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Case Reports & Case Series

Spontaneous intracranial hypotension complicated by refractory subdural hematomas in a patient with coagulation factor XIII deficiency

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ABSTRACT

Spontaneous intracranial hypotension (SIH) is often secondary to an occult cerebrospinal fluid (CSF) leak in the neuroaxis. It is sometimes associated with chronic subdural hematomas (SDH). The authors present the case of a 61 year-old male who presented with SIH from an occult CSF leak and complicated by refractory SDH. The patient underwent treatment for SIH with multiple epidural blood patches, hydration and bed rest. In addition, he underwent evacuation of his SDH twice, once via burr hole drainage and the second via craniotomy. Once the diagnosis of coagulation factor XIII (CFXIII) deficiency was made, the patient was treated with recombinant factor XIII followed by an epidural blood patch. He subsequently made a complete recovery. In this report, the authors propose management strategies for treating patients with SIH complicated by refractory SDH and coagulopathies by reviewing the literature.

1. Introduction

Spontaneous intracranial hypotension (SIH) is often secondary to an occult cerebrospinal fluid (CSF) leak in the neuroaxis. It is sometimes associated with chronic subdural hematomas (SDH). The combination of clinical symptoms and neuroimaging should alert the practitioner of SIH and therefore guide the practitioner to the appropriate treatment [4,6]. The treatment of choice in this situation is an epidural blood patch if conservative measures such as bedrest and hydration fail. For larger SDH, evacuation may be necessary in conjunction with an epidural blood patch. However, refractory subdural hematomas may complicate the situation despite appropriate management [7]. Coagulation factor XIII (CFXIII) is a clot stabilizing factor in the clotting cascade and has also been shown to contribute to wound healing by encouraging the migration and proliferation of fibroblasts into the clot [1,5]. We present a case of a healthy 61 year-old male who presented with SIH from an occult CSF leak and complicated by refractory SDH. He did not respond to treatment until the diagnosis of CFXIII was made and treated appropriately.

2. Case presentation

A 61 year-old right handed male with no past medical history presented to our emergency department with 3 days of progressively worsening headaches. The headaches were severe and positional in nature. He had a nonfocal neurologic examination. A noncontrast commuted tomography (CT) of the head showed bilateral hypodense subdural collections (Fig. 1A). At this point, spontaneous intracranial hypotension (SIH) as a result of occult cerebrospinal fluid leak (CSF leak) was suspected based on clinical and radiological exam.

As part of his work-up for an occult CSF leak magnetic resonance imaging (MRI) of the neuroaxis (brain, cervical, thoracic and lumbar spine) was obtained which demonstrated several small nerve root sleeve cysts in the midthoracic region. A CT myelogram was subsequently obtained with no further findings. Since no obvious source of leak was found, the decision was made to manage him conservatively with bed rest, intravenous fluids, and caffeine pills. His headaches improved substantially after 2 days and he was discharged home after a CT demonstrated stability of his subdural collections.

He returned one week later with severe positional headaches. A follow-up CT scan demonstrated an increase in the size of the subdural

