Anesthesia and Perioperative Management for Kasabach-Merritt Syndrome

— Report of a Case —

Hideo HORIKAWA, Toshihiko TADA and Osamu HONDA

(Key words: anesthesia, syndrome, management)

Kasabach-Merritt syndrome was first reported in 1940 as a capillary hemangioma associated with thrombocytopenic purpura, and is also called hemangioma-thrombocytopenia syndrome[1]. This thrombocytopenia is thought to be due to the consumption of platelets by local intravascular coagulation in the hemangioma. Derangement of the coagulation-fibrinolysis complex can be a serious problem during major surgical operations and anesthesia.

Case report

A 38-year-old female, 152 cm and 48 kg, with a complaint of malaise, was first found to have a gigantic hepatic hemangioma in June, 1989, and was transferred to Yamagata University Hospital in August, 1989. On admission, no purpura was detected (purpura, however, was noticed thereafter on occasions of venipuncture). Her abdomen was markedly distended with the lower edge of the liver reaching the right inguinal region. This finding, together with a set of abnormal values in the coagulation-fibrinolysis complex, represented most remarkably by a low platelet count (table 1), led to a diagnosis of Kasabach-Merritt syndrome. Computer-tomographic, nuclear, angiographic and sonographic studies all indicated that she was a candidate for a surgical removal of the left lobe of the liver. Partial hepatic resection was scheduled on the 55th day after admission.

Preoperatively, gabexate mesilate (FOY) was infused via the right subclavian vein at a rate of 2 $mg \cdot kg^{-1} \cdot hr^{-1}$ for 14 days, and 5 units of fresh frozen plasma (FFP) were given daily for 6 days preceding the operation.

Anesthetic and Operative Procedures

Anesthesia was induced with i.v. thiopental, fentanyl and vecuronium, and was maintained with a nitrous oxide-oxygen-enflurane combination and pancuronium. A sheath (Arrow International, Reading, PA) for an 8.5 Fr Swan-Ganz catheter was placed in the right internal jugular vein and was used as an infusion route for whole blood and FFP. A double-lumen (18G and 14G) catheter (Arrow) was inserted into the left internal jugular vein, and was used for continuous infusion of FOY 2 $mg \cdot kg^{-1} \cdot hr^{-1}$, d-butyric cyclic AMP (DBcAMP) 5–10 $\mu g \cdot kg^{-1} \cdot min^{-1}$ and prostaglandin E_1 (PGE_1) 0.01 $\mu g \cdot kg^{-1} \cdot min^{-1}$. Preoperatively a 7 Fr Swan-Ganz catheter was

Department of Anesthesia, Yamagata University School of Medicine, Yamagata, Japan

Address reprint requets to Dr. Horikawa: Department of Anesthesia, Yamagata University School of Medicine, Yamagata, 990-21 Japan

Table 1. Laboratory data on coagulation and bleeding

	Normal Value	On Admission	Immediate Preoperative	On Discharge
Hb ($g \cdot dl^{-1}$)	13	9.8	7.8	8.6
Platelet ($/mm^3$)	270,000	106,000	82,000	170,000
PT (%)	>80	54.0	41.3	53.0
APTT (%)	>80	83.6	73.9	85.5
Fibrinogen ($mg \cdot dl^{-1}$)	200–400	131.4	135.2	302.1
FDP ($\mu g \cdot ml^{-1}$)	<10	20–39	20–39	20–39
Bleeding Time (min)	<3	4.5	5.5	

