Figure 2 Cyst wall showing ciliated columnar epithelium and circular smooth muscle (haematoxylin and eosin; original magnification $\times 520$).

typically located in the posterior wall of the left ventricle.¹¹ Differences in timing of such misplacement probably account for the varying location and composition of foregut cysts.

Haemopericardium leading to intrauterine death is a rare event. Emboli from placental vessels causing acute myocardial infarction with resultant right ventricular rupture was described by Wolf *et al.*¹³ Other cases have followed a motor vehicle accident¹⁴ and amniocentesis.¹⁵ The case described here is unique in that it is the first in which an intrapericardial foregut cyst was

the cause of intrauterine death. We have not been able to find an account of rupture of the heart secondary to this lesion, but given the histogenesis and frequent presentation in early childhood, it is not altogether surprising that a foregut cyst may cause intrauterine death.

- 1 Singer DB, Sung CJ, Wigglesworth JS. Fetal growth and maturation: with standards for body and organ development. In: Wigglesworth JS, Singer DB, eds *Textbook of fetal and perinatal pathology*. Boston: Blackwell 1991:11–48.
- 2 Burke AP, Virmani R. *Tumors of the heart and great vessels*. Atlas of tumor pathology, 3rd series, Fascicle 16. Washington, DC: Armed Forces Institute of Pathology, 1996:111–12.
- 3 Gomes MN, Hufnagel CA. Intrapericardial bronchogenic cysts. *Am J Cardiol* 1975;36:817–22.
- 4 Shimizu M, Takeda R, Mifune J-I, Tanaka T. Echocardiographic features of intrapericardial bronchogenic cyst. *Cardiology* 1990;77:322–6.
- 5 Deenadayalu RP, Tuuri D, Dewall RA, Johnson GF. Intrapericardial teratoma and bronchogenic cyst. Review of literature and report of successful surgery in infant with intrapericardial teratoma. *J Thorac Cardiovasc Surg* 1974;67:945–52.
- 6 Hayashi AH, McLean DR, Peliowski A, Tierney AJ, Finer NN. A rare intrapericardial mass in a neonate. *J Pediatr Surg* 1992;27:1361–3.
- 7 Thomas R, Van Wesep R. Intrapericardial epithelial cyst in association with an atrioventricular canal defect. *Am J Cardiovasc Pathol* 1990;3:325–8.
- 8 Dabbs CH, Berg R, Peirce EC. Intrapericardial bronchogenic cysts. Report of two cases and probable embryologic explanation. *J Thorac Surg* 1957;34:718–35.
- 9 Jellen J, Fisher WE. Intrapericardial teratoma. *Am J Dis Child* 1936;51:1397–402.
- 10 Machens G, Vahl CF, Hofmann R, Wolf D, Hagl S. Endothelial inclusion cyst of the tricuspid valve. *Thorac Cardiovasc Surgeon* 1991;39:296–8.
- 11 De Lacroix WF, Hubner G. Ciliated epithelial inclusion cyst of the heart. *Beitr Pathol* 1974;151:103–10.
- 12 Todd JL, Silverman ME, Kirby ML, Gray SW, Skandalakis JE. Heart. In: Skandalakis JE, Gray SW eds. *Embryology for surgeons*, 2nd edn. Baltimore: Williams and Wilkins, 1994: 912–57.
- 13 Wolf PL, Jones KL, Longway SR, Benirschke K, Bloor C. Prenatal death from acute myocardial infarction and cardiac tamponade due to embolus from the placenta. *Am Heart J* 1985;109:603–5.
- 14 Sherer DM, Abramowicz JS, Babowski R, Metlay LA, Ron M, Woods JR. Extensive fetal intrathoracic injuries sustained in a motor vehicle accident. *Am J Perinatol* 1993; 10:414–16.
- 15 Berner HW, Seisler EP, Barlow J. Fetal cardiac tamponade. A complication of amniocentesis. *Obstet Gynecol* 1972;40: 599–604.

Gastric outflow obstruction caused by gall stones and leading to death by complex metabolic derangement

C O Wight, M Seed, W W Yeo, T A McCulloch

Departments of
Histopathology and
Medicine, Royal
Hallamshire Hospital,
Glossop Road,
Sheffield S10 2JF, UK

Correspondence to:
Dr Wight, Department of
Histopathology, Nottingham
City Hospital NHS Trust,
Hucknall Road, Nottingham,
NG5 1PB, UK.

Accepted for publication
2 September 1997

Abstract

A 67 year old woman was admitted with a three week history of vomiting, having become increasingly confused for three days. Investigations revealed deranged serum biochemistry consistent with a combination of a diabetic non-ketotic hyperosmolar state and a metabolic alkalosis consistent with gastric outflow obstruction. She was treated with intravenous saline, intravenous insulin, and subcutaneous heparin, but did not improve clinically and

had an asystolic cardiac arrest the following day; she was transferred to the intensive care unit and despite treatment with inotropes she died 40 hours after admission. Necropsy revealed that the stomach was massively dilated with gas and stomach contents, and contained many small black faceted gall stones. In addition a large non-faceted brown-yellow gall stone was wedged in the pyloric antrum causing total obstruction. The patient had died from a complex metabolic derangement including

Figure 1 A large gall stone is impacted in the gastric antrum (arrow). A probe is present in the common bile duct (ST, stomach; L, liver).

non-ketotic hyperosmotic diabetic coma and metabolic alkalosis precipitated by the acute gastric outflow obstruction complicated by previously undiagnosed type II diabetes mellitus.

