RETROPERITONEAL TUMOURS
A Study of Five Cases

J. A. MUSCAT, M.D., F.R.C.S.
Senior Surgeon
MARIE THERESE PODESTA', M.D.
House Surgeon
St. Luke's Hospital

Retroperitoneal tumours and cysts are often regarded as a nebulous group of lesions with a sinister reputation, probably through their uncommon incidence coupled with the fact that their potential site of origin, is anatomically extensive and may be remote. The many and varied pathological classifications put forward in the literature add little to help one form a sharper mental picture of the clinical problem they pose. The present communication is a study of five cases which have all come under our care in the space of a few months.

Case 1. P.B. Male aged 29.

First admitted to hospital in Melbourne, Australia in February 1970 with a history of progressive loss of appetite coming on in December 1969 followed, within a month, by continous pain in the back felt on both sides but worse on the left. He also complained of epigastric pain felt immediately on eating. Three weeks previous to admission he had had a bout of melaena, and four days before admission he had haematuria with passage of clots per urethram. There was no haematemesis, no haemoptysis and no chest pain or cough.

On examination he looked pale. There was no cervical node enlargement. Pulse 80, regular; B.P. 140/80.

There was good chest expansion, good air entry throughout and normal breath sounds. The heart sounds were normal.

Abdominal examination revealed a tender mass in the left loin. Special investigations were as follows:

Ba. Meal: No free oesophageal reflux or hiatus hernia. Stomach was reasonably well filled and outlined. No obvious organic lesion could be seen. Unable to fill the duodenal cap sufficiently to exclude pathological states.

Chest X-Ray — Pleural fluid left base. No obvious pulmonary changes.

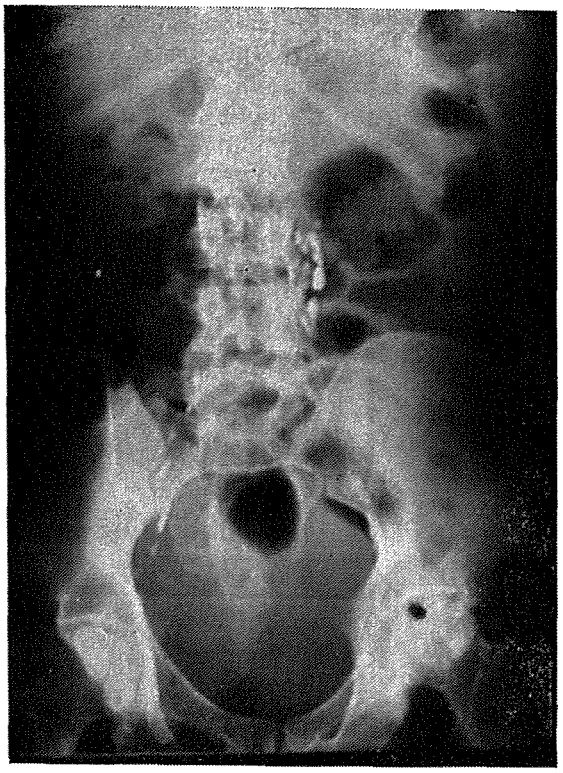
Hgb. 9 G., Prothrombin 80% Serum Bilirubin 0.3 mgm %.

At operation, preliminary cystoscopy, revealed blood and clots coming from left ureteric orifice. Laparotomy showed a large hard craggy retroperitoneal mass extending from the level of L.1 beneath the spleen, across the pancreas to beneath the liver. The mass was involving the left ureter and lymph nodes in the greater omentum. The small bowel was clear of tumour.

Biopsy material was reported upon thus: A large firm lymph node measuring 1.5 by 2 cms. Microscopically almost all the lymphoid tissue is replaced by diffuse proliferation of cells with large round to oval vesicular nuclei with prominent nucleoli. Mitotic figures are numerous. There is a fair amount of reticular formation. The tumour is also infiltrating the surrounding adipose tissue. Diagnosis: Reticulum cell sarcoma.

