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Personal Practice

Screening for Bleeding Tendency in Suspected Child Abuse

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Abstract Children who have been physically abused or victims of Munchausen syndrome by proxy may occasionally be complicated by coagulopathy. Specific bleeding diathesis presenting as suspected child abuse is rarely reported in the medical literature. More commonly, however, screening tests are used to exclude an underlying bleeding tendency in suspected cases of child abuse. Under most circumstances, ordering a platelet count, prothrombin time and activated partial thromboplastin time would be sufficient for clinical differential diagnosis or medico-legal purposes. Bleeding time may be included if a platelet function defect is suspected. However, the value of a thorough history and comprehensive physical examination cannot be overemphasized. The identification of a bleeding tendency does not exclude child abuse. A maltreated child who presents with bruises that are clearly indicative of an inflicted injury does not require coagulation screening unless indicated otherwise.

Key words Child abuse; Haemostasis; Coagulopathy; Thrombocytopenia

Introduction

In Tuen Mun Hospital, an average of more than one child are admitted every week for evaluation of child abuse.[1] Over 50% of these children are admitted because of suspicion of physical abuse, either alone or in association with other forms of maltreatment such as neglect, sexual or psychological abuse.[2] Bruises or haematomas, the cutaneous manifestation of extravasated red cells from damaged blood vessels, are the most noticeable sign of non-accidental injury. The injury may be produced from force, with or without the use of an instrument, from heat, or from intoxication. In the majority of cases, the diagnosis of physical abuse is obvious from the clinical history and physical findings.[3]

In the evaluation of suspected physical abuse when cutaneous bruises are the major clinical features, the major differential diagnosis is to distinguish inflicted injury from unintentional trauma. Because of the occasional reports that children with bleeding tendency may be confused with victims of child abuse, clinical evaluation becomes necessary whenever there is a suspicion of an underlying medical problem. Screening studies for coagulation and bleeding diathesis are therefore required for a number of reasons.[4] (1) The caretaker often alleges a history that the victim bruises easily. (2) A medico-legal documentation is required when the cause of bruises is not apparent. (3) Bleeding diathesis may just be one of the manifestations of maltreatment. (4) The history or physical finding suggests an underlying coagulopathy. However, not all physically abused children require coagulation screening.

The present review will be devoted to the discussion of the basic approach on handling the issue of coagulation disorder among children with suspected maltreatment, the use of coagulation screening, and a review of bleeding disorders mistaken as child abuse in the medical literature.

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The Basic Approach on the Use of Coagulation Screening

The principle underlying the clinical approach on the use of coagulation screening in a suspected case of child abuse is similar to other children.[5] A thoroughly taken history would give clues to whether the child has any manifestations of bleeding tendency (Table 1). An underlying bleeding diathesis would be extremely unlikely

Table 1 Important historic features suggestive of bleeding tendency

Excessive/prolonged swelling after immunization
Prolonged bleeding after circumcision or other operation
Unexplained muscle or joint swellings
Recurrent epistaxis or gum bleeding
Recurrent bloody diarrhoea or haematemesis[20]
Family history of bleeding disorders and menorrhagia
Consanguinity

if the patient has undergone surgery without excessive, prolonged or delayed haemorrhage. Operations that are commonly encountered in childhood include circumcision, tonsillectomy, dental extraction and appendicectomy. An uncomplicated craniotomy for removal of blood clots without haemostatic replacement therapy practically excludes any significant coagulopathy.

The evaluation of a child referred for suspected abuse should always include a comprehensive physical examination.[6] In particular, the distribution, discoloration, size, shapes, and surrounding soft tissue injuries of all bruises or haematoma should be noted. Scars from previous injuries are often helpful clues. Congenital pigmented lesions such as Mongolian spots can be easily differentiated from bruises.[7] However, confusion may arise if they occur in the limbs in isolation or when the child is not adequately exposed. Diseases with vasculitis (e.g. Henoch Schonlein purpura[8]) or vascular fragility (e.g. Ehlers-Danlos syndrome[9]) may also present with bruises. The purpuric rash associated with Henoch Schonlein purpura has a characteristic dependent distribution and papular appearance, and patients with Ehlers-Danlos syndrome can be distinguished by other associated features such as hyperextensibility and paper tissue scars.

Many of the inflicted injuries produce characteristic patterns that are indicative of the mode of injury.[6] These patterns may be modified according to the amount of force used, the actual direction of the impact, and the contour of the region of the body being hit. Slapping and hitting with a stick or a rod will leave parallel tracks of bruises. These tracks of bruises occuron both sides of, rather than the actual point of, impact as it is those parts of the skin that are subjected to maximal tensile forces. Beating with a loop of electric cord or a clothes hanger will produce curved or curvilinear marks that correspond to the shape of the weapon. Also, telltale signs of bruising can be found after beating with a belt depending on whether the child is injured by the buckle or the leather strap. When such patterned bruises are evident on examination, they are clear signs of inflicted injury. Coagulation screening is not indicated unless there are suggestive clues from the history.