(*J Clin Pathol* 1997;50:963–965)

Keywords: gastric outflow obstruction; gall stones; diabetes mellitus

Gall stone ileus occurs when a gall stone erodes into part of the gastrointestinal system and causes intestinal obstruction. It is a well recognised but rare complication of cholelithiasis, resulting in 1–2% of all adult small bowel obstructions.¹ The importance of gall stone ileus lies in its high mortality (12–27%), which is thought to be due to the patients being elderly and suffering from concomitant diseases.² The most frequent site of impaction of the gall stone is in the terminal ileum following erosion of the gall stone into the duodenum.¹ Gastric outlet obstruction by gall stones is a very rare but recognised complication of cholelithiasis resulting from erosion of a gall stone into the gastric antrum.³

We report a case of death by non-ketotic hyperosmolar coma resulting from pyloric obstruction caused by perforation of a large gall stone into the pyloric antrum and complicated by previously undiagnosed non-insulin dependent diabetes. To the best of our knowledge, this is the third reported case of gastric outlet obstruction by a gall stone and the first complicated by non-insulin dependent diabetes.

Case report

A 67 year old woman was admitted with a three week history of vomiting, having become increasingly confused for three days. She had longstanding hypertension that was treated with methyl-dopa, atenolol, and a combination of amiloride and cyclopentiazide. Otherwise, past medical history was unremarkable.

On examination she was obese, drowsy, and very dehydrated. Her blood pressure was 70/40 mm Hg and she had atrial fibrillation. There was clinical suspicion of an epigastric

mass, which was thought to be a tumour, but her abdomen was non-tender and bowel sounds were present.

Investigations revealed deranged serum biochemistry consistent with a combination of a diabetic non-ketotic hyperosmolar state (calculated osmolality 347 mmol/l) and a metabolic alkalosis consistent with gastric outflow obstruction (sodium 118 mmol/l, potassium 3.9 mmol/l, bicarbonate 43 mmol/l, urea 69.5 mmol/l, creatinine 441 mmol/l, glucose 41.5 mmol/l).

She was treated with intravenous saline, intravenous insulin, and subcutaneous heparin, but did not improve clinically and had an asystolic cardiac arrest the following day after an episode of vomiting. Cardiac output was restored with adrenaline and atropine and she was transferred to the intensive care unit. Despite treatment with inotropes she steadily deteriorated and died 40 hours after admission.

PATHOLOGICAL EXAMINATION

At postmortem examination the stomach was massively dilated with gas and stomach contents, and found to contain many small black faceted gall stones, each approximately 5 mm in maximum diameter. In addition a large non-faceted brown-yellow gall stone measuring $5 \times 3.5 \times 3.5$ cm was wedged in the pyloric antrum causing total obstruction (fig 1). There was a 2 cm diameter fistula running directly from the gall bladder into the pyloric antrum, the mucosa of which was eroded and the wall appeared fibrotic. The gall bladder also contained multiple small faceted black gall stones. The common bile duct and left and right hepatic ducts were dilated but contained no gall stones. The liver appeared normal on external surface but was congested on slicing. Histology was unremarkable. The rest of the gastrointestinal system was within normal limits. The only other postmortem findings were a minimal degree of atheroma in the large arteries and some pulmonary oedema.

We concluded that the patient had died from a complex metabolic derangement including non-ketotic hyperosmotic diabetic coma and metabolic alkalosis precipitated by the acute gastric outflow obstruction complicated by previously undiagnosed type II diabetes mellitus.

Discussion

Gall stone ileus has a high mortality, which is thought to result from the elderly age of many patients,⁴ concomitant disease, and delay in seeking medical care after intestinal obstruction. In one study 50% of patients presenting with gall stone ileus had significant pulmonary disease, 33% had cardiac disease, and 40% had diabetes mellitus.⁵ In another study the number of patients having concomitant diabetes mellitus was as high as 50%.⁶ In this case diabetes mellitus was previously undiagnosed and the combination of the gastric outflow obstruction and diabetes led to a complex metabolic picture resulting in death. Furthermore, less than half of patients presenting with gall stone ileus have known biliary tract disease, and so there is often a delay in diagnosis⁶ once medical help is sought.

This case is important as it illustrates the combination of an unusual site of gall stone impaction, which was not diagnosed in life, and the effects of age, delay in seeking medical care, and concomitant previously undiagnosed disease leading to fatality. We believe it to be only the third reported case of gastric outflow obstruction by a gall stone and the first combined with non-insulin dependent diabetes mellitus causing death by this complex metabolic derangement.