Following operation the patient developed melaena, the Hgb falling to 3.6 G. He was transfused with 8 pints of blood. He was later transferred to Peter MacCallum Clinic for Mega Voltage Therapy.

He was at this Clinic for a month from 25.2.70 to 24.3.70 Amongst the special investigations he had there was a lymphogram, reported upon as follows: A bilateral injection of contrast was made. The nodes and vessels in inguinal, iliac and para-aortic regions were outlined. The upper one third of the thoracic duct appeared moderately dilated but the significance of this finding was not evident. The iliac nodes on either side appeared within normal limits. Two groups of para-aortic nodes were demonstrated one on the right side opposite L2-L3 and one on the left opposite L3-L4. The nodal tissue appeared abnormal and involvement by the malignant lymphoma has almost certainly occurred. The absence of nodal tissue above the level of L3 on the left side would suggest surgical removal above this level. The appearance of the inguinal nodes on either side remains equivocal. Lymphatic stasis is demonstrated adjacent to these nodes and patchy filling is seen within them. These changes may be related to the malignant process or they may have resulted from previous infection.



Case 1: **Lymphogram**

On 14.3.70 he returned from weekend leave complaining of severe left chest pain. X-Ray chest showed further elevation of left hemi-diaphragm and partial collapse of the left lower lobe together with a small left pleural effusion. The appearances were considered to be due to a pulmonary infarct or consolidation, collapse resulting from bronchial occlusion. IVP showed kidneys normal in outline and function. The left pelvicalyceal system appeared attenuated and the upper one third of the left ureter appeared somewhat irregular. These appearances would suggest that early infiltration of the kidney and ureter by the lymphoma may have occurred but these findings are not diagnostic.

On 21.4.70 he was admitted to St. Luke's Hospital, Malta. He was then complaining of severe pain in low back and occasional vomiting. His general condition was poor. He showed wasting. No enlarged lymph nodes were present in the neck, axillae or groins. His abdo-

men was tensely distended, shifting dullness and a fluid thrill being present. There was bilateral ankle oedema. Hgb 81%, PCV 40%, WBC 16,400, ESR 40 mm. Chest X-Ray L. pleural effusion. On 22.4.70 nine pints of straw coloured turbid fluid were removed from his abdomen.

He was started on a course of i.v. cyclophosphamide and given opiates for the persistent pain. His subsequent course was relentlessly downhill. At no time did he show any response to the cytostatic drugs afforded to him and these included chlorambucil, vinblastine and prednisone.

There was reaccumulation of fluid in his abdomen and left chest necessitating repeated tappings.

His pain never left him and he complained bitterly of it as it affected the left lower chest wall which was often tender on palpation.

On 1.8.70 he developed paraplegia. On 25.8.70 he died.

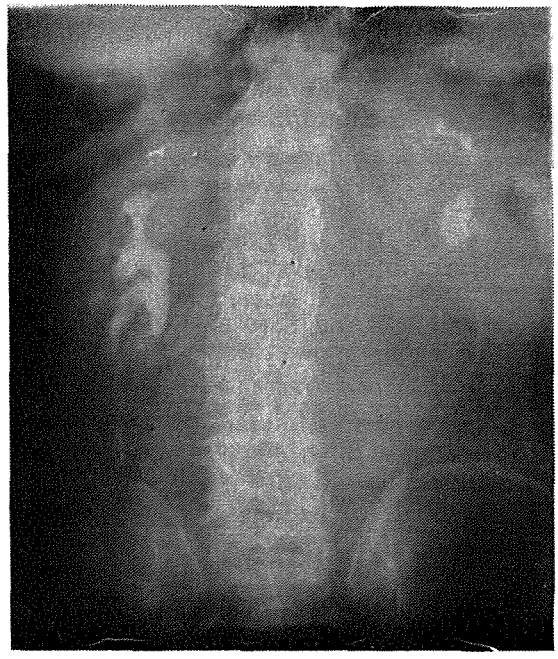
Permission for post mortem examination was not given.

Case 2. S.T. Man aged 74.

