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SPONTANEOUS ACUTE SUBDURAL HEMATOMA AND CHRONIC EPIDURAL HEMATOMA IN A CHILD WITH F XIII DEFICIENCY

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□ **Abstract—Factor XIII (F XIII) deficiency is a rare autosomal recessive congenital disorder that can cause spontaneous subdural or epidural hematomas. Due to its low incidence, F XIII deficiency may well be under-diagnosed. A 7-year-old girl with no history of medical problems presented with progressive headache of 3 days. Cerebral computed tomography (CT) scans revealed a large right acute parietooccipital subdural hematoma with a significant midline shift. After an emergent parietooccipital craniotomy and evacuation of the subdural hematoma, a screening test for factor XIII was performed. The results of the test were abnormal. She had full recovery and was discharged with a follow-up treatment of monthly transfusion of fresh frozen plasma as the replacement and prophylactic therapy. Ten months later, she was referred to our center with headache after a minor head trauma. Her medical history revealed that she had not received fresh frozen plasma for the last 2 months. CT scan showed a chronic right parietal epidural hematoma beneath the craniotomy flap. The present case indicates that although its incidence is very rare, F XIII deficiency can cause acute or chronic subdural and epidural hematomas. Therefore, in acute or chronic subdural and epidural hematomas with no underlying cause, the presence of a potential F XIII deficiency should be suspected as a cause of hemorrhagic diathesis.** © 2010 Elsevier Inc.

□ **Keywords—F XIII deficiency; epidural hematoma; subdural hematoma; pediatric age**

INTRODUCTION

Spontaneous acute subdural hematoma (SDH) is a rare condition (1,2). Potential predisposing factors are hypertension, anticoagulant therapy, bleeding and coagulation disorders, cocaine-induced vasculitis, and bleeding from intracranial tumors or aneurysms (1,3,4). Epidural hematoma (EDH) often results from a local injury (5). Factor XIII (F XIII) is a transpeptidase that is necessary for normal blood hemostasis. In F XIII deficiency, blood clot is weak and thus unable to maintain a hemostatic plug, leading to hemorrhages (6,7). Intracerebral hemorrhages due to factor XIII deficiency have been shown in various case reports (8–12). To the best of our knowledge, this is the first case of acute SDH and chronic EDH 10 months after the first operation.

CASE REPORT

A 7-year-old girl was admitted to our hospital with complaints of progressing headache, nausea, and vomiting for the last 3 days. She had no significant past medical history. At the time of admission, she was disoriented but able to converse. Computed tomography (CT) scans showed a large hyperdense parietooccipital SDH with a significant midline shift. The hematoma was evacuated immediately (Figure 1). Her post-operative course was uneventful with complete neurological recovery. In consultation with the Department of Pediatrics, further investigations were made to determine the etiology of subdural hemorrhage. Platelet count, prothrombin time, activated partial thromboplastin time, international normalized ratio, bleeding time, and all coagulation factors were normal except for

