CASE REPORT

Spontaneous Intracranial Haemorrhage in Children with Chronic Immune Thrombocytopenic Purpura

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SUMMARY

Spontaneous intracranial haemorrhage (ICH) is a rare complication of chronic immune thrombocytopenic purpura (ITP) in children. We report four patients with cITP who developed ICH. The latency between onset of ITP and ICH varied from 1-8 years. All our patients were profoundly thrombocytopenic (platelet count of <10 x 109/l) at the time of their intracranial bleed. The presenting features and management are discussed. All patients survived, three had complete neurological recovery while one had a minimal residual neurological deficit.

KEY WORDS:
Chronic immune thrombocytopenic purpura, intracranial haemorrhage, children

INTRODUCTION

Intracranial hemorrhage (ICH) is a potentially fatal manifestation of immune thrombocytopenic purpura (ITP). This complication however, has been described only rarely in children with chronic ITP. We report 4 patients with severe chronic ITP who developed ICH.

CASE REPORT

Case 1

A 12-year-old girl with chronic ITP had recurrent gum bleeding. She presented with a sudden onset of monoparesis of the right upper limb. Clinically, she was pale and had gum bleeding. Cranial computed tomographic scan (CT scan) of brain showed intracranial haemorrhage of the left fronto-parietal region. She was treated with intravenous immunoglobulin (IVIG), methylprednisolone and platelet support. A splenectomy was performed. Following this, she had a full neurological and haematological recovery.

Case 2

A 9-year-old girl was diagnosed with chronic ITP. She did not show any response to steroids or IVIG. On the day of presentation, she complained of sudden onset of unilateral right parietal headache associated with vomiting. She had left 7th cranial nerve palsy and left hemiparesis. CT scan brain revealed a right internal capsule haemorrhage (Figure 1). She was treated with IVIG, methylprednisolone and platelet support. Her platelets normalised but she was left with a residual left hemiparesis and facial nerve palsy.

Case 3

A 17-year-old female was diagnosed with chronic ITP. She was a steroid responder. She presented with petechiae, menorrhagia and gum bleeding for a week. She was pale and had multiple bruises. She was transfused with platelets and packed cells and started on steroids. Two days later, she developed severe headache and neck pain associated with drowsiness and blurring of vision. Examination showed a retinal haemorrhage. CT scan brain showed haemorrhage in left basal ganglia with midline shift. She was treated with IVIG, methylprednisolone, and platelet support. She made a full haematological and neurological recovery.

Case 4

An 11-year-old girl with chronic ITP developed headache and vomiting for a week and right eye strabismus 2 days prior to presentation. Examination showed right 6th nerve palsy with diplopia on right lateral gaze. CT scan brain revealed right fronto-parietal subdural haemorrhage (SDH) (Figure 2). She was treated with methylprednisolone, IVIG and platelet support. The SDH was drained. Following this, the 6th nerve palsy resolved. She underwent splenectomy 3 weeks later. On follow up, her platelet was stable between 30-90x1010/l.

DISCUSSION

Our series consists of 3 patients with intraparenchymal bleed and one with isolated SDH. Subdural haematoma and subarachnoid haemorrhage usually occur as extensions of ICH.[1] Isolated SDH is very rare. All of our patients are adolescent girls. In contrast, Mohsen *et al* found a male predominance.[2] There was no difference in age between the 40 patients with ICH and the 80 case controls (ITP without ICH) in a survey done by Bethan *et al*.[3]

Although trauma has accounted for some reported cases of ICH, most seem to be spontaneous, as seen in our cases. All of our patients had platelet counts <10x109/l. This agrees with most studies who reported median platelet counts <10x10^9/l in most of their cases with ICH. Severe thrombocytopenia appears to be permissive but not sufficient for ICH as higher platelet counts have also been reported. One hypothesis is that other sites of bleeding, such as wet purpura, epistaxis, genitourinary or gastrointestinal haemorrhage in addition to petechiae and ecchymosis in children with ITP, could be a warning sign of impending ICH. A Japanese study reported menstruation as a risk factor in

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Table I: Clinical profile of the patients

