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Case Report

Embolization for Refractory Subacute Subdural Hematoma in a Child with Severe Hemophilia Type A

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Hemophilia is an X-linked hemorrhagic disease due to coagulation factor VIII or IX deficiency with approximately 5–10% incidence of central nervous system bleeding. We present an intriguing case of a refractory subacute subdural hematoma (SDH) controlled with endovascular embolization in a hemophilic patient. A 5-year-old severe hemophilic A boy presented with a life threatening left parietal subcortical hemorrhage, for which he underwent craniotomy and evacuation of the hematoma. Recurrent hemorrhage necessitated a repeat craniotomy. This was followed by three episodes of SDH development at the craniotomy site that were treated surgically, and finally controlled with embolization in the subacute period. This case presents a novel option for treating a refractory SDH in patients with coagulation disorders.

Keywords: embolization, hemophilia, subdural hematoma, endovascular, coagulation disorder

Introduction

Endovascular embolization is reported to be effective for refractory chronic subdural hematoma (SDH). However, no report exists on the efficacy of embolization for subacute SDH. Likewise, the role of embolization in SDH with coexisting blood dyscrasia is unknown. We report our experience on treating a refractory SDH in the subacute stage in a patient with severe hemophilia type A.

Clinical Presentation

A 5-year-old boy with a severe hemophilia A with inhibitor against factor VIII who had been under immune tolerance therapy presented with a left parietal hematoma, for which he underwent craniotomy and evacuation of the hematoma (Fig. 1a, b). The hemophilia was considered severe because less than 1% of the factor VIII was present on laboratory examination. Post-operatively, recombinant factor VIII replacement therapy was initiated because of the low level of the inhibitor.

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On the 28th post-operative day, a recurrent left parietal subcortical hematoma necessitated another craniotomy. Because of the elevation of the inhibitor titer, a recombinant activated factor VII and a freeze-dried sterile human plasma fraction with factor VIII inhibitor bypassing activity (bypass therapy) were added in addition to the recombinant factor VIII. The bypass therapy was repeated for each periprocedural period thereafter. Post-operatively, an asymptomatic collection of subdural cerebrospinal fluid in the left parietal region was seen (Fig. 1c, d).

Two months after the second surgery, the subdural cerebrospinal fluid collection transformed into a chronic SDH, causing change in consciousness and right hemiparesis (Fig. 1e). The hematoma was evacuated by burr hole and irrigation surgery.

Seven days after the third surgery, rapid deterioration in consciousness prompted a head computed tomography (CT) study (Fig. 1f). The study revealed an acute SDH in the left parietal region, for which the patient underwent a craniotomy and evacuation of the hematoma with concomitant coagulation of the left middle meningeal artery (MMA). Post-operative serial head CT scans showed progressive accumulation of a left parietal acute/subacute SDH resulting in a marked midline shift by the seventh post-operative day (Fig. 1g).

Because of the refractory nature of the SDH, endovascular embolization of the dural arteries that supplied the SDH capsule was planned with the aim to change the trajectory of the disease. During the embolization, the left external carotid artery angiogram demonstrated opacification of the SDH capsule that was supplied from the petrosquamous branch of the MMA and the transmastoid and stylomastoid branches of the occipital artery (Fig. 2). A microcatheter was superselectively navigated into each branch and then embolized with fine gelatin particles until complete occlusion of the capsular vessels was obtained (Fig. 3). After the embolization, immune tolerance therapy was resumed.

Following the embolization, the growth of the hematoma was stabilized and subsequently regressed in the course of 1 month. Nine-month post-embolization follow up CT demonstrated complete resolution of the SDH (Fig. 4).

