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Case report

Malakoplakia of the pancreas

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Summary. We report a case of malakoplakia arising in the pancreas of a 45-year-old woman. Clinical and histological features are described. Malakoplakia was present with chronic pancreatitis and adenocarcinoma. The aetiology of malakoplakia at this site may be due to a stromal response to carcinoma or an abnormal localised histiocytic response to infection or inflammation. This case and previous reported cases suggest that the aetiology and pathogenesis of pancreatic malakoplakia is not the same for all patients.

Key words: Malakoplakia – Pancreas

Introduction

Malakoplakia is a rare condition which was first described in the urinary tract by Michaelis and Gutmann in 1902. It is characterised by aggregates of histiocytes containing diastase-resistant, periodic-acid-Schiff-positive granules (Hansemann cells) and laminated calcospherites (Michaelis-Gutmann bodies) (Michaelis and Gutmann 1902; von Hanseman 1903). The urinary tract is the most common site of involvement, but it has been described in many other sites including the male and female reproductive tracts, gastrointestinal tract, retroperitoneum, lymph nodes, brain, lungs, bones, skin, adrenals, conjunctiva, tonsils and middle ear (McClure 1983; Azadeh and Ardehali 1983). Pancreatic involvement by malakoplakia is rare, and only three cases have been previously reported. We present a case of malakoplakia arising in a background of chronic pancreatitis and pancreatic adenocarcinoma.

Case report

A 45-year-old woman presented to her general practitioner several years ago, complaining of recurrent attacks of upper abdominal pain. Prior to this she had been in good health. In May 1988 her abdominal pains became worse and she developed anorexia, nausea, weight-loss and steatorrhoea. There was no history of jaundice, hyperlipidaemia, hypercalcaemia or excessive alcohol consumption. Computed tomographic scanning revealed a probable pancreatic neoplasm.

In September 1988 she underwent a Whipple's procedure. No abnormality of the biliary tract was found at laparotomy. Postoperatively she did well. However, in June 1989 her condition deteriorated. Liver biopsy showed metastatic adenocarcinoma. She died in July 1989, but permission for autopsy was declined.

Macroscopic findings. The specimen consisted of a 21-cm loop of duodenum attached to an irregular ($8 \times 6 \times 4$ cm) mass of firm, white tissue which on section had a nodular appearance. Scattered, small, orange-red foci were noted as well as cysts up to 0.2 cm in diameter which contained thick, white secretion. There was no evidence of necrosis or haemorrhage. Several lymph nodes were present within surrounding fatty tissue.

Microscopic findings. Histological sections were processed from tissue fixed in 10% formol-saline by standard technique and stained with haematoxylin and eosin. Sections through the head of the pancreas showed features of chronic pancreatitis (Fig. 1). Within these areas, centred predominantly around ducts, were foci of histiocytes containing scattered lymphocytes (Fig. 2). The histiocytes had granular cytoplasm (Hansemann cells) and lying within and between these cells were laminated Michaelis-Gutmann bodies. These structures stained positive after diastase digestion with periodic acid-Schiff and also for calcium (Figs. 3, 4). No organisms could be identified in Gram-stained sections. Also present in the head of the pancreas was a moderately well-differentiated adenocarcinoma which invaded into the wall of the duodenum reaching the submucosa (Fig. 5). One of four lymph nodes examined contained metastatic adenocarcinoma.

Discussion

The aetiology of malakoplakia remains unknown. It is seen in three different clinical situations (McClure 1983). In the first and largest group, patients present with genitourinary symptoms. Many have recurrent urinary tract infections due to *Escherichia coli* and these patients may have a defect in host histiocyte response to infection by Gram-negative coliform bacteria. Secondly, malak-

