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Sequential combined bypassing therapy for refractory bleedings in two adolescent hemophiliacs with inhibitors

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OBJECTIVES

Hemophilia patients who have developed inhibitors often experience several bleeding episodes, and 10% to 20% of these episodes are extremely difficult to manage with bypassing agent monotherapy alone. Few reports have described the use of sequential therapy with recombinant factor VIIa (rVIIa) and activated prothrombin complex concentrate (APCC) within 12 hours of a bleeding event in this patient group (1,2). Here we report our experience with this treatment for two hemophiliacs with inhibitors.

METHODS

We present two patients with severe hemarthrosis and muscle hematomas that cannot be controlled with bypassing agent monotherapy. The first patient was an 18-year-old male with severe hemophilia A with high inhibitor titers (range, 35- 95 BU) since the age of 3, During the 3 months prior to presentation, he had experienced frequent bleeding episodes in his right knee and had developed joint damage that was assessed as grade III according to the WFH grading system. Despite monotherapy with rVIIa or APCC, he had developed chronic synovitis and difficulty walking. The second patient was a 19-year-old male with hemophilia B with high inhibitor titers since age 5. At the most recent admission, the patient had presented with psoas bleeding and a hematoma in his right arm muscle. The hematoma did not regress despite 1 month of on-demand treatment with rVIIa or APCC monotherapy. Multiple small cysts formed in the muscle tissue around the hematoma.

RESULTS

Sequential combined bypassing therapy for first patient was administered using alternating doses of APCC and rVIIa within 12 hours of a bleeding event (i.e., one dose of APCC (25-50 U/kg) followed by one to three doses of rVIIa (90-120 µg/kg) depending on occurrence of heavy bleeding). After 2 week of this therapy, the youth had excellent joint function. For second patient, this sequential therapy was administered as first-case treatment. The patient was followed up weekly, and the hematoma gradually decreased and none of the cysts enlarged.

CONCLUSIONS

Our experience with these two cases indicates that for hemophilia patients with inhibitors, sequential combined bypassing therapy is likely a good choice for treating bleeding episodes that cannot be controlled with bypassing agent monotherapy.

References

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