Acute Abdominal Pain in Sickle Cell Disease

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ABSTRACT

A prospective study of paediatric patients with sickle cell disease (SCD) admitted to Qateef Central Hospital (QCH) because of abdominal pain was carried out between July 1990 and October 1992. Out of 59 admissions, 53 had acute abdominal painful crises (APC) and 6 patients had acute surgical abdomen (ASA). History of abdominal pain was found in 66% of APC. Vomiting was found in 66.7% of ASA and 26% of APC. Diffuse abdominal pain was found in all ASA and 11.3% of APC. Rigidity, involuntary guarding and decreased bowel sounds were found in 66.7% of ASA and 9.4% of APC. Rebound renderness and increased bowel sounds were found only in ASA (33.4%). Accompanying painful bone crises was found in 56.6% of APC and in 33.3% of ASA. Past history of abdominal pain, diffuse abdominal pain and accompanying painful bone crises were features of APC. On the other hand, signs of peritoneal irritation were features of ASA.

Sickle cell disease (SCD) is one of the commonly inherited disorders in the Eastern Province of Saudi Arabia[1], and has variable clinical course. Acute abdominal pain is one of the common problems in patients with SCD which has been known since the early description of Washburn[2]. At Qateef Central Hospital 10.7% of paediatric patient admissions with SCD were due to acute abdominal pain (unpublished data). The differentiation of this pain from acute surgical abdomen remains a challenging clinical problem for both the paediatrician and the paediatric surgeon. This difficulty is further complicated by the administration of narcotic medications which may mask the progression of signs and symptoms of acute surgical abdomen.

The aim of this study is to delineate the pattern of clinical features, causes, and course of paediatric patients with SCD presenting with acute abdominal pain and analyse this in relation to clinical and laboratory variables.

METHODS

All the paediatric patients with SCD admitted to QCH with acute abdominal pain from July 1990 to October 1992 were studied prospectively. The patients were evaluated by both a paediatrician and a paediatric surgeon. The diagnosis of SCD was confirmed by a positive sickling test and haemoglobin electrophoresis[3] using Helena Laboratories Super Z electrophoresis kit. A detailed history was obtained from each patient including past history, site of abdominal pain and associated musculoskeletal pain. A thorough physical examination was done. The following investigations were obtained: complete blood count (CBC) and differential count, urine analysis and culture, stool analysis, chest x-ray, abdominal x-ray and ultrasound. Low grade fever was defined as a temperature ranging between 37.5°C and 38.9°C[4-5]. High grade fever was defined as a temperature greater than 38.9°C[4]. The unpaired t-test was used to compare the blood test values in patients with simple abdominal crisis, complicated abdominal crisis and acute surgical abdomen.

RESULTS

During the study period, 38 patients with SCD had 59 admissions with acute abdominal pain. Of these, 24 were males and 14 were females. Their age ranged between 2½ to 12 years (mean 6.4).

Of these, 6 were found to have acute surgical causes; acute appendicitis[2], intestinal obstruction[2], acute cholecystitis[1] and salmonella splenic abscess[1]. Six other admissions of acute abdominal crisis were found to be

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Table 1
Site of localised abdominal pain

Site	*APC* *No*	*ASA* *No*
Epigastric	2	–
Rt. Hypochondria	–	2
Lt. Hypochondria	1	1
Umbilical	2	1
Rt. Iliac Fosa	–	2
Hypogastrium	1	–

associated with acute chest syndrome[4], acute splenic sequestration[1], priapism[1] and salmonella septicaemia[1]. Gall stones were detected by ultrasound in one patient and Giardiasis were isolated in another.

Past history of abdominal pain was found only in patient with APC. This was obtained from 35 admissions (66%). On the other hand vomiting was a frequent presentation (66.7%) in those with ASA when compared to 26% of those with APC. Diffuse abdominal pain was found in 88.7% of APC admissions. Localised abdominal pain was found in all patients with ASA while only 11.3% of admissions with APC had localised abdominal pain. The sites of localised abdominal pain are shown in table 1.

Associated painful bone crises were found in 56.6% of admissions of APC and only in 33.3% of ASA admissions. The sites of painful bone crises are shown in table 2.

Temperature was normal in 56.6% of APC admissions and in 50% of ASA admissions. Low grade fever was found in 41.5% of APC admissions and in 50% of ASA admissions, while high grade fever was found in one patient with APC.

