

Subdural Hematoma Associated with Immune Thrombocytopenic Purpura in Two Different Clinical Settings

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Abstract

Subdural hematoma (SDH) is a rare complication of immune thrombocytopenic purpura (ITP) the incidence being around 2%.¹ Although SDH usually occurs secondary to trauma, in bleeding disorders it may occur spontaneously. Here we report subdural hematoma in two uncommon settings, one patient with systemic sclerosis developing secondary ITP and consequently subdural hematoma and the other patient with chronic ITP developing subdural hematoma in the inter hemispheric region. In both the settings the patients recovered spontaneously without any surgical intervention.

Introduction

Neurological complications of immune thrombocytopenic purpura (ITP) are mainly intra cerebral hemorrhages which are often fatal and are dependent on the severity of thrombocytopenia. Subdural hematoma is rare in a setting of ITP and only few cases have been documented.¹ Here we report two such interesting cases.

Case Report 1

A 33 year old female, presented to us with headache, severe vomiting, hematuria and diplopia since last 3 weeks. She also complained of menorrhagia since last menstrual cycle, dimness of vision in both eyes since last 9 month. She had a history of skin tightening (mainly facial, peri orbital, both upper and lower limbs proximal as well as distal and in trunk), Raynaud's phenomenon, symptoms suggestive of Gastro esophageal reflux disease (without any history of shortness of breath or joint pain, malar rash and muscle weakness) since last 5 years. She was previously diagnosed to be a case of diffuse cutaneous scleroderma and was under appropriate therapy.

On examination: Patient was conscious and alert. General examination was unremarkable except for mild pallor and the skin tightness as described above. Pulse: 72/minute, BP:130/80 mmHg. She also had diffuse hypo pigmented patches over the thickened skin areas. Her neurological examination revealed bilateral VI nerve palsy, visual acuity was 6/6 but blurring of vision was present. There were no focal neurological deficits, neck rigidity was absent. Fundoscopy revealed bilateral retinal hemorrhages but there was no papilloedema. Other systemic examination including Respiratory system, CVS and GI was normal.

Her investigations revealed: Hb – 8 gm %, TLC – 11,000/cmm, Random blood sugar – 107 mg/dl. Platelet count was 20,000/cmm. Routine urine examination revealed plenty of RBC's. Her renal and liver function tests, PT and APTT were normal. Her anemia was further investigated and was found to be iron deficiency anemia. HIV ELISA, IgM Anti HCV reports were negative. ANA (hep2) was positive with 1: 640 titer speckled pattern. Serum Scl 70 was also strongly positive. Coombs test

was done to rule out Evans syndrome which was found to be negative. Serum electrophoresis was normal. CT scan of her brain revealed bilateral hypo dense lesions in temporo parietal region suggestive of chronic subdural hematoma. Subsequently, a bone marrow study was done to evaluate her low platelet count. It revealed megakaryocytes increased in number and size, usually smooth forms with single nuclei, suggestive of ITP. Antiplatelet IgG antibodies were not done as they have not yet been demonstrated to be important for clinical diagnosis and management decisions, also these assays are very expensive.

Considering the temporal profile of her events, a diagnosis of subdural hematoma secondary to chronic ITP in a background of diffuse cutaneous scleroderma, was made.

Patient was started on Oral prednisone 1 mg/kg body weight in tapering dose along with other supportive therapy. She was also given 20 units random platelet transfusion. After 7 days her platelet count rose to 1,60,000 and neurosurgery consultation suggested conservative management regarding subdural hematoma since patient's headache, vomiting gradually subsided post admission and no other features of raised ICT developed in her hospital course. She was discharged when her platelet count rose to 2, 10,000 and head ache significantly decreased.

Case Report 2

A 45 year old female presented to us with headache of sudden onset localized to the vertex, since last one week. There was no history of vomiting, altered sensorium, seizures or neuro deficits. There was no past history of any chronic headache. But she gave a history of easy bruisability, intermittent purpura in lower limbs and occasional gum bleeding since last 10 years. There was no history of any joint pain, skin rash or fever preceding the episode. She was otherwise healthy.

On examination patient was conscious and alert. General examination was unremarkable. Pulse: 88/ minute, BP: 110/70. Neurological examination revealed plantar extensor on right side. Power in all 4 limbs was 5/5. Other systems including Respiratory system, CVS and GI were found to be normal.