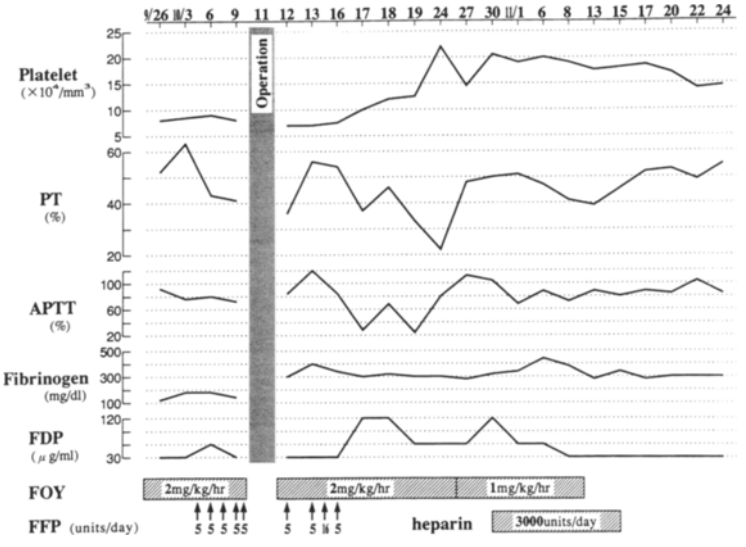

Fig. 1. The postoperative course of the patient with Kasabach-Merritt syndrome

in place through the right antecubital vein, and was used for perioperative hemodynamic monitoring.

The left lobe of the liver was successfully removed and hemostasis was done nearly perfectly. There were no remarkable events during anesthesia and the operation. Hemodynamic variables were reasonably stable. The time for the operative procedures was 7 hr 20 min and that for anesthesia was 8 hr 45 min. The measured total blood loss was 3,841 ml and the amounts of FFP and fresh whole blood transfused were 2,380 ml and 2,200 ml, respectively. The resected lobe of the liver (3,090g) was occupied by a gigantic cavernous hemangioma, but no sign of malignancy was found histologically.

After the operation, the patient was sent to the intensive care unit, where the treatment regimen was centered on the normalization and maintenance of the coagulation function. The postoperative course is shown in figure 1. FFP was infused and FOY was also given at a rate of 2 $mg \cdot kg^{-1} \cdot hr^{-1}$. Postoperative bleeding from the liver was 60 $ml \cdot hr^{-1}$ in the beginning but it decreased rapidly. The patient was transferred to the surgical ward on the 4th postoperative day.

In the ward coagulation tests were carried out repeatedly and, together with FOY, 3,000 units of heparin was used daily for 17 days. On the 46th postoperative day, the patient was discharged from the hospital with fair improvement in the coagulation function (table 1).

Discussion

Kasabach-Merritt syndrome is usually presented as a benign hemangioma. Eighty per cent of the patients with this syndrome are under one year of age[2]. In adults hemangioma is often seen in the liver and tends to be malignant. Reports concerning the surgical operation for gigantic hepatic hemangioma have been very few[3–5], and none gave a detailed account of the anesthetic management.

For hemangiomas large-dose steroid therapy[6], blood vessel ligation[7], cryotherapy[8], radiation therapy[9], embolization of the hepatic artery[10], etc., have been tried with varying degrees of success. It has been pointed out that in these patients such minor insults as a biopsy and blood sampling could lead to aggravation of bleeding tendency and even to hemorrhagic shock[11]. Our patient already demonstrated an established coagulaopathy and malaise. Therefore, surgical operation was chosen, omitting invasive examinations preoperatively.

In order to suppress the initiation of intravascular coagulation in the gigantic hepatic hemangioma as much as possible, FOY was given intravenously at a rate of 2 $mg \cdot kg^{-1} \cdot hr^{-1}$ for 14 preoperative days. The normalization of the prothrombin time (PT), activated partial prothrombin time (APTT), fibrin and fibrinogen degradation products (FDP) and fibrinogen was hoped for from this treatment. These items, however, worsened, rather than improved (table 1). The patient required daily administration of 5 units of FFP for the 6 immediate preoperative days. Heparin may have been indicated in this case but it was not used for fear of the massive bleeding that may accompany an extensive resection of the hepatic lobe.

One problem which demands the utmost attention is the development of bleeding tendency. Both anesthesia and surgical operation have been reported to trigger disseminated intravascular coagulation and accompanying profuse bleeding[12]. Epidural analgesia, routinely combined with inhalation anesthesia for intra-abdominal procedures in our institution, was not used for fear of possible epidural bleeding. As procurement of reliable intravenous infusion routes in the extremities was anticipated to be extremely difficult in this case, large-bore catheters were placed in the bilateral internal jugular veins. It may have been more justifiable to choose venous cutdowns in the extremities than to insert catheters blindly into the deep neck veins in the presence of coagulopathy, but no serious problem arose from this procedure.

