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A Case Report On Severe Haemophilia-A Presented With Right Sided Inter Muscular Haematoma Secondary To Fall

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Abstract

Background:- The term Haemophilia comes from a student of Zurich university, Friedrich Hopff and his professor, Dr Schonlein, who came up with the term 'Haemorrhaphilia' which became "Haemophilia" in 1828.

Case Presentation :- A 3 year old male patient came to the hospital with acute history of fall while playing with kids in farm. Patient developed swelling over abdomen and fever and were no complaint of vomiting, ENT bleed or loss of consciousness.

Conclusion :- The patient was initially treated with PCV transfusion and transferred to other hospital for the infusion of high quality factor VIII supplementation, which helped save valuable time and improved his prognosis.

Key words :- Haemophilia A, Haematoma , Factor VIII deficiency, PCV, Mucosal Haemorrhage

Introduction

Haemophilia A is a rare coagulopathy caused by inhibitors of blood coagulation factor VIII. This condition is characterized by spontaneous and post traumatic subcutaneous bleeding and mucosal hemorrhage[1]. It affects 1 to 1.5 per 1 million people each year and affects both men and women. Up to 50% of acquired haemophilia cases could be idiopathic and is also associated with condition such as malignancy, diabetes, pregnancy, infection and autoimmune disorder[2]. Haemophilia is X linked recessive benign haematological disorder associated with bleeding manifestation and coagulation abnormalities[3]. Here, The inter muscular hematoma is internal bleeding between muscle fascia and interstitial spaces when muscle and fascial vessels are damaged. An intramuscular hematoma usually causes greater loss of function and more persistent swelling. Typically, bruising and swelling caused by a collection of blood appear distally to the damaged area 24- 48 hours after the injury due to gravity[4].

Case report

A 3 year old male patient came to the hospital with acute history of fall while playing with kids in farm. After 2 days of fall , patient developed swelling over abdomen and fever since past 1 day. There were no complaint of vomiting, ENT bleed or loss of consciousness .

Past history

The patient had similar complaint of development of swelling over various body parts after minimal trauma which usually regressed spontaneously in the past and seeked no treatment for the same. The guardian of patient decided to seek medical help this time due to presence of fever along with swelling over abdomen following trauma. Patient immunization was up to date and there's absence of major medical illness

General examination

On examination patient was conscious, crying , playful and well oriented to place .

General examination revealed no abnormal findings in the patient.

Local Examination

Inspectomy findings revealed slit like umbilicus with a (15 × 15) cm sized swelling in left flank region extending from the right rib medially in left flank laterally.

Swelling does not move with respiration with stretched, shiny, dark res to blackish skin over it. Palpation revealed presence of raised temperature, tenderness and stony consistency. Percussion revealed dull more and bowel sounds were audible on auscultation.

USG Abdomen + Pelvic

USG Abdomen + Pelvic was suggestive of large localized hematoma involving right lower anterior chest and abdominal wall with intra muscular extension within rectus sheath. No use evidence of intra peritoneal extension.

CT Scan

CT Scan of abdomen found swelling with large hypodense collection noted in inter muscular plane of abdominal wall on right side extending from right hypochondriac region to right iliac fossa region. Surrounding fat stranding is noted. Size of collection is about 7.7*2.4*9.0cm=vol77cc.

Laboratory Reports

Time		**13:00**	**9:18**	**19:35**
Date	**14-Mar**	**15-Mar**	**16-Mar**	**16-Mar**
Hb		4.0	8.1	
RBC		2.35	3.64	
WBC	18	18	17.4	15.4
Neutrophils		72		
Lymphocytes		24		
Monocytes		3		
Eosinophils		1		
Basophils		0		
Platelets		418	224	263
MCH		17	22.3	21.6
MCHC		2708	30.2	31
MCV		67.3	73.6	69.8
Hematocrit		14.4	26.8	32.3
RDW		42		58.8
PT test			14.8	12.9
INR			1.26	1.09
APTT test		38	51	52.3
AST (SGOT)	17			
ALT (SGPT)	17			
Ser. ALP	134			
Bili. Total	0.37			
Bili. Direct	0.1			
Bili. Indirect	0.27			
Total Protein	6.79			
Albumin	3.2			
Gobulin	3.59			
A/G Ratio	0.89			
CRP	92.2			
Na+	135			138
K+	3.7			3.8
Ca				1.27
Cl	101			
Ser. Creatinine	0.34			
Factor VII		<1%		
Factor IX		72.1%		
Fibrinogen		411		