Her investigations revealed: Hb – 11 gm %, TLC – 9,000/cmm, Platelet count was 70,000/cmm, Random blood sugar – 107 mg/dl. Routine urine examination was normal. Her renal and liver function tests, PT, APTT reports were normal. HIV, IgM Anti HCV, ANA (hep2) and Coombs test reports were negative. Serum electrophoresis was normal. CT scan of her brain revealed a subdural hematoma in the falx cerebri with extension in the left tentorium cerebelli which was further confirmed by an MRI Scan considering the very unusual location of the bleed. It showed subacute SDH in the inter hemispheric fissure with the extension

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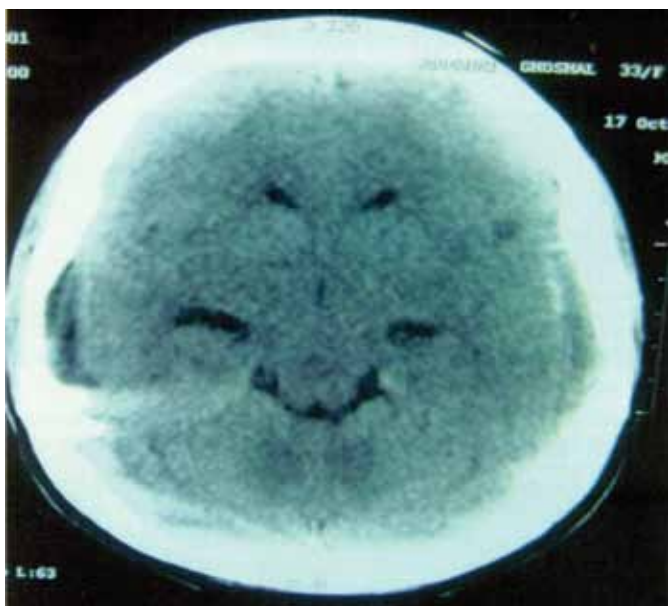


Fig. 1 : CT Brain showing bilateral hypo dense lesions in temporo-parietal region.

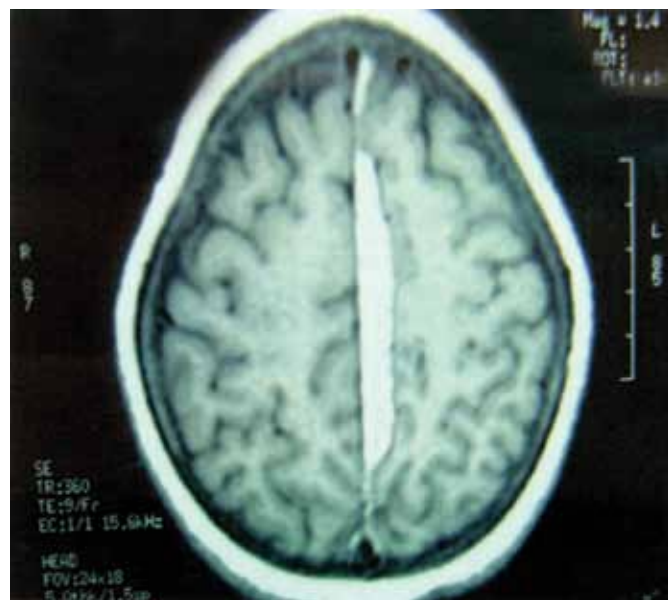


Fig. 2 : MRI brain showing the presence of hyperdense blood interdigitating within the sulci indicating acute subdural hematoma which has collected between the leaves of the falx cerebri

over left parietal cortical sulci. Mild SAH was also seen. The bone marrow revealed features suggesting ITP.

Patient was managed conservatively. She was started on Tab Prednisolone 1 mg/kg body weight with gradual tapering. There was no worsening of her neurological signs and after 2 weeks her platelet count rose to 1,40,000/cmm. Patient came for follow up after 6 weeks. This time she had neither any neurological problems nor any bleeding manifestations and her platelet count was 1,46,000/cmm. Her follow up CT scan showed considerable resolution of the inter hemispheric subdural hematoma.

Discussion

ITP (*first described by Werlhof in 1735*) is an acquired disorder which leads to immune mediated destruction of platelets characterized by low platelet count and normal coagulation studies. Secondary ITP usually accounts for 5–10 percent cases. Some underlying causes are CLL, drugs (e.g. quinine, heparin), HIV, HCV infection, SLE etc. Some autoimmune disorders causing ITP are Myasthenia gravis, Scleroderma, Rheumatoid arthritis, Sjogrens syndrome, Sarcoidosis etc. The most serious and life threatening complication of ITP is intracerebral hemorrhage.