Even when the bruises are not typical of a recognized pattern, the older child can often tell how the injuries were sustained when asked. Sometimes the perpetrator may admit his beating and confirm the child's account of injury. If this information is available, coagulation screening may be omitted. However, when the abuser or the victim is not willing to disclose, or when a legal procedure is probable, screening for bleeding tendency will be indicated.[4]

Screening for Bleeding Tendency

The purpose of conducting coagulation screening in suspected cases of child abuse is to detect any significant bleeding diathesis as suggested by the history or physical examination, and to exclude common illnesses that may be confused with non-accidental injury. The tests are directed at both the intrinsic and extrinsic pathways of the coagulation cascade and platelet haemostasis. Complete blood counts (CBC) including platelet count, prothrombin time (PT), and activated partial thromboplastin time (APTT) are recommended as the first screening tests.[10,11] Liver and renal function tests, and measurement of bleeding time (BT) are useful if indicated. Thrombocytopenia, common coagulopathies such as haemophilia A or B, and vitamin K deficiency bleeding are easily ruled in or ruled out. Von Willebrand disease, estimated to be present in 1% of the population, can be detected by a prolongation of APTT and/or the bleeding time. More specific assays such as ristocetin cofactor and factor VIII coagulant activities are required to confirm the diagnosis. Measurement of bleeding time may also be indicated if there are other concerns of qualitative platelet disorders. These may be congenital as in the case of Glanzmann's thrombasthenia, or may be acquired in the case of salicylate ingestion. Examination of platelet size and morphology, platelet function tests, and analysis of platelet glycoproteins will be required for confirmation. The use of laboratory tests in this regard is summarized in Table 2. Specific reference describing the use of each of the laboratory test is available.[12]

When the CBC, PT, APTT and BT are normal and a

Table 2 Coagulation screening in suspected cases of child abuse

1. Detailed history and physical examination (see Table 1)
2. First line screening :
Complete blood counts (CBC)
Prothrombin time (PT)
Activated partial thromboplastin time (APTT)
3. Bleeding time, if platelet function disorder suspected
4. Renal and liver function tests, if indicated
5. Specific factor assay or platelet function tests

bleeding diathesis is strongly suspected because of recurrent significant bleeding and/or a positive family history, further evaluation for deficiencies of factor XIII, α2 antiplasmin or plasminogen activator inhibitor type 1 should be pursed.[5] The clinical and laboratory features of these disorders are summarized in Table 3.

Table 3 Significant inherited bleeding disorder with normal coagulation screening tests (normal PT, APTT, and platelet count)

Disorder	Frequency	Mode of inheritance	Clinical manifestation	Laboratory diagnosis
**Von Willebrand disease*[21,22] (Types 1 & 2)	from 125 / million up to 1%	Autosomal dominant (rare recessive forms)	mucocutaneous bleeding (epistaxis, gum bleeding, easy bruising, menorrhagia, bleeding after dental extraction)	vWF:Ag, ristocetin cofactor activity, factor VIII activity, ristocetin-induced platelet aggregation, multimer analysis, mutation detection in Type 2
Factor XIII deficiency[23]	very rare (> 100 patients reported)	Autosomal recessive (parental consanguinity common)	bleeding from umbilical stump, delayed and repeated bleeding from wounds, intracranial bleeding, spontaneous abortion	clot solubility in urea or monochloracetic acid, immunological assay of FXIII subunits
a_2-antiplasmin deficiency[24]	very rare	Autosomal recessive	prolonged bleeding and bruising after minor trauma, spontaneous haemarthrosis	α_2-antiplasmin amidolytic assay
Plasminogen activator inhibitor-1 deficiency[12]	very rare	Autosomal recessive	Recurrent bleeding after surgery or trauma	shortened euglobulin lysis time assay for PAI-1 antigen and activity
**Platelet dysfunction*[25]				
Glanzmann's thromboasthenia	rare	Autosomal recessive	mucocutaneous bleeding, menorrhagia	platelet aggregation test, platelet glycoprotein analysis
Storage pool disease	rare	Autosomal recessive (some dominant forms)	mucocutaneous bleeding, associated defects (e.g. albinism in Hermansky -Pudlak syndrome)	platelet aggregation test, platelet nucleotide assay, electron microscopy
Defects in TxA_2 generation and signal transduction	very rare	Variable	mucocutaneous bleeding (mild)	platelet aggregation test, specific enzyme assay and metabolic test

* Conditions with prolonged bleeding time.