Treatment Chart

Drug	Dose/Route/Frequency
IV Fluids	IV/60 ML per HR
Inj Augmentin	30 mg/IV/1-1-1
Inj Metronidazole	30 ML/IV/1-1-1
Inj Tramadol	1-2 mg/Kg/IV/4-6 Hrly
Inj Paracetamol	10 mg/kg/iv/sos
Inj Ondensetron	2 mg/IV/1-1-1
Inj KCL	10 mEq/Twice a day
Inj Pantoprazole	40mg/IV/1-0-1
Thrombophobe	20 mg/Tropical/BD

Discussion

The patient is diagnosed with Haemophilia A with right sided intermuscular hematoma secondary to fall. Haemophilia A is a X linked bleeding disorder resulting in a deficiency of coagulation factor VIII or coagulation factor IX Haemophilia B due to gene mutation in the respective clotting factor genes [5]. Haemophilia is classified as severe (factor level < 0.01 IU/ml), moderate (factor level < 0.05 IU/ml), and mild (factor level ≥ 0.05 and < 0.4). Haemophilia A, which occurs in approximately 1 in 5000 live male births, is more common and more likely to be severe, and morbidity is higher among males than among females.[6]

Patient was admitted in paediatric ICU under joint care with surgery department also, kept NBM, IV fluids started and blood investigation sent and as patient had a large abdominal haematoma. CT Scan was done which was s/o above mentioned findings. Empirically antibiotic (Inj Augmentin and Inj Metronidazole) were started . As blood reports were s/o Low Hb and deranged coagulation profile so 2 units of PCV and 1 unit of FFP was given. On next day blood investigation sent which were s/o hypokalemia so acute correction was given and continued in maintainance dose and coagulation profile which was sent which was again deranged so again Inj FFP was transfused and factor VIII and IX were sent which were s/o severe deficiency of Factor VIII (<1%) , so 4 units of cryoprecipitates were transfused . Relatives have been counselled regarding this bleeding tendency , need to transfuse specific factor as per deficiency.

As patient was vitally stable, taking and tolerating feeds well so transferred to paediatric ward for further observation.

Conclusion

The patient was initially treated with PCV transfusions and was transferred to other hospital for the infusion of high-quality factor VIII supplementation, which helped save valuable time and improve his prognosis. Therefore, the provision of factor VIII is a very important factor in the treatment of hematoma patients with a medical history of haemophilia A. We recommend monitoring for the coagulation function and also determining the medical history when the patient is admitted, especially in the emergency department.

Abbreviations :-

1.	PCV	Packed cell volume
2.	NBM	NIL By Mouth
3.	FFP	Fresh Frozen Plasma
4.	Hb	Haemoglobin
5.	ICU	Intensive Care unit
6.	APTT	Activated Partial Thromboplastin clotting time
7.	MCH	Mean Corpuscular Haemoglobin
8.	MCHC	Mean Corpuscular Haemoglobin concentration
9.	MCV	Mean Corpuscular Volume
10.	RDW	Red cell Distribution Width
11.	AST	Aspartate Aminotransferase
12.	ALT	Alanine Aminotransferase

Declaration of patient consent

The authors certify that they have obtained appropriate patient consent form. The patient's parents understand that their child name and initials will not be published and due efforts will be made to conceal his identity.

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Conflict of interest

All authors declare that they have no conflict of interest.

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