Leiomyosarcoma of the duodenum is a rare malignant tumour. The majority of reported cases were not diagnosed preoperatively (Waugh, Harp, and ReMine, 1963; Lie and Sinclair, 1966), and even at operation the correct diagnosis was often missed (von Salis, 1920; Nielsen and Mahin, 1962). Many cases were diagnosed by histological examinations (Dodds and Beahrs, 1963) and quite a number at necropsy (Starr and Dockerty, 1955). The high operative mortality and the generally poor prognosis which characterized this neoplasm in the past were due partly to wrong preoperative diagnosis and partly to inadequate awareness of the behaviour of this uncommon neoplasm. The standard textbooks make little or no reference to it but recently there has been an increased number of publications on this tumour. Also the operative mortality has been reduced due to improvement in the surgical technique of pancreatico-duodenectomy, safer anaesthetic methods, and advances in supportive therapy before, during, and after operation (such as transfusion of blood and plasma expanders and antibiotics). A review of recent publications shows that preoperatively the correct diagnosis of leiomyosarcoma of the duodenum still eludes many surgeons, and it is hoped that by reporting this case, together with a review of the world literature, the diagnostic problems will be underlined and the salient features in the diagnosis of the tumour so outlined that preoperative diagnosis will be made easier.

CASE REPORT

A 61-year-old Nigerian woman was admitted to University College Hospital, Ibadan, Nigeria, on 11 February 1966 with a history of a swelling and pain in the upper part of the abdomen, weakness and breathlessness on exertion, and swelling of the legs for about four months. The abdominal pain was intermittent and worse on movement and it occasionally radiated to the right flank of the trunk. There was nausea with occasional vomiting, and 11 days before admission she vomited altered blood. She had passed black stools on and off for about three months. There was no history of jaundice or urinary trouble, and no loss of weight. In 1960 an amoebic liver abscess was drained surgically in the same hospital and she had remained perfectly healthy until the present illness.

On examination, she was pale, obese, and ill-looking. She had an old 3 in. scar in the right subcostal region. There was a large tender mass in the right hypochondrium extending from the subcostal margin to just above the umbilicus. The mass was cystic and moved very little with respiration. One could just get above it. She was not jaundiced. The pulse rate was 124/min, regular, and of full volume; blood pressure 110/50 mm Hg. Jugular venous pressure was raised. Pitting oedema of both legs and feet was present. The heart was slightly enlarged and there was a soft systolic murmur over the left side of the heart. There were fine crepitations over both lung bases.

FIG. 1. Barium meal examination showing distortion and displacement of the distal stomach and first part of the duodenum. Barium-filled cavity in the mass, 'fistula' directed downwards and to the right.
Faecal occult blood was positive, and there was only a trace of urobilinogen in the urine. Liver function tests, electrolytes, and serum amylase were normal. She was O Rh positive, PCV 25%. An ECG showed sinus tachycardia and flat T waves. A chest film showed the right dome of the diaphragm to be raised; and a plain film of the abdomen confirmed a large soft tissue mass on the right side of the abdomen. The mass did not appear to be related to the kidney or the liver. In the barium meal films the pyloric antrum was displaced upwards, there was considerable distortion of the duodenal cap and a large ‘fistula’ running downwards and to the right. The ‘fistula’ was thought to be an abscess cavity communicating with an old perforated duodenal ulcer (Fig. 1).

Clinically, the differential diagnosis of a pancreatic cyst was also considered. After the patient had been adequately prepared a laparotomy was undertaken. At operation on 15 March, a large retroperitoneal vascular tumour, about 12 cm by 18 cm, was found situated in the right hypochondrium. The distal part of the stomach and first part of the duodenum were adherent and stretched over the upper part of the tumour. The rest of the duodenum could not be visualized. The transverse mesocolon and a greater part of the transverse colon were adherent and infiltrated by the tumour. The right lobe of the liver, the gall bladder, and the head and body of the pancreas were infiltrated and adherent to the tumour. There were no discrete secondary deposits in the liver and no peritoneal seedlings. The lymph nodes were not enlarged. The tumour was assessed unresectable, and when a biopsy was being taken the tumour ruptured and a mass of necrotic greenish material gushed out. The spillage and the inside of the tumour were sucked dry but it made very little difference to the size and mobility of the tumour. A biopsy was taken at the site of the rupture and the defect in the tumour repaired. The peritoneal cavity was drained and the abdomen closed. The patient developed a faecal fistula on the sixth post-operative day and died 21 days after operation.