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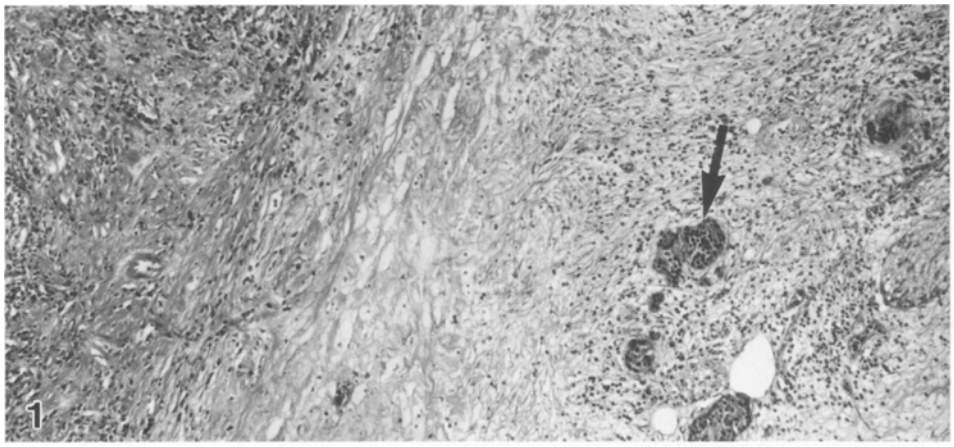
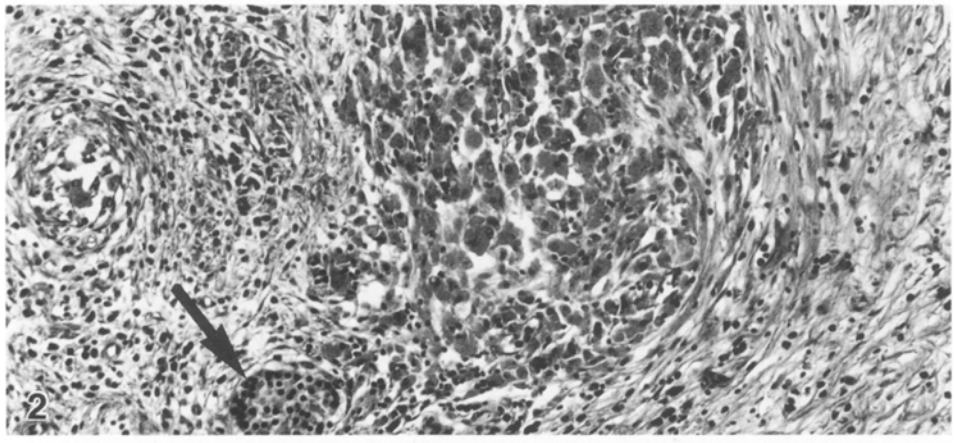




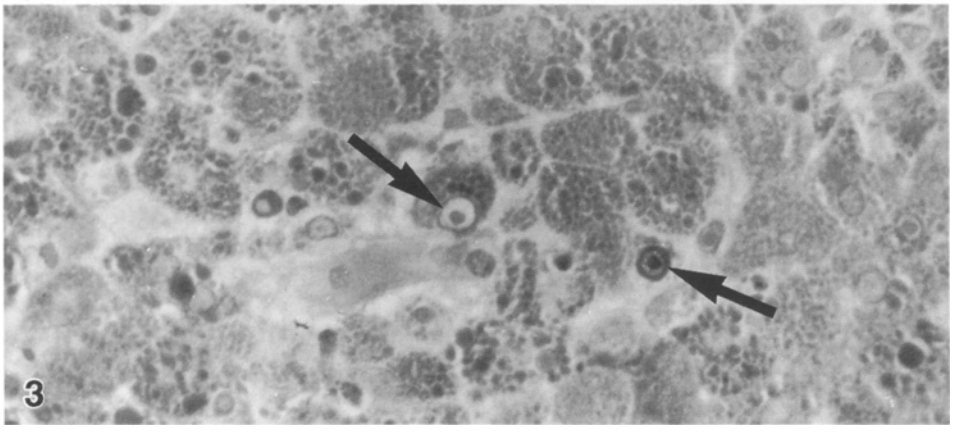


Fig. 1. Chronic pancreatitis with atrophy and loss of acini, preservation of islets of Langerhans (*arrowed*), increase in fibrous tissue and a chronic inflammatory cell infiltrate. H&E, ×20