This patient presented on 22.7.70 complaining of a gradual increase in abdominal girth which he had noticed over the previous three months with "hardening" of his abdomen. There was no abdominal pain of any note but occasional mild dragging pain was present. His bowel habit was not disturbed though he did tend to being occasionally costive. His appetite had fallen off and he thought he had lost some weight. There was no nausea and no vomiting. He had never noticed passing any blood with his stools or his urine. He had no difficulty with urination but was experiencing a slight (3 to 5) nocturnal frequency in the last few months.

He was admitted to St. Luke's Hospital on 23.7.70. On examination he looked slightly pale and drawn; T. 99° F Pulse 100/min, regular and good volume. B.P. 195/100 There was no cervical node enlargement, and no jugular congestion Examination of the chest showed good air entry on both sides; some fine crepitations were audible over both bases. The apex beat was in the 6th interspace outside the mid-clavicular line. There was a soft systolic murmur localised to the apex and the left para-sternal region.

The abdomen was grossly distended. Its entire cavity seemed to be filled by a large, well defined, bilobular, rubbery, hard, non-tender mass which showed a fair degree of lateral mobility. Normal bowel sounds were audible. There was a left inguinal scar, no enlargement of inguinal lymph nodes. A right inguinal hernia was present. Rectal examination showed slight enlargement of the prostate.



Case 2: Showing displacement of left ureter

The following investigations were carried out: Hgb. 72% PCV 39% No proteinuria, no glycosuria, Blood Urea 14 mgm %.

Serum electrolytes Na-126 m.Eq., K-4.4 m.Eq., Plasma chloride 93 m.Eq./ Litre. Modified Glucose Tolerance Test: Fasting blood glucose 78 mg./100ml. 2 Hour Blood Glucose Level: 155 mg./100 ml. Urine glucose absent.

Faecal Occult Blood: Positive in 1 instance in 3. I.V.P.: Mass in mid-abdomen mainly to left side with irregular calcification anteriorly and to the right. Re-

troperitoneal, not connected to renal tract. Both kidneys concentrate the dye well. Left Ureter displaced. No abnormality in kidneys.

Operation was performed on 5th August 1970. G.A. Dr. J. Psaila, and Dr. Alex Galea.

Through a left paramedian incision a large solid and highly vascular retroperitoneal tumour was removed. It was seen to be arising from the anterior aspect of the abdominal aorta seeming to have burrowed into the sigmoidmesocolon. In the course of its dissection parts of the mesocolon and adjoining segment of colon were inadvertently contused but the bowel was not opened. Haemorrhage was at times difficult to control. A tube caecostomy was performed prior to closure with drainage. Six pints of blood were given over the operation and immediate post operative period. The tumour weighed $12\frac{1}{2}$ lbs. or 5750G.

On the third day after operation he developed intractable hiccup, and on the sixth day post-operation a faecal discharge was evident through the site of his tube drain. Meanwhile the caecostomy had already begun to function.

His further post-operative recovery was slow but unattended by other complications and he was discharged home with wound healed and a dry abdominal wall on 7th September 1970.

He was next seen at Follow Up Clinic on 26.9.70. His general condition had begun to pick up, he was eating better and had no amdominal pain or discomfort. He was however troubled by having to go to stood 6 to 8 times a day to pass soft normal coloured faeces. He was put on an opiate mixture.

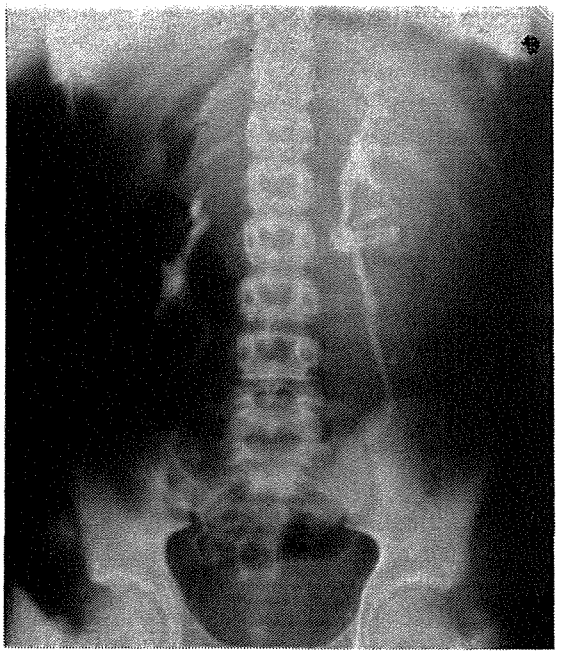
On 10th October when he next reported he complained of constipation, not having had a motion for four days.