It has been reported[13] that with inhalation anesthesia with enflurane the hepatic blood flow decreases and hepatic venous blood saturation declines markedly at the time of skin incision. In the present case, where epidural block was not used, it was apprehended that general anesthesia alone might not be sufficient to prevent this decrease in the hepatic blood flow. Adverse effects of low cardiac output and hypotension accompanying massive bleeding on the hepatic metabolism were also to be dealt with. DBcAMP and PGE_1 preparations were used in the hope of alleviating these undesirable effects of bleeding.

Bleeding from the operative surface of the liver requires the closest attention in the period of postoperative management. Substantial amounts of

FOY and FFP used postoperatively resulted in gradual increases in platelets and fibrinogen. Normalization of FDP, however, was rather slow. Heparin was used for 16 days at a dose of 3,000 $U \cdot day^{-1}$ to accelerate the decline of FDP. It has been reported in a multi-institutional study[14] that a combination of FOY and heparin was more effective in these circumstances than using these substances separately to prevent this complication.

In summary, operative and anesthetic management of a very rare pathology, Kasabach-Merritt syndrome, was successfully achieved with meticulous perioperative care including: normalization of coagulation function with FOY and FFP preoperatively, stabilization of hemodynamics with FOY, DBcAMP, PGE_1, fresh whole blood and FFP intraoperatively, prevention of blood loss with FFP, FOY and heparin postoperatively, and, above all, close observation of the patient.

(Received Oct. 1, 1991, accepted for publication Feb. 3, 1992)

References

1. Kasabach HH, Merritt KK: Capillary hemangioma with extensive purpura: Report of a case. Am J Dis Child 59:1063–1070, 1940
2. Henricksson P, Nilsson IM, Bergentz SE, et al: Giant hemangioma with a disorder of coagulation. Acta Pediat Scand 60:227–234, 1971
3. Miura K, Yoshida K, Matsubara Y, et al: Surgical treatment of cavernous hemangioma of the liver with disseminated intravascular coagulation. Jpn J Gastroenterol 81:112–116, 1984
4. Watzke HH, Linkesch W, Hay U: Giant hemangioma of the liver (Kasabach-Merritt syndrome): Successful suppression of intravascular coagulation permitting surgical removal. J Clin Gastroenterol 11:347–350, 1989
5. Klompmaker IJ, Slooff MJH, van der Meer J: Orthotopic liver transplantation in a patient with giant cavernous hemangioma of the liver and Kasabach-Merritt syndrome. Transplantation 48:149–151, 1989
6. Özsoylu S, Irken G, Gürgey A: High dose intravenous methylprednisolone for Kasabach-Merritt syndrome. Eur J Pediat 148:403–405, 1989
7. de Lorimer AA, Simpson EB, Baum RS, et al: Hepatic artery ligation for hepatic hemangiomatosa. N Engl J Med 277:333-337, 1967
8. Kawahara M, Takeshita T, Akira S: Anesthetic management of a patient with Kasabach-Merritt syndrome. Anesth Prog 34:17–19, 1987
9. James DH Jr, Tuttle AH: Congenital hemangioma with thrombocytopenia. J Pediat 59:234–237, 1961
10. Vomberg PP, Buller HA, Marsman JWP, et al: Hepatic artery embolization: Successful treatment of multinodular haemangiomatosis of the liver. Eur J Pediat 144:472–474, 1986
11. Mimaya J, Nagao T, Hanawa Y, et al: Four cases of giant hemangioma with coagulopathy (Kasabach-Merritt syndrome). Jpn J Clin Hematol 14:1046–1053, 1973
12. Lang PG, Dubin HV: Hemangioma thrombocytopenia syndrome: A disseminated intravascular coagulopathy. Arch Dermatol 111:105–107, 1975
13. Kainuma M, Fujiwara Y, Kimura N, et al: Monitoring hepatic venous hemoglobin oxygen saturation in patients undergoing liver surgery. Anesthesiology 74:49–52, 1991
14. Kosaski G, Kambayashi J, Hirayama A, et al: A study on the therapeutic effects of FOY against disseminated intravascular coagulation. —A multi-institutional study—. J Clin Exper Med 124:144-154, 1983 (in Japanese)