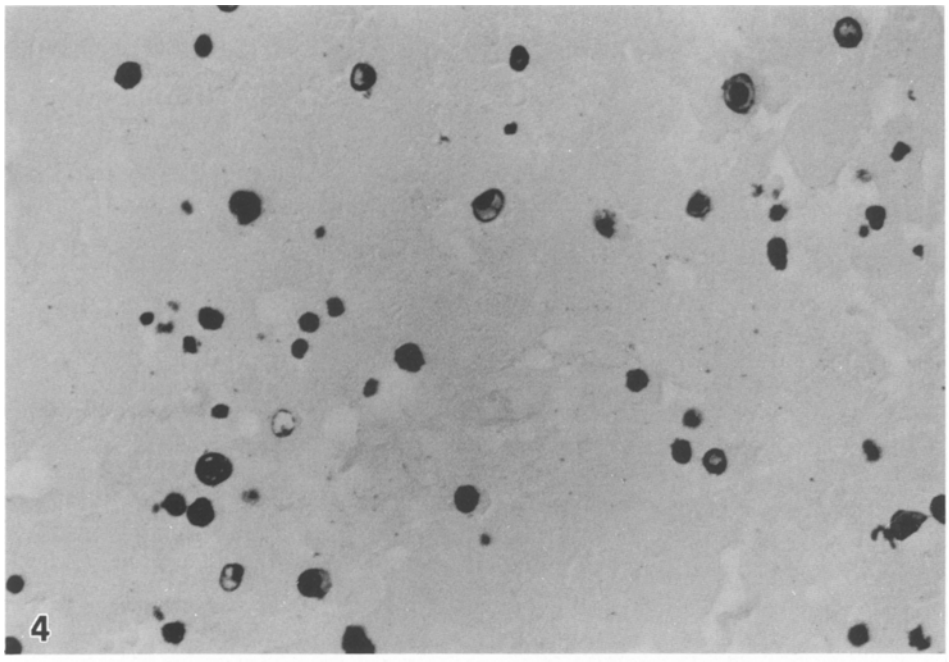
Fig. 20. Histiocytic aggregate lying near pancreatic ductal remnants (*top left*) and an islet of Langerhans (*arrowed*) H&E, ×40

Fig. 3. Histiocytes containing diastase-resistant, PAS-positive granules. Laminated Michaelis-Gutmann bodies *arrowed.* PAS diastase, ×160

oplakia presents outside the genitourinary tract, for example, in the retroperitoneum, gastrointestinal tract, bone, lung, skin and brain. As in the first group it may be due to an abnormal, localised histiocytic response to chronic infection or inflammation, but some have an underlying malignancy and malakoplakia may be produced by a stromal response to the tumour. The third group usually present with disseminated malakoplakia and these patients usually have a primary or secondary immunodeficiency.

Three cases of malakoplakia involving the pancreas have been reported (Table 1). Sinclair-Smith et al. (1975) described a case of malakoplakia in the adrenal gland and colon with extension into the pancreas and perinephric adipose tissue. The patient was a 6-week-old male with thymic and lymphoid hypoplasia who developed fatal miliary tuberculosis. They concluded that malakoplakia developed at this site is a response to extravasated erythrocytes or to erythrocytes and bacteria. In 1978, Colby reported a case in which the tail of the pancreas was involved by a large retroperitoneal malakoplakic lesion. This 49-year-old male had steroid-treated haemolytic anaemia, but no conclusion was given for the aetiology of malakoplakia at this site. Kulatunga and Kyllönen (1987) presented a case arising as a solitary mass in the head of the pancreas in a healthy 75-year-old male which was discovered at elective cholecystectomy. They concluded that malakoplakia was produced by chronic inflammation.