Histology of the biopsy showed cells which were pleomorphic but spindle-shaped cells predominated. The nuclei were hyperchromatic and here and there were arranged in pailissades. Mitotic figures were many and myofibrils were demonstrated by phosphotungstic acid haematoxylin staining. Leiomyosarcoma of the duodenum was diagnosed (Fig. 2).

At necropsy there was a mass measuring 10 × 15 cm situated anteriorly in the second part of the duodenum. There was a central haemorrhagic cavity communicating through an ulcer with the lumen of the duodenum. The tumour contained areas of calcification. It had infiltrated into the right lobe of the liver, gall bladder, stomach, pancreas, and transverse colon. Faeces had leaked through the site of the biopsy.

**DISCUSSION**

Leiomyosarcoma of the duodenum was first reported by von Salis in 1920, and since then there have been a number of reviews and reports of isolated cases in the literature. Bradham (1959) reviewed 54 cases including one of his own, and Starzl, Bernhard, and Heneger (1960a) reviewed 61 cases including two of their own. The publications on this neoplasm since the last comprehensive review by Starzl et al (1960b) are summarized in Table I.
### TABLE I

**SUMMARY OF PUBLISHED CASES**

<table>
<thead>
<tr>
<th>Author, Year</th>
<th>Age</th>
<th>Sex</th>
<th>Symptoms</th>
<th>Findings</th>
<th>Radiology</th>
<th>Gross Pathology</th>
<th>Site</th>
<th>Treatment</th>
<th>Result</th>
</tr>
</thead>
<tbody>
<tr>
<td>Amerson et al (1959)</td>
<td>65</td>
<td>M</td>
<td>Epigastric pain, weakness, nausea and vomiting, gastrointestinal bleeding</td>
<td>Anaemia</td>
<td>—</td>
<td>Tumour 3 × 4 cm with mucosal ulceration; no metastases; sinus formation</td>
<td>2nd</td>
<td>Peptic ulcer regimen, blood transfusion</td>
<td>Died</td>
</tr>
<tr>
<td>Amerson et al (1959)</td>
<td>60</td>
<td>F</td>
<td>Indigestion, weakness, melena</td>
<td>Anaemia</td>
<td>Filling defect 3rd part of duodenum associated with a mass</td>
<td>Not stated</td>
<td>Resection of 3rd part of duodenum</td>
<td>Doing well 3 months later</td>
<td></td>
</tr>
<tr>
<td>Bradham (1959)</td>
<td>47</td>
<td>M</td>
<td>Rectal bleeding, weakness, dyspnoea, episodic syncope</td>
<td>Anaemia</td>
<td>Filling defect in 3rd part associated with a mass</td>
<td>Tumour 6:5 × 4:5 cm, haemorrhagic degeneration, no metastases</td>
<td>3rd</td>
<td>Resection of 3rd part of duodenum</td>
<td>Well yr later</td>
</tr>
<tr>
<td>Nielsen and Mahin (1962)</td>
<td>72</td>
<td>M</td>
<td>Weakness, loss of weight, melena</td>
<td>Anaemia, mass</td>
<td>Fistula</td>
<td>Cystic mass, central necrosis, fistula</td>
<td>Not stated</td>
<td>Duodenectomy</td>
<td>Well after 1 year</td>
</tr>
<tr>
<td>Vieta (1962)</td>
<td>55</td>
<td>M</td>
<td>Malaise, fever, abdominal pain</td>
<td>Generalized peritonitis</td>
<td>Perforation of viscus</td>
<td>Large mass with perforation</td>
<td>4th</td>
<td>Excision of tumour with adjacent duodenum</td>
<td>Well</td>
</tr>
<tr>
<td>Dodds and Beahrs (1963)</td>
<td>72</td>
<td>M</td>
<td>Epigastric pain</td>
<td>Anaemia</td>
<td>Filling defect</td>
<td>Tumour mass with central calcification</td>
<td>2nd</td>
<td>Pancreateco-duodenectomy</td>