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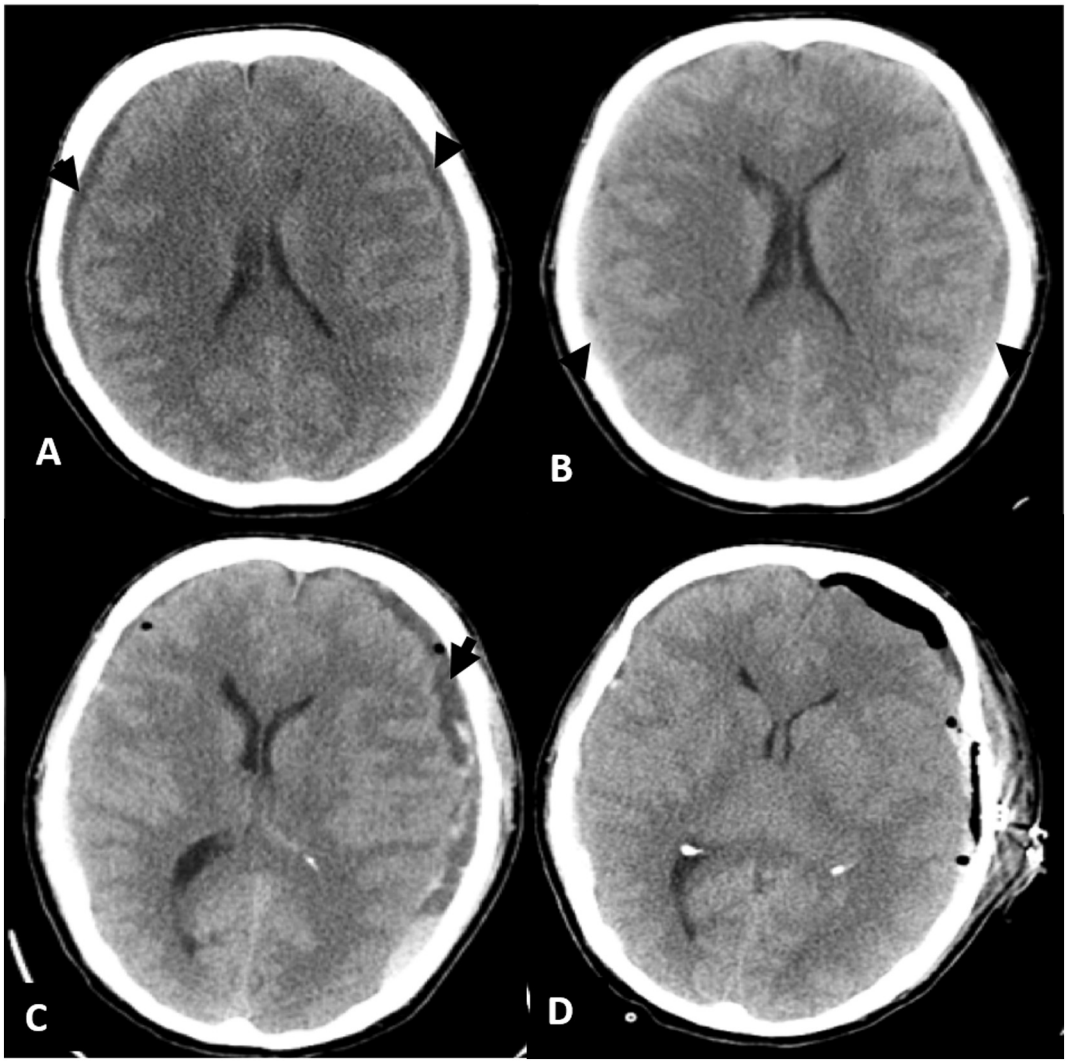

Fig. 1. Axial CT of the head on presentation (A), 7 days after presentation (B), 4 days after bilateral burr hole drainage (C), and post-craniotomy (D) demonstrating the subdural hematomas.

collections with additional acute blood (Fig. 1B). Although nonfocal on his initial neurologic exam, he became lethargic despite intravenous fluids and lying flat for 24 h. The decision was then made to take him to the operating room and drain the subdural hematomas with bilateral burr holes. On postoperative day one his symptoms improved and a postoperative CT demonstrated significant reduction in the subdural fluid collections. However, by the second postoperative day, his symptoms of severe positional headaches and lethargy recurred. At this point, a blood patch was performed using 60 ml of autologous blood injected through a spinal needle into the epidural space of the lumbar spine.

The patient's headaches did not improve after the blood patch. In addition, he developed word finding difficulties. Due to the new focal neurologic deficit, a CT scan was performed which showed an increase in size of the left subdural collection with associated mass effect and acute blood (Fig. 1C). Due to the reappearance of acute blood in the subdural collection, a hematology consult was obtained. He was taken to the operating room for a left sided craniotomy and evacuation of the subdural hematoma. A postoperative CT demonstrated evacuation of the hematoma (Fig. 1D) and a second blood patch was performed. The patient initially improved.

By postoperative day 2, despite being kept flat, he became progressively obtunded and required intubation. An MRI of the brain demonstrated downward displacement of the cerebellar tonsils and splenium of the corpus callosum with severe crowding of the foramen magnum. Progressive sulcal, third and fourth ventricle, and basal cistern effacement was seen consistent with severe intracranial hypotension. The patient was placed in 30 degrees Trendelenberg and within 15 min he regained consciousness. At this time, his hematology work-up revealed a severe coagulation factor XIII deficiency, at a level of 23% in his plasma (normal range, 70–140%). The patient was given recombinant factor XIII intravenously followed by a third blood patch. He subsequently made a complete recovery. A surveillance CT scan was performed as an outpatient and demonstrated complete resolution of the subdural collections.

