

Solitary gumma in a neonate

A case report

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Summary

An unusual case of congenital syphilis is reported. A week-old baby presented with abdominal distension, small-bowel obstruction, bilious vomiting and a large left upper quadrant mass. Histopathological examination of a specimen of the mass confirmed the clinical diagnosis of gumma formation. Primary resection with end-to-end anastomosis was carried out. The baby made an uneventful recovery.

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By definition a gumma is a 'soft, gummy tumor, such as that occurring in tertiary syphilis, made up of tissue resembling granulation tissue'.¹

This short report serves to rebut the presumption that gumma formation does not occur in primary syphilis or in newborn babies, and endorses the belief of renowned medical scientists of the past that syphilis remains the great imitator in medicine.

Case report

A 1-week-old coloured baby girl, weight 1 600 g, born up-country to a proven syphilitic mother, was referred to our unit because of bilious vomiting, abdominal distension and a large left upper quadrant abdominal mass.

On examination the baby was mildly dehydrated and anaemic and the sclera revealed a mild degree of jaundice. There was gross distension of the abdomen, snuffles, and excoriation of the skin, which was accentuated in the region of the mouth and the peri-anal area. The pulse rate was 140/min and the haemoglobin concentration 8,5 g/dl. Palpation of the abdomen revealed a 5 cm hepatomegaly and a fixed rubbery hard intra-abdominal mass, approximately 5 cm in diameter, in the left upper quadrant. The rest of the clinical examination was within normal limits.

Laparotomy revealed a solitary intra-abdominal mass about 5 cm in diameter and the peritoneal cavity contained a small quantity of turbid fluid. Blunt dissection could not separate the bowel from the main mass and further mobilisation and inspection of the mass revealed penetration and perforation of the wall of all three loops of small bowel.

The small bowel was resected and primary end-to-end anastomosis performed. The main mass was mobilised and appeared to arise from the retroperitoneal region. The tumour was removed and the cut surface revealed a well-encapsulated greyish white mass, homogeneous in appearance on the peri-

phery, resembling neuroblastoma. The centre of the mass consisted of softish tumour. The abdomen was closed in layers and the tumour dispatched for light and electron microscopic examination. Histopathological examination of the tumour confirmed the diagnosis of gumma formation. No growth was obtained from the peritoneal fluid. The baby's postoperative course was uneventful.

Discussion

Information gleaned from published reports² indicates that gumma formation in the neonate is an extremely rare phenomenon of babies born to syphilitic mothers, such as the case reported here. Mascola *et al.*³ reported 50 infants with congenital syphilis diagnosed and treated in the state of Texas. Condylomas, osteochondritis, periostitis, snuffles or haemorrhagic rhinitis were considered major clinical signs essential in the diagnosis but no case of gumma formation was encountered. Minor clinical signs consisted of fissure of lips, cutaneous lesions similar to the case described here, mucous patches, hepatosplenomegaly, generalised lymphadenopathy, haemolytic anaemia, central nervous system signs and an elevated number of cells or protein in the cerebrospinal fluid.³

Of 460 cases of congenital syphilis reported by a US study group,² osteochondritis and perichondritis were the most common clinical manifestations.

In our case, jaundice was of a mild nature and physiological jaundice could not be ruled out as the underlying cause. On admission the serum bilirubin value was 78 $\mu\text{mol/l}$, the liver projected 5 cm below the costal margin and because of the left upper quadrant mass the spleen could not be palpated.

Excoriation of skin was obvious with accentuation around the mouth and anal areas. A discharge from the nose reminiscent of snuffles and a fixed mass in the left upper quadrant together with the other clinical manifestations contributed to the tentative pre-operative diagnosis of congenital syphilis.

Histological examination (light microscopy) of the mass revealed the presence of giant cells, epithelioid cells, areas of caseation and necrosis in the centre of the lesion. The histopathological findings, together with the clinical manifestation of congenital syphilis and the serologically positive mother, made gumma of the peritoneal cavity the most likely diagnosis. Culture of the centre of the tumour was negative. Radiography of the skull and long bones was undertaken to rule out the osteochondritis and periostitis commonly encountered in reported cases^{2,3} but not found in our patient.

Long-term monitoring of syphilitic infants is vitally important. It is recommended by Mascola *et al.*³ that infants be re-examined at 3, 6 and 9 months and again at 1 year of age. This follow-up schedule is mandatory since infected infants may be asymptomatic at birth because expression of the disease may be delayed or the clinician may be misled by a negative radiographic examination, which does not rule out congenital syphilis.

