malignancy and congestive heart failure as a contributing factor to the existing hepatomegaly. However, the recognition of GSD was delayed in our patient because all symptoms were initially attributed to underlying SCA. Both SCA and GSD could present with repeated infections, failure to thrive, hepatomegaly with abnormal liver enzymes and cardiomegaly. Therefore, when symptoms change in SCA or acquires an atypical clinical course a concurrent illness should be considered in the differential diagnosis; as early recognition of a co-existing disease such as GSD is important to institute appropriate therapy and prevent potential complications.[2] The clinical presentation, biochemical derangement and liver biopsy were supportive of GSD; in addition, the absence of neutropenia excludes GSD-1b[3] and the presence of lactic acidemia and hyperuricemia makes GSD type III a remote possibility.[4] Since enzymatic assay was not available in our institute, our management plan was directed towards the most common form GSD-1a, in addition to conventional management of sickle cell disease.

In conclusion, since SCA is commonly seen by pediatricians, it is important to have a high index of suspicion when a common disorder has an atypical presentation in order to avoid preventable complications.

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References

1. Schubert TT. Hepatobiliary system in sickle cell disease. ***Gastroenterology*** 1986; 90: 2013-2021.
2. Rake JP, Visser G, Labrune P, Leonard JV, Ullrich K, Smit GP. Glycogen storage disease type I: diagnosis, management, clinical course and outcome. Results of the European Study on Glycogen Storage Disease Type I (ESGSD I). ***Eur J Pediatr*** 2002; 161 Suppl 1: S20-34. Epub 2002 Aug 22.
3. Bashan N, Hagai Y, Potashnik R, Moses SW. Impaired carbohydrate metabolism of polymorphonuclear leukocytes in glycogen storage disease Ib. ***J Clinic Invest*** 1988; 8: 1317-1322.
4. Amin AS, Kasturi L, Kulkarni AV, Ajmera NK. Glycogen storage disease type III. ***Indian Pediatr*** 2000; 37: 670-673.

Omphalitis and necrotizing fasciitis in neonates

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Although, the incidence of infective umbilical cord /periumbilical tissue has decreased in the developing countries,[1-3] but a spectrum of complication related to infection of unhealthy umbilical stump including neonatal omphalitis (NO) and necrotizing fasciitis (NF) is still seen. We present the experience of these infections during the last 5 years at the Royal Hospital (RH), Muscat, Sultanate of Oman the only tertiary center with facility of pediatric surgery. During the recent years (January 2000 to January 2004), a total of 15 cases with the diagnosis of NO/NF were admitted to RH. Out of the 15 cases, 8 had NO while 7 had NF. The differentiation is made according to the severity[3] (Category 1: Umbilical discharge/ malodorous unhealthy umbilical stump, Category 2: Omphalitis with periumbilical erythema, Category 3: Omphalitis with systemic sepsis, Category 4: Omphalitis with fasciitis and systemic sepsis). The case summaries are depicted in **Table 1**.

All babies were born at term and all received parenteral antibiotics. Blood cultures were negative in all cases except in case 12 (*Staphylococcal epidermidis*). Majority of the cases was referred from other hospitals (60%). Only one case was born at home (case 13). The mean age at presentation was 5 days and the mean hospital stay was 8 days. There were 8 males and 7 females. Out of 7 serious stage/category 4 NF cases, 3 died, giving a mortality of 42% (overall mortality was 20%, 3 out of 15), which shows a reduction by 50% from our previous experience a decade back.[4] The improvement noted could be a reflection of better maternal and child health care, decreased home birth, early recognition, inpatient care with use of parenteral antibiotics and following baby friendly hospital initiative, as suggested by Sawardekar.[3] However, further improvement could be achieved by more aggressive approach, as shown by Garner et al.[5] The use of local agents may also have potential role. A recent randomized clinically trial has also shown the benefits of triple dye/alcohol regime in comparison to dry cord care.[6]

In conclusion, it is evident from the data presented that incidence and mortality related to

Table 1 - Case summaries.

Case number	Year of admission	Gestation	Place of Birth	Sex	Age (days) at presentation	Stay (days)	Outcome	Diagnostic category
1	2000	Term	Out born	Male	5	1	Died	NF
2	2000	Term	Out born	Female	6	4	DC home	Omphalitis
3	2000	Term	In born	Female	11	7	DC home	Omphalitis
4	2000	Term	In born	Male	2	3	DC home	Omphalitis
5	2000	Term	Out born	Female	7	1	Died	NF/Shock
6	2000	Term	Out born	Male	2	10	DC home	Omphalitis
7	2001	Term	Out born	Male	5	15	DC hospital	NF
8	2001	Term	Out born	Male	7	27	DC home	NF
9	2001	Term	In born	Male	1	7	DC home	Omphalitis
10	2001	Term	In born	Female	3	7	DC home	Omphalitis
11	2003	Term	In born	Male	5	4	DC PNW	Omphalitis
12*	2003	Term	In born	Female	5	7	DC home	Omphalitis
13†	2003	Term	Out born	Female	3	13	DC hospital	NF
14	2003	Term	Out born	Female	5	13	DC home	NF
15	2004	Term	Out born	Male	6	3	Died	NF

* *Staphylococcal epidermidis* (umbilical swab and blood culture), †Born at home, DC - discharge, NF - necrotizing fasciitis, PNW - postnatal ward

neonatal omphalitis and necrotizing fasciitis are on a decline in Oman. This could be due to the factors mentioned above. Further, improvement could be achieved with the re-enforcement of the practice of applying antibacterial agents on the umbilical cord soon after birth.

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References

1. Airede AI. Pathogens in neonatal omphalitis. ***J Trop Pediatr*** 1992; 38: 129-131.
2. Guvenc H, Aygun AD, Yasar F, Soylu F, Guvenc M, Kocabay K. Omphalitis in term and preterm appropriate for gestational age and small for gestational age infants. ***J Trop Pediatr*** 1997; 43: 368-372.
3. Sawardekar KP. Changing spectrum of neonatal omphalitis. ***Pediatr Infect Dis J*** 2004; 23: 22-26.
4. Samuel M, Freeman NV, Vaishnav A, Sajwany MJ, Nayyar MP. Necrotizing fasciitis: a serious complication of omphalitis in neonates. ***J Pediatr Surg*** 1994; 29: 1414-1416.
5. Garner P, Lai D, Baea M, Edwards K, Heywood P. Avoiding neonatal death: an intervention study of umbilical cord care. ***J Trop Pediatr*** 1994; 40: 24-28.
6. Janssen PA, Selwood BL, Dobson SR, Peacock D, Thiessen PN. To dye or not to dye: a randomized, clinical trial of triple dye/alcohol regime versus dry cord care. ***Pediatrics*** 2003; 111: 15-20.