3. Discussion

Spontaneous intracranial hypotension (SIH) should be suspected in patients with postural headaches. In patients with SDH who have postural headaches and no history of trauma or coagulopathy, the diagnosis and treatment of SIH is critical for a favorable outcome. Recurrence of SDH associated with SIH is common until treatment of the underlying CSF leak is achieved [3,7]. There is ample evidence that epidural blood patch (EBP) alone is effective in treating patients with chronic SDH associated SIH even when the site of CSF leak has not been identified [3]. Repeated EBP may be required in patients. In the rare refractory patient, surgical repair of a CSF leak is required but is

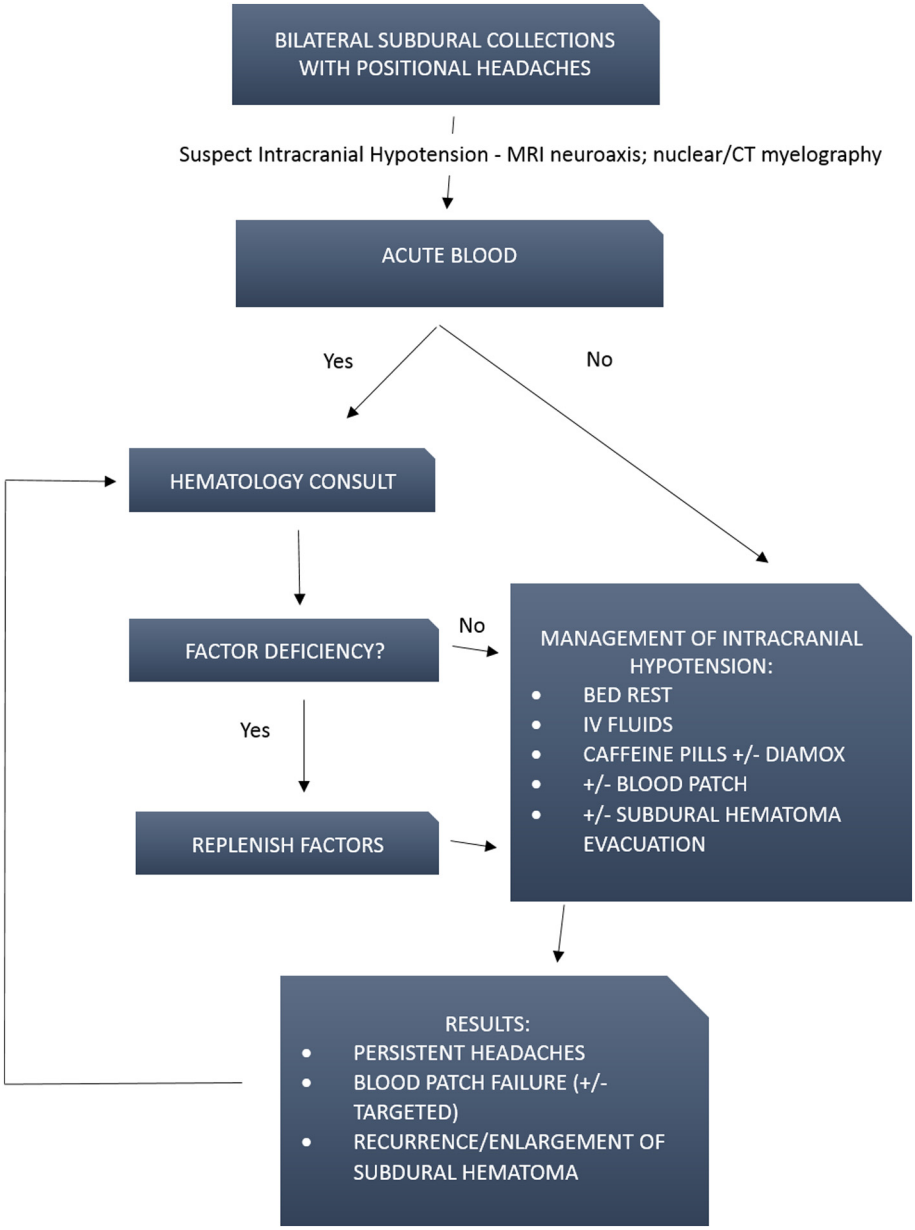

Fig. 2. Proposed algorithm for the management of subdural hematoma associated spontaneous intracranial hypotension, which includes proper diagnosis and treatment of a factor deficiency.

