to hospital because of bleeding; the platelet concentration was 1000/cu mm. Prednisone therapy was reinstituted; subsequent platelet levels proved to be steroid-dependent for several months before the drug was finally stopped. The only neurological residuum has been a mild behavioral disorder.

Case 2

This 14-year-old boy was diagnosed as having chronic ITP after a 5-year history of easy bruising and epistaxis. No therapy had been given. Laboratory data included a platelet count of 6000/cu mm with plentiful megakaryocytes in the bone marrow.

First Operation. An elective splenectomy was performed because of bleeding symptoms; it resulted in an increase in the concentration of platelets to 860,000/cu mm by the fifth postoperative day. The boy was discharged 1 month after surgery with no therapy. Three weeks later he returned to hospital because of bleeding; the platelet concentration was 1000/cu mm. Prednisone therapy was reinstituted; subsequent platelet levels proved to be steroid-dependent for several months before the drug was finally stopped. The only neurological residuum has been a mild behavioral disorder.

Case 3

This 13-year-old girl was diagnosed as having ITP after a 6-week history of purpura, bleeding from the gums, epistaxis, and heavy menstrual flow. Laboratory data included a platelet count of 1000/cu mm and adequate megakaryocytes in the bone marrow examination. No therapy was given and she remained well until 2 weeks later, when she suddenly complained of headache and vomiting and had two generalized convulsions.

Examination. The fundi appeared normal and there was no obvious focal neurological deficit. The platelet count was 1000/cu mm. A clinical diagnosis of intracranial hemorrhage was made and treatment started with intravenous dexamethasone. Computerized axial tomography (CAT) (Fig. 1 left) outlined a right frontal abnormality consistent with an intracerebral hematoma. As the patient's neurological status improved dramatically, she was observed expectantly with the hope that she would show spontaneous recovery from her ITP. Ten days later she became lethargic and developed severe headaches and meningismus. Because of technical difficulties, the CAT scanner was not available, but a carotid arteriogram confirmed that the only site of intracranial bleeding remained that within the right frontal lobe.

Operation. Following an emergency splenectomy and administration of 10 units of platelet concentrate, bilateral carotid arteriograms (performed under the same anesthetic) outlined a space-occupying lesion in the left occipital lobe, with evidence of transtentorial herniation.
30 ml of blood removed from a cavity in the medial part of the frontal pole. The surrounding brain was edematous, and the dura was thus left open and the bone flap out. Hemostasis was adequate.

**Postoperative Course.** Postoperatively, the platelet count was 100,000/cu mm. The patient was fully alert with no neurological deficit. Corticosteroids were stopped after 3 weeks and a further CAT scan showed complete removal of the clot (Fig. 1 right). The patient has remained well with normal platelet counts. Her bone flap was subsequently replaced without difficulty.

**Case 4**

This 10-year-old girl presented with a 1-week history of purpura.

**Examination.** The platelet count was 3000/cu mm and the bone marrow aspirate contained many megakaryocytes. A diagnosis of ITP was made and no therapy given. Five days later, she developed headache and vomiting and progressively over 24 hours, drowsiness, meningismus, and a left hemiparesis. Intravenous dexamethasone brought about no improvement.

**Operation.** Following emergency splenectomy, platelet concentrates were given without any increase in the platelet count. Under cover of further platelet concentrates, a right carotid angiogram showed a space-occupying lesion in the region of the right internal capsule. The location of the clot seemed initially to militate against its surgical removal. The next day because of deteriorating neurological status, and in spite of continuing thrombocytopenia, a temporal burr hole was placed. Fifteen ml of old blood was aspirated from the temporal lobe followed by fresh bleeding through the brain needle. Despite further platelet concentrate administration, hemostasis was never achieved and the brain needle was removed.

**Postoperative Course.** Mannitol (20%) was given postoperatively, and the child's neurological status improved markedly. After surgery, prednisone and daily platelet transfusions were given with no improvement in platelet counts, but there was little bleeding problem. The patient was discharged 1 month
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postoperatively, without neurological residuum. She has remained moderately thrombocytopenic, with or without prednisone therapy.