<td>Died after 1 year. Secondaries</td>
</tr>
<tr>
<td>Dodds and Beahrs (1963)</td>
<td>50</td>
<td>M</td>
<td>Weakness, abdominal pain, gastrointestinal bleeding, flatulence</td>
<td>Mass</td>
<td>Fistula</td>
<td>Large mass infiltrating other structures</td>
<td>Not stated</td>
<td>Pancreateco-duodenectomy</td>
<td>Well 18 mth later</td>
</tr>
<tr>
<td>Spinazzola and Gillesby (1963)</td>
<td>64</td>
<td>M</td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Barlebo and Mathiesen (1963)</td>
<td>42</td>
<td>F</td>
<td>Epigastric pain, melena, fainting</td>
<td>Anaemia</td>
<td>Filling defect</td>
<td>Mass, submucosal, ulcerated</td>
<td>2nd</td>
<td>Pancreateco-duodenectomy</td>
<td>Well</td>
</tr>
<tr>
<td>Recordier, Legré, Vignoli, and Gaujoux (1963)</td>
<td>28</td>
<td>M</td>
<td>Epigastric pain, haematemesis</td>
<td>Mass</td>
<td>Duodenum normal, extrinsic pressure on antrum</td>
<td>Mass spreading around ampulla</td>
<td>2nd</td>
<td>Pancreateco-duodenectomy</td>
<td>Biliary-pancreatic fistula; died 6 wk post-operatively</td>
</tr>
<tr>
<td>Rosenberg (1964)</td>
<td>64</td>
<td>M</td>
<td>Weakness, melena</td>
<td>Mass, anaemia</td>
<td>Soft mass displacing transverse colon and ureter (R)</td>
<td>Cystic haemorrhagic tumour, 11 cm diameter</td>
<td>3rd</td>
<td>Resection of tumour with adjacent duodenum</td>
<td>Well at first; died 1 year later with secondaries</td>
</tr>
<tr>
<td>Peterson, Franklin, and Peterson (1964)</td>
<td>57</td>
<td>F</td>
<td>Abdominal pain, weakness, melena</td>
<td>Tenderness in epigastrium, anaemia</td>
<td>Active duodenal ulcer</td>
<td>No evidence of duodenal ulcer, mass 5 × 7 cm</td>
<td>3rd</td>
<td>Resection of tumour with adjacent duodenum</td>
<td>Well</td>
</tr>
<tr>
<td>Peycelon, Jegou, and Delore (1965)</td>
<td>40</td>
<td>M</td>
<td>Loss of weight, melena</td>
<td>Anaemia</td>
<td>Filling defect</td>
<td>Large hard mass, 4th dumb bell</td>
<td>2nd</td>
<td>Pancreateco-duodenectomy</td>
<td>Survived</td>
</tr>
<tr>
<td>Lie and Sinclair (1966)</td>
<td>42</td>
<td>M</td>
<td>Epigastric pain, gastrointestinal bleeding, weakness</td>
<td>Tenderness in epigastrium, anaemia</td>
<td></td>
<td>Woody, hard tumour, central cavity</td>
<td>2nd</td>
<td>Pancreateco-duodenectomy</td>
<td>Well</td>
</tr>
<tr>
<td>Perry and McClelland (1966)</td>
<td>71</td>
<td>F</td>
<td>Generalized abdominal pain</td>
<td></td>
<td></td>
<td>Intrapancreatic haemorrhage, mass 2 × 3 cm</td>
<td>4th</td>
<td>Subtotal duodenectomy</td>
<td>Well</td>
</tr>
<tr>
<td>Serrano and McPeak (1966)</td>
<td>Reported (a)</td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Olurin and Solanke (1968)</td>
<td>61</td>
<td>F</td>
<td>Seven cases clinically diagnosed</td>
<td>No individual details</td>
<td>Filling defects, 'fistula'</td>
<td>Tumour 15 × 10 cm, ulceration, central necrosis</td>
<td>2nd</td>
<td>Laparotomy, unresectable</td>
<td>Died 21 days after operation</td>
</tr>
</tbody>
</table>

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(a) Includes one case found at necropsy.
(b) One case presented at Association Internationale pour l'étude de la Tumeur du Pancreas.
83 cases have now been collected from the world literature to which we add a case of our own.