Table 2
Associated painful bone crisis

Site	*APC*		*ASA*	
	No	*%*	*No*	*%*
Limbs	19	35.9	–	–
Joint	2	3.8	–	–
Chest	1	1.8	–	–
Back	15	28.3	2	33.3

Table 3
Abdominal Examination

	APC		*ASA*	
	No	*%*	*No*	*%*
Tenderness	53	100	6	100
Distension	20	37.7	3	50
Rigidity	5	9.4	4	66.7
Guarding	5	9.4	4	66.7
Rebound Tenderness	–	–	2	33.4
Bowel Sounds	5	9.4	4	66.7
Bowel Sounds	–	–	2	33.7

The results of abdominal examination are shown in table 3. Rectal examination revealed tenderness in 50% of ASA admissions while only one patient of APC admissions had tenderness on rectal examination.

Haemoglobin, white blood counts and differential counts were compared between patients with uncomplicated APC and those with complicated APC but there was no significant difference apart from band counts (Table 4). These values were also compared between those with uncomplicated APC and those with ASA. There was no significant difference between the two except for band counts (Table 5).

Table 4
Comparison of blood tests between uncomplicated complicated APC

	Uncomplicated APC	*Complicated APC*	*P-Values*
No. of admissions	47	6	
Mean HB ± SD	8.4 ± 1.7	7.6 ± 3.1	NS
Mean WBC ± SD	13.8 ± 5.8	14.8 ± 6.8	NS
Mean polys ± SD	60.3 ± 15	58.1 ± 11.6	NS
Mean bands ± SD	2.9 ± 2.8	10.8 ± 15.1	S

NS = No significant difference
S = Significant difference

Table 5
Comparison of blood tests between uncomplicated APC and ASA

	Uncomplicated APC	*ASA*	*P-Values*
No. of admissions	47	6	
Mean HB ± SD	8.4 ± 1.7	9.8 ± 1.96	NS
Mean WBC ± SD	13.8 ± 5.8	13.6 ± 5.7	NS
Mean polys ± SD	60.3 ± 15	63.3 ± 12.5	NS
Mean bands ± SD	2.9 ± 2.8	6.8 ± 2.2	S

NS = No significant difference
S = Significant difference

All patients received intravenous fluids and simple analgesia. Strong analgesia was required by 9 patients (19.2%) of APC admissions and 3 (50%) patients of complicated APC and 6 (100%) patients of ASA admissions. Blood transfusions were required in 4 admissions (8.5%) of uncomplicated APC, 6 admissions (100%) of complicated APC and in 2 admissions (33.3%) of ASA. The mean duration of hospitalisation was 4.5 days in uncomplicated APC (range 2-9), 9.8 days in complicated APC (range 7-14) and 11 days in ASA (range 3-17).

DISCUSSION

Acute abdominal crisis is a common complication of SCD, but there are other causes of acute abdominal pain in these patients. The distinction between acute abdominal pain of SCD origin and other causes may be difficult[6], but it is important to detect those conditions which may require surgical intervention. Delayed surgical interventions in patients with ASA may lead to further complications, while inappropriate laparotomy during APC may result in avoidable morbidity[7] and mortality[8].

The clinical features of APC may stimulate any abdominal pain, but in most instances differentiation is possible. We found the majority of patients with APC had no vomiting or signs of abdominal rigidity, involuntary guarding or rebound tenderness, which is consistent with findings of others[9,10]. They usually have diffuse abdominal pain, past history of APC and accompanying painful bone crises. On the other hand the majority of patients with ASA had vomiting, localised abdominal pain and peritoneal irritation.

Fever was not found to be a good indicator of serious illness. Low grade fever was found in less than 50% of APC which is similar to other reports[11].

Haemoglobin, white blood count (WBC) and differential WBC were studied but no significant difference between uncomplicated APC and either complicated APC or ASA could be found except for band counts. There was no significant difference in band count when we compared between complicated APC and ASA. This may suggest that increase in band count may suggest serious illness rather than uncomplicated APC. This is similar to other previous reports[12].

Strong analgesia was required for all patients with ASA, 50% of complicated APC and less than 20% of uncomplicated APC. This may suggest that the more severe abdominal pain is more likely to be ASA or complicated APC.

CONCLUSION

Acute abdominal pain with SCD is a challenging clinical problem. It may be extremely difficult to differentiate between APC of SCD and ASA. Past history of abdominal pain, diffuse pain and accompanying painful bone crisis suggest abdominal crisis. On the other hand, localised abdominal pain and signs of peritoneal irritation may suggest acute surgical abdomen, but it should be emphasised that the most effective way of management of these patients is a combined clinical evaluation by an experienced paediatrician and an experienced paediatric surgeon who are also experienced in SCD.

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