Discussion

About 80% of patients with acute ITP of childhood recover spontaneously in the first few weeks of illness without any therapy. Aside from easy bruising and a petechial rash, most children are completely asymptomatic during that interval and restricted physical activity is the sole form of management. The use of corticosteroid therapy in patients without gross bleeding complications is debatable and does not seem to be necessary in most cases. While steroids do not appear to shorten the course of ITP, their use may lessen bleeding by a direct vascular action or by improving platelet survival by blocking phagocytosis in the reticuloendothelial system.

In our experience, splenectomy for children with acute ITP is reserved for uncontrolled bleeding, since platelet transfusions alone seldom result in an increment in circulating platelet counts or ameliorate hemorrhage, probably because of rapid peripheral destruction of the platelets. However, platelet survival generally increases following splenectomy, thereby permitting transfused platelets to effect hemostasis at that time.

Previous authors have commented on the relative rarity of intracranial bleeding in ITP. In those reported instances of cerebral hemorrhage occurring in ITP, the diagnosis was uncertain, since specific investigative measures designed to delineate the hematoma were not instituted. Hirsch and Dameshek reported three patients with "mild cerebral hemorrhages" but did not define how the diagnosis was made. Lusher and Zuelzer documented two patients whose deaths were thought to be due to intracranial bleeding, but at autopsy there were "cerebral vascular lesions reminiscent of thrombotic thrombocytopenic purpura."

Furthermore, there has been no specific written comment concerning the management of patients with ITP who suffered intracranial hemorrhage. One patient referred to by Kowrower and Watson survived a spontaneous intracranial hemorrhage and was treated with splenectomy but remained blind and hemiplegic. Another patient in Hirsch and Dameshek's review suffered a "severe cerebral hemorrhage" and died from it following splenectomy. As intracranial bleeding is the most dreaded complication of ITP, we thought it valuable to examine our experience and outline a treatment plan that confirms the diagnosis and permits surgical evacuation of the intracerebral hematoma.

Since 1950, 413 children at the Hospital for Sick Children have been diagnosed as suffering ITP, but only six have suffered intracranial hemorrhage. In two, the diagnosis was presumed; four others are reported here. The threatening intracerebral hemorrhage in these children presents an urgent problem for which a successful outcome will be obtained only if a coordinated plan of management involving hematologist, general surgeon, radiologist, and neurosurgeon is followed using orthodox neurosurgical techniques. Based on our experience, we have developed the following approach. First, the diagnosis of ITP, if not previously established, must be confirmed by bone marrow aspiration. Simultaneously, traditional methods to reduce cerebral edema using intravenous dexamethasone and mannitol (20% solution) should be started. In addition to its beneficial effect on cerebral edema (which in two of the children was severe), dexamethasone was administered during surgery and for a variable time thereafter to facilitate the platelet response. Lumbar puncture is contraindicated.

The subsequent surgical and investigative management can, if necessary, be conducted under a single anesthetic. Emergency splenectomy should be carried out and followed by infusion of platelet concentrates, which then seem to have more hemostatic effect since an increment in platelet counts can usually be obtained. Cerebral angiography may then be performed to outline the site, size, and nature of the intracerebral clot. After radiological examination, in critical situations, the patient may be transferred under anesthesia back to the operating room for craniotomy and clot evacuation. The availability of computer tomography permits the pre-splenectomy identification of the cerebral hematoma. Then, splenectomy followed by craniotomy can be performed under a single anesthetic in one operating room.

Splenectomy alone (Case 2), or with corticosteroids (Cases 1 and 3) appeared to be
useful. Falling platelet counts after cessation of corticosteroid therapy (Case 1) implied that the site of reticuloendothelial destruction shifted from spleen to liver. In Case 4, although the success of splenectomy and steroids was not obvious from the platelet counts, the absence of further bleeding may mean that the platelet survival was increased enough to reduce bleeding episodes.

In the three surviving children, there are no major neurological sequelae despite the fact that all the hemorrhages were intracerebral.

Acknowledgment

We wish to thank Dr. Harold J. Hoffman for permission to include the patient in Case 4, who was under his care.

References


Address reprint requests to: Robin P. Humphreys, M.D., Division of Neurosurgery, Room 2223, Hospital for Sick Children, 555 University Avenue, Toronto, Ontario M5G 1X8, Canada.