Case	1	2	3	4
Age/Sex	12 years Female	9 yrs 6 mo Female	18 yrs Female	11years Female
Platelet at time of ICH (x 109 /L)	8	8	7	6
Latency between onset of ITP and ICH	1 year	1year 4 months	8 years	2 years
Presenting features	Right upper limb monoparesis	Left hemiparesis and facial nerve palsy	Headache, neck pain and blurring of vision	Headache, vomiting and right 6th nerve palsy
Other bleeding symptoms	Ecchymosis and gum Gum bleeding	Gum bleeding	Gum bleeding, menorrhagia	bleeding
Site of ICH	Left fronto- parietal	Right internal capsule	Left basal ganglia	Right fronto- parietal SDH
Management of ICH	Steroids/IVIG/Platelets, splenectomy	Steroids /IVIG/Platelets	Steroids /IVIG/Platelets	Steroids, IVIG, platelets and craniotomy: burrhole and drainage, splenectomy
Outcome	Complete neurological recovery	Residual left hemiparesis and facial nerve palsy	Complete neurological recovery	Complete neurological recovery

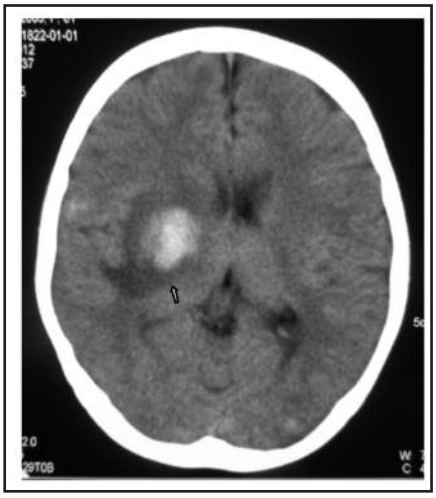

Fig. 1: (Case 2): CT scan brain showing right internal capsule haemorrhage and surrounding oedema.

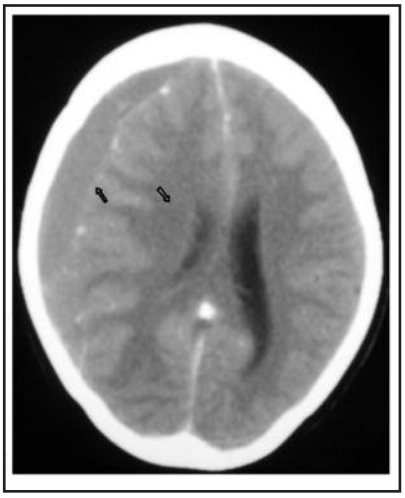

Fig. 2: (Case 4): CT scan brain showing right fronto-parietal subdural haemorrhage with mass effect (midline shift).

ICH [4] and the Egyptian study found 70% of their cases had associated bleeds [2]. All of our patients had mucosal bleeds and one of them had heavy menstruation prior to ICH.

ICH in ITP is a medical emergency. Our patients showed variable responses to both IVIG and steroids during the acute and chronic phases of ITP: steroid partial response (case 1), IVIG and steroid response (case 2) and IVIG or steroid non-response (case 3 and case 4). Despite that, all our patients showed good platelet responses to both methylprednisolone and IVIG in the management of ICH. Corticosteroids have a direct effect on platelet levels and also help to reduce the oedema that accompanies ICH.

Emergency splenectomy is employed in cases of ITP refractory to conservative therapy or if life-threatening haemorrhage occurs. The response of platelet counts to splenectomy varies from 70-90%. Two of our patients underwent splenectomy and showed responses in platelet counts. One of our patients required neurosurgical intervention as she developed signs of neurological deterioration and there was mid-line shift. SDH is often managed surgically because of the poor outcome associated with conservative management and a higher chance of recurrence. The risk of perioperative bleeding and potential worsening of intracranial haemorrhage in patients with ITP limit the role for surgery. [1] The mortality rate of ICH varies from 12.5-25%. [2-4] Bethan *et al* reported that 25% of their patients had neurological sequelae. [3] There was no mortality in our series and only one of our patients had a residual 7th facial nerve palsy and hemiparesis.

CONCLUSION

Our cases demonstrate that successful treatment of ICH in ITP can occur, provided early diagnosis and prompt, aggressive management is instituted. It should be emphasised to patients that as long as severe thrombocytopenia persists, the risk of ICH remains, hence regular follow-up and early presentation to hospital is advised.

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