Bleeding Disorders Presenting as Suspected Physical Abuse

Bleeding disorders may occasionally be confused with child abuse, but they rarely masquerade as child maltreatment to the experienced paediatrician (Table 4). In a series of 2,578 cases evaluated by the child abuse team in Leeds, only five (0.2%) children were found to have coagulopathies including idiopathic thrombocytopenic purpura, haemophilia A, and vitamin K deficiency bleeding of the newborn.[13] Single cases of Glanzmann's thrombasthenia, haemophilia A, and acquired inhibitors to factors VIII and IX were reported in which the affected children were initially thought to be victims of child abuse.[14,15] However, the clinical histories and physical findings in these cases were sufficiently indicative of the underlying bleeding diathesis. Specific tests that followed led to the correct diagnosis.

In a review of conditions mistaken for child abuse, Bays quoted another seven examples of "occult" coagulopathies including haemophilia, acute lymphoblastic leukaemia, rodenticide-induced coagulopathy, two cases of vitamin K deficiency secondary to cystic fibrosis, and two cases of disseminated intravascular coagulation complicating meningitis.[10] Harley also reported recently two cases of idiopathic thrombocytopenic purpura and one case of haemophilia B who were initially though to have been maltreated.[16] Although the history might be non-specific in some of these cases, the coagulation screening as suggested in the previous discussion was sufficient to pick up these abnormalities.

It should be emphasized that the finding of a coagulation defect does not exclude the diagnosis of child abuse. Prolongation of PT or APTT is not uncommon among victims of the shaken baby syndrome, presumably as a complication of the underlying brain injury. Of 101

Table 4 Reported cases of bleeding disorders mistaken as child abuse

Reference	Sex/Age	Bleeding disorder	Case description
10	M/10 m	Haemophilia A	Widespread bruises of different ages; Old fracture of clavicle
	M/4 m	Vitamin K deficiency & Cystic fibrosis	Recurrent bruises, petechiae, failure to thrive, and anaemia
	M/1 y	Vitamin K deficiency & Cystic fibrosis	Generalized bruises
	F/2 y	ALL	Recurrent bruises for 1 month; Died with widespread leukaemic infiltration and a haemoglobin level of 1.8 g/dl
	M/4 y	Meningitis & DIC	Multiple bruises, fever, lethargy and death
	F/2 y	Meningitis & DIC	Bruises on cheek, thigh, fever and death
13	M/3 y	Haemophilia A	Multiple bruises
	Not mentioned	3 cases of ITP	Widespread bruising
	Not mentioned	HDN	Not mentioned
14	M/10 m	Haemophilia A	Recurrent spontaneous bruises and ecchymosis, epistaxis and oral mucosal bleeding
	F/9 m	Glanzmann's disease	Recurrent bruises and epistaxis
15	F/1 y 7 m	Acquired inhibitor to Factors II, VIII & IX	Spontaneous bruises and ecchymosis for 2 weeks after a diarrhoeal illness
16	M/5 y	ITP	3-day history of unexplained bruises
	F/2 y	ITP	2-day history of unexplained bruises
	M/8 m	Haemophilia B	Insidious onset of unexplained bruises, especially when the child attempted to walk

Abbreviations: ALL, acute lymphoblastic leukaemia; DIC, disseminated intravascular coagulation; HDN, haemorrhagic disease of newborn; ITP, idiopathic thrombocytopenic purpura.

children with parenchymal brain injury studied by Hymel et al.,[17] 54% had mild prolongation of PT, and 24% had prolongation of APTT. Deliberate poisoning with rodenticide (superwarfarin) has been reported as Munchausen syndrome by proxy.[18] However, malicious intent may be difficult to prove in extremely unkempt living environment.[19] In their elegant study of 50 children with suspected abuse, O'Hara and Eden found mild, transient abnormalities of coagulation in five cases. They also diagnosed platelet aggregation disorder and von Willebrand disease in two other patients, respectively. However, the diagnosis of physical abuse was obvious from the history and social enquiry in these children in spite of the coagulation defect.[15]

Summary

It is important for the paediatrician handling suspected cases of child abuse to recognize the occasional patient with an underlying bleeding diathesis. It is equally important that a bleeding tendency be reasonably excluded when the occurrence of bleeding is unexplained. Every child who is a suspected victim of child maltreatment should have a thorough history and detailed physical examination to delineate the account and characteristics of the injuries. In this respect, the general paediatrician is well qualified to handle these children in their practice. Laboratory investigations should be used judiciously and haematologist should be consulted when there are uncertainties in the use or interpretation of tests. It should be recapitulated that physical child abuse is often a clinical diagnosis. The finding of a coagulation abnormality is by no means exclusive of an abusive incident.

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