Leiomyosarcomatosis of probable uterine origin with long survival—a case report

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Summary
A case of leiomyosarcomatosis is presented. Over a period of 15 years the patient underwent seven operations to remove eleven tumours, the largest as big as a football, before dying of widespread metastases: between operations the patient was remarkably well. The disease was almost certainly of uterine origin.

References
GLYNN, J.M. (1964) The action of cardiac glycosides on ion movements. Pharmacological Reviews, 16, 381.

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and consequent leakage of potassium from the myocardial cells (Glynn, 1964). Attempts to control the hyperkalaemia, including using haemodialysis, may be unsuccessful (Reza et al., 1974; Smith and Willerson, 1971). Little attention was paid to potassium balance in the present patient owing to the rapidity of events, but this probably made little difference to the fatal outcome. Other unfavourable factors in this case include the previous maintenance therapy, which may have saturated the drug-binding sites, and the relative intolerance of diseased myocardium to digoxin.

One of the commonest cardiac arrhythmias is bradycardia with varying degrees of atrioventricular block. Atropine may be tried, but pacing is usually required. In the presence of hyperkalaemia, a pacemaker should be inserted as a prophylactic measure even if the ECG is normal (Bismuth et al., 1973). However, in fatal cases there is often terminal resistance to pacing. In the present case, there was complete cardiac standstill despite direct electrical stimulation of the myocardium. This too seems to be due to failure of the myocardial cell membrane pump (Reza et al., 1974).

The recent description of digoxin-specific antibodies and their use in severe digoxin poisoning may be a significant advance (Smith et al., 1976); in the case they report, hyperkalaemia and pacing resistance were both reversed and the patient survived.

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the Whittington Hospital as an obese woman with small bowel obstruction. At emergency laparotomy, adhesions of small intestine to a fibroid were found to be the cause. A fist-sized fibroid was removed by myomectomy.

In 1962 she re-presented complaining of a swelling in the abdomen and suprapubic pain. Sub-total hysterectomy and bilateral salpingo-oophorectomy were performed, the uterus containing a huge fibroid 23 × 24 × 10 cm which was densely adherent to ileum.

In 1969 she was re-admitted because of abdominal pain and a mass was palpable per abdomen. A 17 × 14 × 11 cm retroperitoneal tumour overlying the pelvic brim and densely adherent to much of the small bowel was found and 'shelled out', no other procedure being possible.

In 1970, she complained of diarrhoea, and rectal examination revealed a polypoid mass projecting into the rectum. At laparotomy, a sessile lobulated tumour 10 × 7 × 5 cm was found lying in the lumen of the rectum (Fig. 1) and a Hartmann's procedure (removing the rectum via the abdominal incision, closing the rectal stump, and bringing out a terminal colostomy in the left iliac fossa) carried out. There were also three tumours of up to 5 × 2-5 × 2-5 cm lying in the mesentery and wall of the small intestine, and these were 'shelled out'.

In 1971 she developed an intestinal fistula through the previous laparotomy incision. Three tumours of 11, 7 and 5 cm diameter respectively were found involved with small intestine (Fig. 2), one of which had necrosed the anti-mesenteric border by pressure. The tumours and involved small intestine were resected.

In 1972, sub-acute small intestinal obstruction again developed together with iron-deficiency anaemia (Hb 6-8 g/100 ml). Obstruction was found to be due to a tumour 13 × 11 × 9 cm which was resected together with involved small intestine.

In 1973, at laparotomy for obstruction, a mass 16 × 10 × 10 cm was resected, together with small intestine and abdominal wall.

In 1974, she complained of further abdominal pain. In the epigastrium a mass was palpable and two other masses could be felt suprapubically. A coin lesion was seen in the left lung on chest X-ray. She grew progressively weaker and finally died in April 1974.