Discussion

Hemophilia A and B are X-linked inherited disorders of the coagulation factors characterized by spontaneous bleeding. Approximately 5–10% of all severe hemophilic patients develop at least one episode of intracranial hemorrhage (ICH: epidural, subdural, subarachnoid, and intracerebral) in their lifetime.[1–5)]

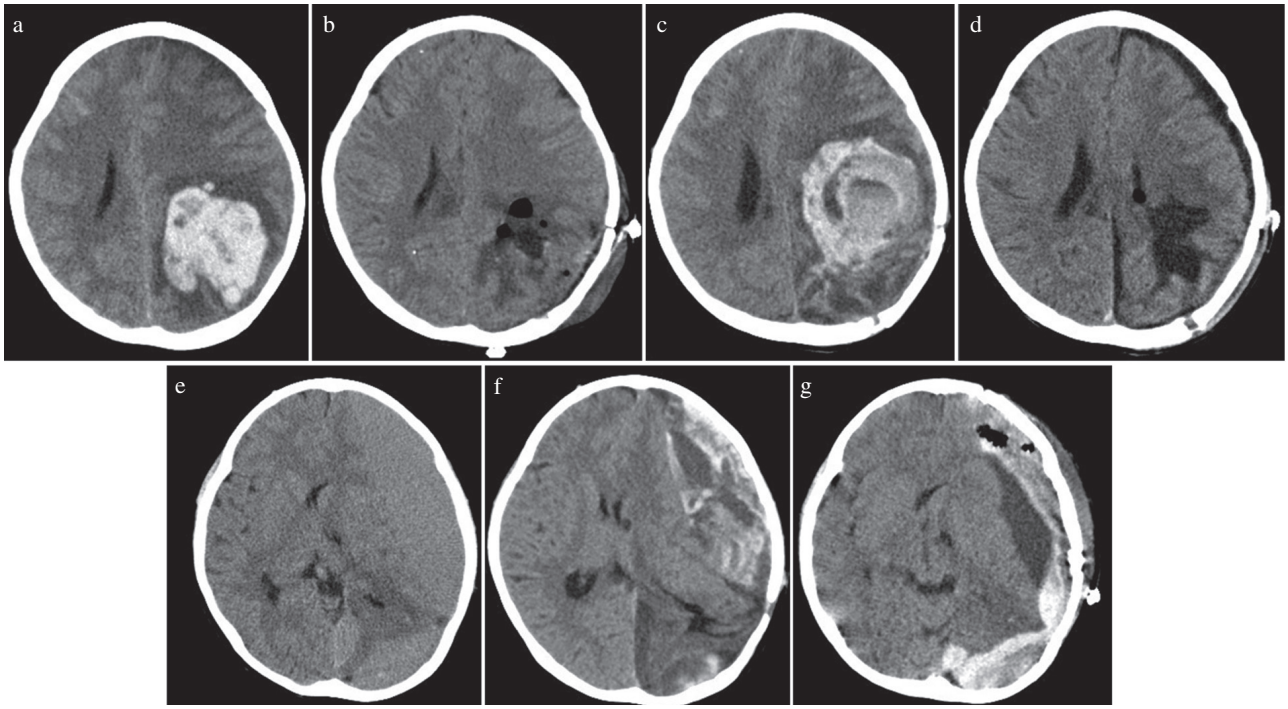

Fig. 1 Head CT scans demonstrating the course of the intracranial hemorrhage. Head CT at presentation (a) demonstrates a left parietal subcortical hematoma that was evacuated by craniotomy (b). Head CT obtained on the 28th post-operative day after sudden deterioration in consciousness shows a recurrence of the left parietal subcortical hematoma (c) and the patient underwent another craniotomy. Post-craniotomy image shows collection of subdural fluid at the craniotomy site (d). Two months after the second craniotomy, the subdural effusion developed into a symptomatic chronic subdural hematoma (SDH) (e), which was evacuated by a burr hole surgery. Seven days later, the patient developed an acute SDH (f), for which he underwent a third craniotomy. Post-operative serial CT scans demonstrated progressive development of an acute/subacute SDH (not shown) resulting marked midline shift on the seventh day (g).