The origin of chronic subdural hematomas are usually traumatic, how ever in patients with a bleeding disorder like ITP, subdural hematoma usually occurs spontaneously without any history of bleeding as was found in the review of ten cases of chronic SDH with ITP by Seckin H et al in *Surgical Neurology* 2006.² The clinical features are mainly headache, hemiparesis, signs of raised intracranial tension, altered consciousness. Neurological signs were reported to occur more frequently in patients younger than 20 years of age.

The mean platelet count of patients with ITP who developed ICH and SDH was found to be 14.3 ± 7.7 and $26.7 \pm 6.1 \times 10^3 / \text{mm}^3$ respectively in the study of 24 patients reported by Lee and Kim in *Neurology* 1998.³ Usually, subdural hematomas occur around the top and side of the frontal and parietal lobes but they may also occur in the posterior cranial fossa, near the falx cerebri and tentorium cerebelli. The development of subdural hematoma usually depends on low platelet levels but the rate

of enlargement of the hematoma is independent of the platelet count. Hence there exists a relatively safe period for pre operative evaluation and treatment in patients with normal neurological findings.

Subdural hematoma in the interhemispheric location is a rare and interesting clinical entity. Analysis of seven cases by Satoh et al in 1986⁴ revealed the following: All of the patients were male between 23 to 74 years of age. The mechanism of the hematoma formation in such region remains unclear, but seemed to be caused partially due to rotational cerebral injuries. Characteristic clinical symptoms were crural hemiparesis or monoparesis. In our report we have described non traumatic interhemispheric bleed in a 36 year old female with no neurological signs and head ache as the only symptom which recovered spontaneously.

The association of ITP with systemic sclerosis is rare and not too many cases are found in the literature although there are several reports of Thrombotic thrombocytopenic purpura with Systemic sclerosis. Leivovici et al⁵ has reported a patient with generalized morphea and thrombocytopenia. The thrombocytopenia responded promptly to corticosteroid and immunosuppressive therapy. Similarly thrombocytopenia has also been reported in localized scleroderma. In a study of 180 cases about hematological abnormalities in scleroderma by Frayha RA et al⁶ it was shown that anemia was detected in 25% and was attributed mainly to chronic inflammatory disease. Interestingly, in this study thrombocytopenia was found to be often a manifestation of microangiopathy. There has also been a report of thrombocytopenia induced by D Penicillamine, used in the treatment of scleroderma. The occurrence of thrombocytopenia in scleroderma suggests an autoimmune mechanism (by the response to steroids and the presence of positive auto antibodies) however non immune mechanisms can not be ruled out. This possible association emphasizes the need to look for hematological disorders in patients with systemic, as well as localized scleroderma.

Usual management strategy is surgical evacuation of the hematoma although in our present two cases as well other cases previously reported by Sreedharan et al 1999¹ and H Seckin et

al 2006² has shown that conservative management also led to spontaneous recovery. Overall the decision depends on the site and size of hematoma, extent of neurological damage involved and pre operative assessment of the patient.

Regarding medical therapy of ITP with or without subdural hematoma, a platelet count below 20,000/cmm is an indication for treatment. Treatment usually is initiated with intravenous steroids, IVIg or a combination of these drugs. Platelet transfusion (preferably single donor) alone may slow down the bleeding but it seldom results in a sustained rise of platelet count due to peripheral platelet destruction.²

After the platelet count has stabilized, an orally administered steroid, such as prednisone (1–2 mg/kg per day) is usually prescribed. However any deterioration of neurological status inspite of adequate treatment of ITP will warrant a repeat neuroimaging (CT or MRI scan) to decide urgent surgical exploration.

As sub dural hematoma is a dreaded and rare complication of ITP, we thought it would be valuable to share our experience in these cases with a successful outcome through medical therapy only. Early diagnosis and prompt, aggressive management of the thrombocytopenia is extremely important.

Abbreviations

ITP – immune thrombocytopenic purpura SDH – subdural hematoma ICH Intracerebral hemorrhage CVS – cardiovascular system GI – gastrointestinal system PT – prothrombin time APTT – activated partial thromboplastin time. HIV – human

immunodeficiency virus HCV – hepatitis C virus Hb – hemoglobin TLC – total leukocyte count ANA – anti nuclear antibody CT – computed tomography MRI – magnetic resonance imaging ELISA – enzyme linked immunosorbent assay.

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