At post-mortem, there was a mass 18 cm in diameter involving the terminal ileum, and another 5-cm mass lay in the pelvis. The liver weighed 3700 g, and contained multiple secondary tumours, the largest of which was 40 cm in diameter and replaced the entire left lobe of the liver. A solitary 1-5 cm diameter metastasis was present in the upper lobe of the left lung. No lymphatic invasion was found, and no other secondary deposits. Microscopically, all the abdominal tumours, liver and lung deposits were leiomyosarcoma.

The histological sections of successive tumours have been reviewed recently. The tumours removed in 1960 and 1962 have the appearance of cellular fibromyomas, whilst the tumours removed in 1969 and subsequently are sarcomas of similar origin (Figs 3 and 4). The possibility that there was progression from cellular fibroid to frank leiomyosarcoma is clear, especially, as will be pointed out, when considered in relation to the clinical picture.

**Discussion**

The organ of origin of the leiomyosarcomatosis in this patient is not immediately obvious.
The clinical course suggested primary and subsequently metastatic leiomyosarcoma of the uterus, despite the fact that the uterine fibroids of 1960 and 1962 were considered histologically to be benign. In fact the border-line between benign and malignant smooth muscle tumours of the uterus is finely drawn, so that histology alone is often equivocal. Thus, whilst the most important features in distinguishing between leiomyoma and leiomyosarcoma are generally agreed to be (a) nuclear abnormalities and (b) mitoses, their relative importance vis-à-vis one another is contentious. Spiro and Koss (1965) invariably found nuclear abnormalities in leiomyosarcoma, but mitoses of variable occurrence, whilst, on the contrary, Taylor and Norris (1966) found the presence of mitoses the most reliable guide to malignancy. If the fibroids were sarcomatous, the fact that this was not diagnosed histologically at the time is therefore not surprising. Other factors have to be taken into account. For instance, Spiro and
Koss (1965) noted that if a uterine leiomyosarcoma was adherent to an adjacent organ, the malignancy was high grade behaviourally if not histologically and the outcome was invariably fatal: here the fibroids were adherent to ileum and the outcome likewise fatal. Silverberg (1971) similarly stressed the importance of gross appearance in assessing the degree of malignancy. It is also of note that whilst fibroids may cause intestinal obstruction, they do so normally by pressure as in Mowat and Miller’s (1973) case, rather than by adhesions, as in this.

Plainly, therefore, the fibroids of 1960 and 1962 could have been sarcomatous, and all subsequent tumours secondary to them. Lymphatic spread would have to be invoked to account for the retroperitoneal tumour; it is unusual but does occur in uterine leiomyosarcomas (Spiro and Koss, 1965); the rectal tumour would then be an implantation secondary in the pouch of Douglas, by seeding at the time of hysterectomy; and all other tumours implantation or blood-borne metastases.

The long time interval between the second operation, hysterectomy, and the third, for retroperitoneal tumour, might be thought to be a bar to this interpretation. But, a prolonged symptomless interval in uterine leiomyosarcoma is well known; for instance, Friedell (1962) reported a 5-year survival with known metastases. Laffargue et al. (1966), reporting a pulmonary metastasis 7 years after hysterectomy for leiomyosarcoma, noted latent intervals in reports of uterine leiomyosarcomas of 8 years (Laffargue, Cabanne and Nosny, 1966), 8 years (Stearns and Sneed, 1966), 8 years (Spiro and Koss, 1965), and 19 years (Drake and Dobben, 1959).

Similar cases, incidentally, are recorded amongst leiomyosarcomas of the alimentary tract, with perhaps the most remarkable of all, Hart, Soots and Yoshida’s (1972) case of 45 years’ survival with leiomyosarcoma of the stomach. First diagnosed at the age of 8 by vomited papillomatous material, operations for the tumour were performed at ages 10, 13, 16, and 27; between ages 37 and 52 she had two further laparotomies and several episodes of melaena, and finally died of the tumour. Starr and Dockerty (1955) reported a patient alive and well 19 years after resection of a leiomyosarcoma of the small intestine, with known metastases for 6 years.