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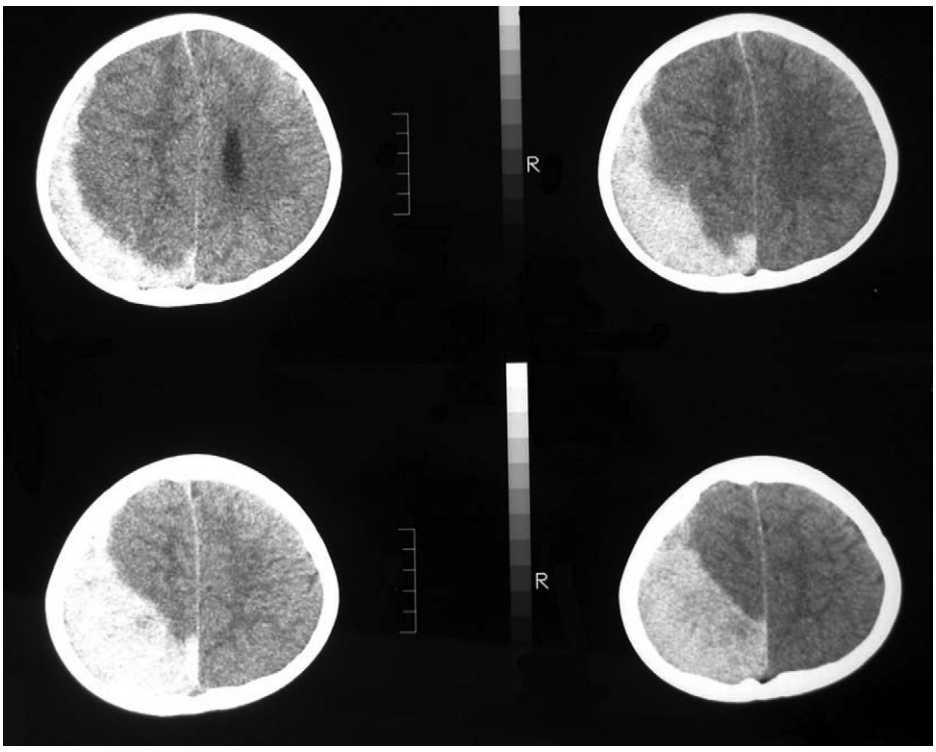

Figure 1. Axial view of the large hyperdense parietooccipital subdural hematoma with a significant midline shift.

F XIII. The diagnosis of F XIII deficiency was made based on the positive clot solubility test using 5 M urea. However, no reliable data about the history of prolonged bleeding from the umbilical stump, a common symptom of F XIII deficiency at birth, were available. The patient was discharged without any neurological deficits, with a treatment of monthly transfusion of fresh frozen plasma as the replacement and prophylactic therapy. There was no residual or recurrent hemorrhage on control CT scans (Figure 2). Ten months after her discharge, the patient was referred to our clinic with a 3-week history of mild headache that had started after a minor head trauma. CT scan revealed a chronic right parietal EDH beneath the craniotomy flap (Figure 3). The patient's neurological functions were intact. The hematological tests showed F XIII levels as low as in her first presentation. A detailed inquiry revealed that the patient had not received fresh frozen plasma for the last 2 months. The EDH was surgically removed while the patient was infused with fresh frozen plasma. Full recovery was achieved, and the post-operative CT scan of the patient demonstrated no residual hemorrhage (Figure 4).

DISCUSSION

Acute SDH occurs in approximately 5–22% of patients with severe head injury and results from bleeding within the subdural space due to stretching and tearing of the bridging veins that traverse the space between the cortical surface and the venous sinuses (13,14). Subdural hemorrhages also can result from injury to the surface of the brain with bleeding from cortical vessels into the subdural space (14). Epidural hematoma is a collection of blood between the inner table of the skull and the dura and occurs in $< 2\%$ of patients admitted with craniocerebral trauma (14). In infants and elderly people, EDHs are rare due to dura that is well attached to the skull, but in young children the dura-to-skull fibrovascular attachments decrease in density as the skull grows. EDHs, therefore, are the most common traumatic intracranial hematoma in young children, with about 3% of those hospitalized with head trauma (5).

Acute spontaneous SDH is extremely rare. Its main etiological factors include anticoagulant therapy, hypertension, cocaine-induced vasculitis, bleeding from intracranial tumors or aneurysms, and bleeding and coagulation disorders (1,3,4). Deficiency of F XIII is a quite rare coagulation disorder and occurs as congenital or in acquired forms (15). The main causes of acquired F XIII deficiency are liver disease, inflammatory bowel disease, disseminated intravascular coagulopathy, Hennoch-Schönlein purpura, and sepsis (15,16). Congenital form is an autosomal recessive disorder of hemostasis, with an estimated prevalence of approximately 1 in 3–5 million individuals (7). In our patient, there were no signs or