Leiomyosarcoma of the duodenum constitutes about 2% of all malignant tumours of the small intestine (Mayo, 1940), about 18% of the leiomyosarcoma of the small intestine (Starr and Dockerty, 1955), and about 10% of all the malignant tumours of the duodenum (Brenner and Brown, 1955; Ochsner and Kleckner, 1957). In 65 cases, including our own, in which details are given in the literature, the site of the neoplasm in relation to the different anatomical subdivisions of the duodenum has been worked out as follows: 8% were situated in the first part, 52% in the second part, 28% in the third part, and 12% in the fourth part (Fig. 3). The neoplasm appears to affect males (53%) slightly more than females (47%) and the age incidence varies from 21 to 80 years. The majority of them (40%) occurred between 40 and 49 years (Starzl et al, 1960a, and Table 1).

The commonest presenting symptoms are abdominal pain and gastrointestinal bleeding. The abdominal pain is usually confined to the upper part, and varies considerably in character. The pain may be vague and ulcer like, a dull, boring pain, or it may be a cramping pain. It is usually constant.

The bleeding is more often of the upper gastrointestinal type, haematemesis being prominent, but melaena is also common. The bleeding may be either chronic and unnoticed until anaemia is well established, or it may be acute and profuse, requiring massive blood transfusion or emergency operation (Amerson and Lumpkin, 1959; Lie and Sinclair, 1966). Other symptoms are nausea and vomiting, weakness, diarrhoea, fever, and jaundice. A large proportion of the patients present with a mass in the upper abdomen.

Anaemia is a common finding. Faecal occult blood is often positive. Of the 56 cases of which details were given in the literature, no less than 36 had anaemia on admission (64%). Our patient was anaemic, she had sinus tachycardia, peripheral pitting oedema, raised jugular pressure, and a soft systolic murmur. A mass is palpable in the right upper quadrant in about 50% of cases (Starzl et al, 1960b and Table I). Jaundice is infrequent; in our review of the literature we found jaundice recorded in five of 57 cases (8.8%). This is surprising, because in about 52% of cases the tumour is situated in the second part of the duodenum, and also because this tumour is characterized by local invasion.

Radiological investigation of the gastrointestinal tract is a very helpful diagnostic aid. The majority of patients show radiological abnormalities such as a soft tissue mass, filling defects, duodenal obstruction, and ulceration. The presence of a sinus or fistula is very significant and almost specific; the sinus may be directed to the retroperitoneal space or form fistulous connexions with other viscera (Starzl, 1960a). Baker and Good (1955) thought that the presence of a fistula or sinus was pathognomonic of malignancy. Leiomyosarcoma of the duodenum is often mistaken for duodenal ulcer (Lie and Sinclair, 1966); carcinoma of the head of the pancreas (Starzl et al, 1960a), and pancreatic cyst (Waugh et al, 1963).

The tumour varies in size; it is generally firm but may be cystic and lobulated or globular. Often conspicuous blood vessels are present on the surface. Leiomyosarcoma of the duodenum is classified as endoenteric, intramural, exenteric, and dumb-bell according to its site of origin in relation to the wall of the duodenum, the exenteric type being the commonest (Lie and Sinclair, 1966). Macroscopically, the cut section is soft greyish yellow with a whorled appearance, often with central necrosis and haemorrhage. The tumour tends to outgrow its blood supply with the result that ulceration, degeneration, necrosis, haemorrhage, cavitation, sinus and fistula formation, and infection may occur. Calcification is also sometimes seen. Microscopically, a definite capsule is often absent, the cells are pleo-

![Site of leiomyosarcoma in different parts of the duodenum in 65 cases.](http://gut.bmj.com/ on October 30, 2017 - Published by group.bmj.com)
of the duodenum must be included in the differential diagnosis of a case of upper abdominal pain, gastrointestinal haemorrhage, anaemia, abdominal mass, and, occasionally, obstructive jaundice. Radiological examination of the gastrointestinal tract is in most cases diagnostically characteristic. Aggressive surgery may be required for extirpation of the neoplasm where the conditions are favourable but it should be stressed that it is a tumour of low malignancy. Prognosis is guarded.

We would like to express our gratitude to the secretaries in the Department of Surgery and the staff of the Medical Illustration Unit for their help. We also thank our colleagues in the Department of Morbid Anatomy for their advice.

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