- 1 Stitt RB, Heslin DJ, Currie DJ. Gall-stone ileus. *Br J Surg* 1967;54:673-8.
- 2 Clavien PA, Richon J, Burgan S, Rohner A. Gallstone ileus. *Br J Surg* 1990;77:737-42.
- 3 Otten VG, Helwing E, Jung D. Cholezysto-gastrale Gallensteinperforation. *Zentralbl Chir* 1978;103:941-2.
- 4 Brockis JC, Gilbert MC. Intestinal obstruction by gall stones. A record of 179 cases. *Br J Surg* 1957;44:461-6.
- 5 Kurtz RJ, Heiman TM, Beck AR, Kurtz AB. Patterns of treatment of gallstone ileus over a 45 year period. *Am J Gastroenterol* 1985;80:95-8.
- 6 Day EA. Gallstone ileus. Review of the literature and presentation of thirty four new cases. *Am J Surg* 1975;129:552-8.

Systemic reactive amyloidosis associated with Castleman's disease: serial changes of the concentrations of acute phase serum amyloid A and interleukin 6 in serum

Shu-ichi Ikeda, Hisanao Chisuwa, Seiji Kawasaki, Junko Ozawa, Yoshinobu Hoshii, Tadaaki Yokota, Tuneto Aoi

Department of
Medicine (Neurology),
Shinshu University
School of Medicine,
Matsumoto 390, Japan
S Ikeda

Department of
Surgery, Shinshu
University School of
Medicine, Matsumoto
390, Japan
H Chisuwa
S Kawasaki

Central Clinical
Laboratory, Shinshu
University Hospital,
Matsumoto 390, Japan
J Ozawa

Department of
Pathology, Yamaguchi
University School of
Medicine, Ube 744,
Japan
Y Hoshii

Department of
Pathology, Kokura
Memorial Hospital,
Kitakyushu 802, Japan
T Yokota

Department of
Medicine, Gamagouri
City Hospital, Aichi
443, Japan
T Aoi

Correspondence to:
Dr Ikeda; email:
ikedasi@gipac.shinshu-u.ac.jp

Accepted for publication
15 July 1997

Abstract

A case is reported of a 21 year old woman who suffered from Castleman's disease and systemic reactive amyloidosis. The serum concentrations of serum amyloid A (SAA) and interleukin 6 (IL-6) were extremely high and amyloid protein was immunohistochemically identified as AA. After surgical excision of a large retroperitoneal lymph node with the pathological findings of plasma cell type of Castleman's disease, both serum SAA and IL-6 declined, showing a similar pattern of reduction curves. All clinical symptoms and laboratory abnormalities greatly improved. The biochemical feature of Castleman's disease is abnormal production of IL-6 and this cytokine continuously may stimulate the synthesis of an amyloid precursor, SAA, causing systemic reactive (AA) amyloidosis. This pathogenetic theory is strongly supported by the present study. (*J Clin Pathol* 1997;50:965-967)

Keywords: Castleman's disease; amyloid; serum amyloid A; interleukin 6

Systemic reactive (AA) amyloidosis usually occurs in patients with chronic inflammatory disorders. Serum amyloid A (SAA), which is an acute phase reactant of hepatic origin, is an amyloid precursor in this type of amyloidosis. It is well known that the serum concentration of SAA dramatically increases in the inflammatory state and this response is mediated by actions of some cytokines including interleukin 1 (IL-1), IL-6, and tumour necrosis factor (TNF).^{1,2}

Castleman's disease is a unique form of lymphoproliferative disorder characterised pathologically by the presence of giant lymph node

hyperplasia with plasma cell infiltration.^{3,4} Patients with this disease commonly have fever, anaemia, hypergammaglobulinaemia, and an increase in the serum concentrations of acute phase reactant proteins, all of which are ascribed to the large amount of IL-6 produced in the hyperplastic lymph nodes.^{5,6} We examined serial changes of serum concentrations of SAA and IL-6 in a patient with Castleman's disease and systemic reactive amyloidosis who was treated with surgical removal of an involved large solitary lymph node.

Case report

The patient was a 21 year old woman with a nine year history of general fatigue, arthralgia, and slight fever. In the past three years she had been suffering from epigastric discomfort and poor appetite. On examination, she looked sick and had an enormously enlarged liver with hard consistency, other physical findings were unremarkable. Abnormal laboratory findings are summarised in table 1. Briefly, she had a raised erythrocyte sedimentation rate (152 mm in the first hour), raised C reactive protein (CRP), anaemia, thrombocytosis, hypoalbuminaemia, hypergammaglobulinaemia, and slight proteinuria. Biopsies of liver tissues and gastric mucosa showed severe amyloid deposits with alkaline Congo red staining: amyloid deposition diffusely involved the parenchymal sinusoidal space of Disse and this was also seen in the lamina propria and muscularis mucosae of the gastric mucosa. After pretreatment with KMnO₄, these amyloid deposits lost the affinity for Congo red dye (data not shown). Computed tomography of the abdomen revealed a mass with small calcifications that mimicked a liver tumour in the caudate lobe. Surgical