dependent on establishing the site of the leakage [6]. There are reports that targeted EBP may be more effective in sealing the leak the first time, especially in patients with concomitant SDH, however, they are associated with a higher complication rate, including spinal cord injury [9]. Occluding the CSF leak is critical in the treatment of SDH associated SIH and therefore may require multiple procedures to accomplish this with or without subsequent evacuation of the SDH in neurologically compromised patients.

Nevertheless, some patients may remain refractory to treatment. There has been an association between coagulopathy and chronic recurrent SDH, particularly in young patients [1,2]. Specifically, low levels of factor XIII have been associated with recurrent SDH. Coagulation factor XIII (CFXIII) stabilizes fibrin in the clotting cascade and protects it against fibrinolysis by mechanically cross-linking fibrin chains and alpha-2 plasmin inhibitor. It is also involved in wound healing by stabilizing endothelial barrier function and vascular integrity. Moreover, the pathophysiology of chronic SDH, with the fragile neovascularization of membranes, can further potentiate decreased CFXIII levels through a consumptive process, thereby starting a vicious cycle in patients who are already compromised [1,2,5,8].

In CFXIII deficient patients, with chronic recurrent SDH, treatment with recombinant CFXIII has been shown to be effective. Furthermore, CFXIII has been used to treat SIH refractory to treatment, extrapolating from wound healing properties studied in the literature. In a retrospective review, 9 patients with SIH, without SDH, received intravenous recombinant CFXIII without complication and favorable outcomes. All patients had normal CFXIII levels prior to treatment [5]. Although an option, one must consider the high cost of recombinant CFXIII and the potential serious thrombotic complications that may occur and should therefore not be used routinely.

CFXIII activity is not routinely assessed on admission in most medical centers unless there is strong medical suspicion of a coagulopathy. Furthermore, results are obtained 2–3 days after the blood is drawn, which may delay diagnosis and treatment. Early recognition for the need to evaluate for CFXIII deficiency is critical in patients with SDH associated SIH. In their series, Shimogawa, et al. retrospectively

identified 206 patients who had CFXIII activity data obtained upon admission to their hospital. Of these patients, they identified nineteen SDH associated with SIH patients who necessitated treatment with EBP with or without SDH evacuation. Seven patients (36.8%) developed SDH exacerbation post-treatment. This group of patients had significantly lower CFXIII activity than those without post-treatment exacerbation. In addition, the CFXIII activity was significantly lower in patients with than without a chronic SDH (42.1% vs. 12.8%). All five patients with low CFXIII activity who developed SDH exacerbation received intravenous recombinant CFXIII and had no recurrence after this treatment [8].

To our knowledge, there have been no treatment algorithms discussed in the literature for patients with chronic SDH associated with SIH and low CFXIII. We report a case of a patient with SDH associated with SIH and CFXIII deficiency. Based on this experience and the literature, we propose an algorithm for patients who present with SDH associated SIH in order to prevent delays in diagnosis and treatment, as well as complications (Fig. 2). If SDH associated with SIH is suspected, one should confirm SIH with diagnostic images such as MRI of the neuroaxis, nuclear medicine myelography, or CT myelography. One should then consider studies to evaluate for a factor deficiency if there is acute blood on imaging. If treatment with an EBP with or without SDH drainage fails, one should also consider evaluating for a factor deficiency. Early identification and treatment in patients with a factor deficiency with intravenous recombinant factors will prevent potential unnecessary multiple procedures and/or complications.

Compliance with ethical standards

There are no conflicts of interest to report in preparation of this manuscript.

There was no participation of human or animal subjects in the preparation of this manuscript.

Consent was obtained from the patient to publish this manuscript.

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