The case of Hart et al. (1972) illustrates that leiomyosarcoma of the alimentary tract can become progressively more malignant, a point also made by Neuman (1952). This is less well documented in uterine tumours; but the change from benign to malignant, as postulated in this case, is well recognized (Novak and Anderson, 1937; Aaro, Symmonds and Dockerty, 1966).

It is clear that a uterine leiomyosarcoma could account for all the facts in this case. But it was never shown that the fibroids were malignant.

A second possible interpretation is that the history really begins in 1969 with the retroperitoneal tumour, which was either the primary, or secondary to the rectal tumour of the following year. But retroperitoneal tumours do not spread to the rectum; and rectal tumours do not usually spread to the peritoneum as, like this one, they are usually endo-enteric (Fig. 1). They also generally lie on the posterior or lateral wall in the lower half of the rectum (Morson and Dukes, 1960) whilst this lay on the anterior wall in the upper half, suggesting the much stronger possibility of implantation of uterine sarcoma at the time of hysterectomy in 1962.

Conclusion

Almost all published series of leiomyosarcomas are marred by insufficient length of follow-up; cases such as this and others mentioned above show that 10 years should be considered the minimum (Skandalakis et al., 1962).

Although the course of leiomyosarcomatosis in some cases can be measured in months, in those with a protracted course repeated palliative operations are justified and worthwhile. Despite the daunting list of seven operations to remove eleven tumours, the patient made a rapid recovery from each procedure, and in between spells in hospital lived a normal life; she did not lose weight, managed her colostomy with complete unconcern, and was uncomplainingly cheerful throughout.

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References


Staphylococcal septicaemia with disseminated intravascular coagulation associated with acupuncture

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Summary
A case of disseminated intravascular coagulation due to staphylococcal septicaemia is described in which the source of infection was likely to have been acupuncture therapy.

Introduction
The ancient oriental art of acupuncture has recently enjoyed a revival of popularity. Whatever the merits of this form of treatment, complications are occasionally reported including serum hepatitis (Hussain, 1974), pneumothorax (Carron, Epstein and Grand, 1974) and renal disease (Keller, Parker and Garvin, 1972). A further hazard is illustrated by the case reported here.

Case report
A previously fit 66-year-old man was admitted with a 2-day history of malaise and fever. Two days before the onset of symptoms he had undergone acupuncture treatment around the knees for longstanding osteoarthritis. During the night before admission he was found on the floor of his bedroom confused and rambling. Examination showed him to be pyrexial (38-5°C), drowsy and disorientated, but there were no localizing neurological signs. A soft ejection type systolic murmur was heard over the aortic valve. The skin, nails and abdomen were unremarkable. During the next 2 days his level of consciousness diminished and he developed mild generalized rigidity with extensor plantar responses. Several septic foci appeared in the skin together with widespread purpura and pyoarthroses of the left elbow and wrist.

Investigations on admission showed normal urine, blood film, haemoglobin, white cell and platelet count. Lumbar puncture yielded clear CSF under normal pressure containing 10 polymorphs, 40 RBCs/ml, and 0-53 g/l protein. Bilateral carotid arteriograms excluded intracerebral abscess or subdural haematoma. Eight blood cultures yielded growths of Staphylococcus aureus after 48 hr incubation. By the third hospital day, the haemoglobin had fallen to 9-3 g/dl and the platelet count to 34 × 10^9/l. During this time the blood film showed a progressive shift of the white cells to the left, with the emergence of crenated (70%), distorted or fragmented red cells. Plasma fibrin degradation products rose from 16 mg/l to 64 mg/l (normal range <10 mg/l). The blood urea rose to a peak of 34 mmol/l. HbAg was negative and immunoglobulins normal. Other investigations were non-contributory.

Treatment was started on the third hospital day