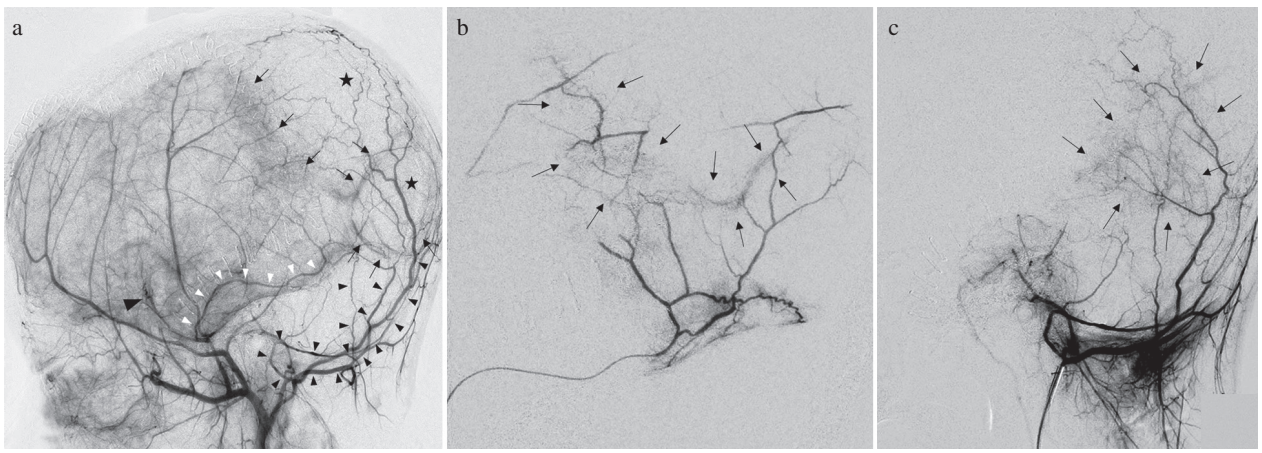

Fig. 2 (a) Left external carotid artery angiogram in lateral view demonstrates the hypervascular capsule of the subdural hematoma (SDH) (arrows). The feeders of the capsule are the petrosquamosal branch of the middle meningeal artery (MMA) (white arrowheads) and the transmastoid and stylomastoid branches arising from the occipital artery (arrowheads). The stars indicate the avascular area representing the SDH. Note the stump of the frontal branch of the MMA that was coagulated during the craniotomy (large arrowhead). (b) Superselective injection of the petrosquamosal branch of the MMA in lateral view demonstrates the hypervascular SDH capsule in the anterior parietal region (arrows). (c) Superselective injection of the occipital artery in lateral view demonstrates the hypervascular capsule in the posterior part of the SDH (arrows).

Management of the ICH in hemophilic patient centers either on conservative treatment with coagulation factor replacement or surgical evacuation.[6,7] The role of endovascular therapy in this setting has never been reported to date. Because endovascular therapy can be safely performed in patients with coagulation disorder and is less invasive, this therapy may be applicable to selected patients with hemophilia and ICH.[8] In the present case, embolization of the responsible arteries achieved immediate stabilization of the SDH. This was subsequently followed by the regression

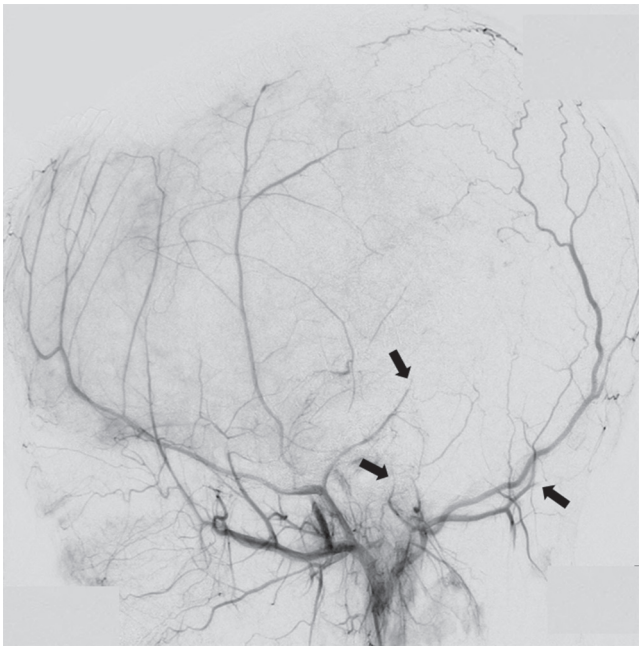

Fig. 3 Post-embolization left external carotid artery angiogram in lateral view. The angiogram shows complete occlusion of the hypervascular hematoma capsule. Note the stump of the embolized vessels (arrows).