Our case differs from those above in that our patient



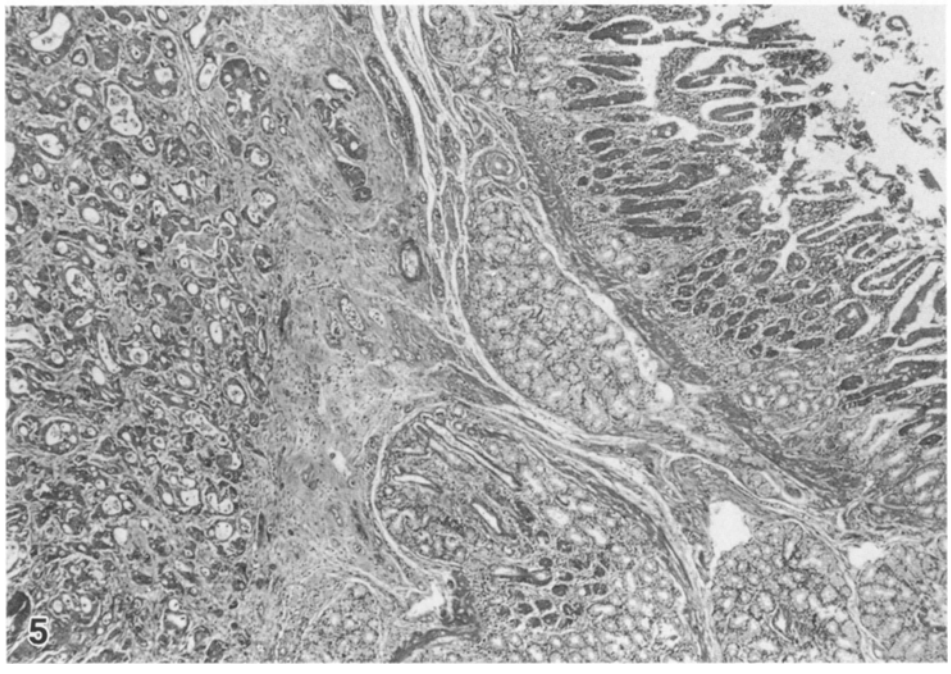


Fig. 4. Michaelis-Gutmann bodies after staining for calcium. Von Kossa, ×160

Fig. 5. Pancreatic adenocarcinoma (*left*) infiltrating into duodenal submucosa. H&E, ×10

Table 1. Details of four patients with pancreatic malakoplakia

Case no.	Age	Sex	Site of malakoplakia	Proposed aetiology	Other features	Evidence of immunodeficiency	Clinical outcome
1[a]	6 weeks	M	Adrenal and colon with spread into tail of pancreas	Response to extravasated erythrocytes and/or bacteria	Thymic and lymphoid hypoplasia	Yes	Died of miliary TB
2[b]	49 years	M	Retroperitoneal with spread into tail of pancreas	None given	Steroid-treated haemolytic anaemia	Yes	Not known
3[c]	75 years	M	Solitary mass in head of pancreas	Response to chronic inflammation	Healthy; lesion found at elective cholecystectomy	No	Well
4[d]	45 years	F	Head of pancreas	Stromal response to carcinoma and/or chronic pancreatitis	Lesion found after Whipple's procedure	No	Died 1 year after diagnosis

[a] Sinclair-Smith, Kahn and Cywes (1975) [b] Colby (1978) [c] Kulatunga and Kyllönen (1987) [d] Our case

is female. The first two cases above show malakoplakia involving the pancreas spreading from adjacent tissues. However, our case is similar to the third case in that malakoplakia appears confined to pancreatic parenchymal tissues. The aetiology of malakoplakia in our patient may be due to a stromal response to infiltrating adenocarcinoma or an abnormal local histiocytic response to infection or inflammation associated with chronic pancreatitis. Clinically our patient would belong to the second group of patients described previously and the three cases described above would most likely belong to categories 3, 3 and 2 respectively.

In conclusion, malakoplakia is rare and is usually seen in three different clinical settings. Pancreatic involvement by malakoplakia has only been described in three other patients and in our case appears to be totally confined to this organ. This is the first reported case in a female. In our patient the aetiology of malakoplakia may be due to a stromal response to adenocarcinoma or an abnormal histiocytic response to infection or inflammation associated with chronic pancreatitis. This case and previous reported cases suggest that the aetiology and pathogenesis of pancreatic malakoplakia is not the same for all patients.

References

Azadeh B, Ardehali S (1983) Malakoplakia of middle ear: a case report. Histopathology 7:129–134

Colby TV (1978) Malakoplakia. Two unusual cases which presented with diagnostic problems. Am J Surg Pathol 2:377–382

Hanseman D von (1903) Über Malakoplakie der Harnblase. Virchows Arch Pathol Anat 173:302–308

Kulatunga A, Kyllönen AP (1987) Malakoplakia of the pancreas. A case report. Acta Pathol Microbiol Immunol Scand [A] 95:127–129

McClure J (1983) Malakoplakia. J Pathol 140:275–330

Michaelis L, Gutmann C (1902) Über Einschlusse in Blasentumouren. Z Klin Med 47:208–215

Sinclair-Smith C, Kahn LB, Cywes S (1975) Malacoplakia in childhood: case report with ultrastructural observations and review of the literature. Arch Pathol Lab Med 99:198–203