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REFERENCES

1. Friel JP, ed. *Dorland's Illustrated Medical Dictionary*, 27th ed. Philadelphia: WB Saunders, 1985.
2. Leads from the *MMWR*. Congenital syphilis — United States 1983 - 1985. *JAMA* 1986; **256**: 3206-3208.
3. Mascola L, Pelosi R, Blount JH, Alexander CE, Cates W jun. Congenital syphilis revisited. *Am J Dis Child* 1985; **139**: 575-580.

Spontaneous bile duct perforation in an infant

A case report

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Summary

A case of spontaneous perforation of the common bile duct in a 4-month-old girl is described. Perforation of the biliary tract is rare, but must be suspected in a child with pale stools, minimal or no jaundice and biliary ascites. Treatment is surgical. Patency of the distal biliary tract must be demonstrated by intra-operative cholecystocholangiography; drainage without repair of the perforation is then appropriate. If there is distal biliary tract obstruction, a biliary-intestinal anastomosis is needed. The difficulty of diagnosis is highlighted by the time between presentation and operation reported in many published cases; suggestions are made to aid a more rapid diagnosis.

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Although perforation of the bile duct in infancy is a rare disorder, it is well documented, with some 70 cases having been reported in English¹⁻⁸ and about 60 cases in Japanese.⁵ Cases have been reported previously in South Africa.^{2,8} Some cases have been secondary to trauma,⁹ cholelithiasis,¹⁰ pancreatitis,⁵ inspissated bile² or torsion of the gall bladder.² Most cases are, however, known as spontaneous infantile perforation of the biliary tract, since no cause can be established. In order to highlight the difficulties of diagnosis and the need for swift and correct surgical management of a disease, which under ideal circumstances should have an excellent prognosis, such a case is reported.

Case report

A 4-month-old baby girl presented to hospital with a 2-week history of abdominal swelling, which had become particularly marked over the preceding few days. The patient was lethargic and feeding poorly. She had been born by caesarean section

for breech presentation; the birth weight was 2 620 g. There was mild neonatal jaundice that settled after 3 days without treatment. At 3 months of age the baby started passing pale stools. Various milk formulas were tried but the pale stools persisted.

On admission the baby was miserable with evidence of wasting, despite a weight of 5 300 g. The skin had a yellow tinge but the sclerae were not jaundiced. The abdomen was tensely distended with ascites. The liver and spleen were both palpable 2 cm below the costal margin. A reducible inguinal hernia was present on the right.

Investigations revealed: haemoglobin 9,7 g/dl; white cell count $26,3 \times 10^9/l$ with 45% neutrophils, 48% lymphocytes, 6% monocytes and 1% eosinophils; platelets $951 \times 10^9/l$; prothrombin index 58%; total protein 72 g/l; serum albumin 34 g/l; serum total bilirubin 18 $\mu\text{mol/l}$; direct bilirubin 10 $\mu\text{mol/l}$; serum alkaline phosphatase 1 168 U/l; serum alanine aminotransferase 58 U/l; serum aspartate aminotransferase 56 U/l.

Paracentesis revealed heavily bile-stained fluid. A perforation of the biliary system was suspected and a laparotomy was performed. Five hundred millilitres of biliary ascites were drained and a thick fibrinous exudate was found covering most of the abdominal contents. The common hepatic duct had a small perforation at the junction with the cystic duct. Intra-operative cholecystocholangiography showed free bile drainage into the duodenum. The abdomen was irrigated, a gall bladder catheter left *in situ* and a Penrose drain inserted near the site of the perforation. The Penrose drain continued to drain for 2 weeks. The drainage then subsided and the Penrose drain and cholecystostomy tube were removed. The stools were now of normal appearance, the hepatosplenomegaly had resolved and the alkaline phosphatase value 10 days after the operation was 421 U/l. The child was discharged well.

Discussion

The cause of spontaneous bile duct perforation is unknown but theories have been proposed implicating a weakness in the bile duct wall, obstruction distal to the perforation or a combination of both (Table I).

The typical history is that of a child initially well who, at the age of 2-12 weeks, presents with pale stools and/or abdominal distension. Jaundice is often absent and if present is usually mild and intermittent. Cases have been reported presenting with an abdominal mass due to pseudocyst formation.^{3,12}

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