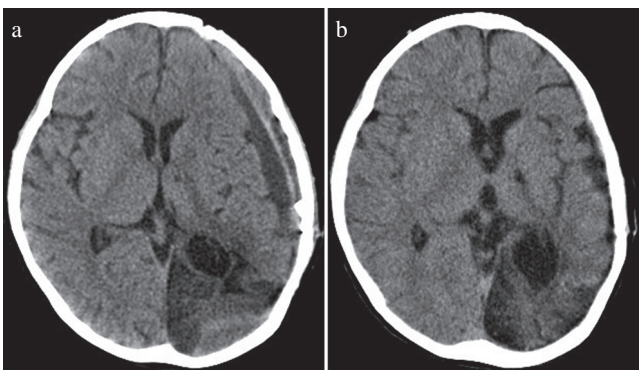

Fig. 4 Head CT on 1-month post-embolization demonstrates regression of the left subdural hematoma (SDH) (a). The SDH continued to regress until complete resolution is seen on 9-month post-embolization CT (b).

of the SDH in the course of 1 month. Since then, the patient remains stable without recurrence for 3 years. We consider that the embolization was able to change the trajectory of the aggressive SDH associated with the underlying Hemophilia A.

In addition, we describe the first case of a subacute SDH treated by endovascular embolization. Previous reports of embolization for the treatment of SDH were applied for refractory chronic SDH. The first report of such case was made by Mandai et al. in 2000.[9] Embolization of the MMA of the affected side resulted in the successful resolution of the chronic SDH. This was followed by a number of studies that demonstrated the efficacy of the treatment.[10–14] The rational for this treatment was based on the findings by Tanaka et al. that many small branches of the dural artery on connected to the fragile microcapillaries in the outer membrane of the chronic SDH capsule.[15–18] Repetitive bleeding and chronic inflammation are considered the cause of the SDH expansion. Therefore, embolization of the dural artery is reasonable for the treatment of refractory chronic SDH. In our case, the treatment was performed in the subacute phase after a craniotomy and durotomy when the outer membrane of the SDH may not have well developed. On the other hand, the subacute hematoma developed in the identical space as the initial chronic hematoma. Because of this, the outer membrane of the hematoma might have already existed when the subacute hematoma developed. Whichever was the case, the effectiveness of the embolization in the subacute SDH may be explained by a similar pathology as in the chronic SDH.

As for the embolic material of choice, we adopted the gelatin particles. The gelatin particles are associated with more recanalization due to absorption of the material.[19] However, we chose the particles because of its higher capability to be sprayed into a wide area of the capillary bed. The other major embolic agent, n-butyl cyanoacrylate, is a permanent embolic material with the potential for more complete embolization than the gelatin particles.[20] However, the risk of proximal feeding artery occlusion and the resultant incomplete embolization could not be negated, and therefore, we chose to use the gelatin particles. The size of the gelatin particles under microscopic inspection was between 100 and 250 μm in diameter. Because of the small size of the particles, there exists a risk of occluding the micro-vessels supplying the cranial nerve, which is approximately 200–300 μm in diameter. In our case, the microcatheter was advanced well beyond the stylomastoid foramen, where embolization proximal to this risks the facial nerve palsy, into the parietal convexity.

A limitation of the endovascular embolization is that the treatment cannot reduce the mass effect and the increased intracranial pressure caused by the SDH. However, whenever the situation allows, since embolization is a less invasive treatment, this may be considered as an option for the followings: 1) a refractory SDH in a patient with a coagulation disorder, 2) refractory subacute SDH. Further investigations are warranted to assess the efficacy and safety of this treatment.

Conclusion

Endovascular embolization was able to change the trajectory of the rapidly expanding refractory SDH in a hemophilia patient and may be considered as a treatment option in selected cases. In addition, embolization was effective in the subacute period when the hematoma showed progressive expansion.

Conflict of Interest Disclosure

The authors declare that they have no conflict of interest.

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