He was now putting on weight and his abdomen was soft and not at all distended. A barium enema was asked for.

The barium Enema report dated 17.10.70 ran as follows: No signs of obstruction in the colon. There is however a fistulous communication in sigmoid region with formation of an irregular cavity and a long narrowed sinus. The outline of the colon is not much altered and the condition is probably due to an inflammatory process which has produced adhesions and perforation of the colon.

Case 3. P.B. Girl aged 12.

This girl was first referred to the Medical Division under the care of Dr. Luis Vassallo on 27.7.70 for abdominal pain and splenic enlargement. She had been quite well until about three weeks before admission when she first began to have intermittent fairly severe pain in left upper quadrant of abdomen. It was severe enough to make the patient take to bed. Her appetite was not impaired and there was no nausea or vomiting. Her bowels were regular. There was no loss of weight. Micturition was normal.



Case 3: Showing tooth rudiment

On examination she was a pale, timid, apprehensive girl. No lymphadenopathy was noted in neck, axillae or groins. Pulse Rate 112/min., B.P. 120/90.

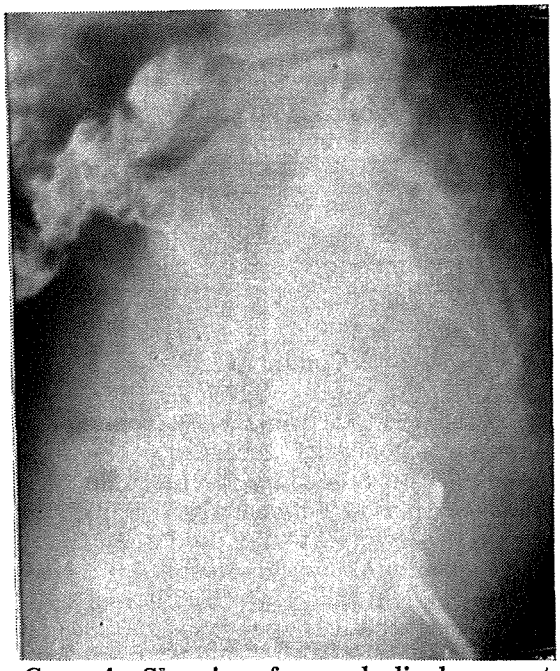
A large mass was palpable on the left side of the abdomen extending back to the loin. It was firm and smooth and dull to percussion. Her blood picture

showed a Hgb of 96% or 14.2G, W.B.C. 8,300, E.S.R. 5 mm. 1st hr., I.V.P. showed a mass in the left hypochondrium separated from the kidney and containing a toothlike structure, suggesting dermoid cyst.

Laparotomy was performed on 11.8.70 under general anaesthesia (Dr. C. Borg). A large retroperitoneal cyst arising from an area situated below the tail of the spleen was removed through a left paramedian incision. The abdominal wound was closed with drainage. She made a good post-operative recovery and was discharge with wound well healed on 30.8.70.

Case 4. Male F.G. aged 70.

27.8.70 Referred to Surgical Out Patient Clinic for "Haemorrhoids". This patient stated that he had been troubled with piles for some 11 years. His more immediate concern however was that of constipation. There was no history of prolapse or anal bleeding. On examination he was a well preserved florid corpulent man. There was no cervical lymphadenopathy. The chest was clinically clear.



Case 4: Showing forward displacement of rectum

The abdomen was well covered and no masses or organs were palpable. Rectal examination revealed a large smooth hard mass occupying the sacral hollow and abutting on the lower rectum. The rectal mucosa could be moved over it though the mass itself was completely fixed. The prostate was small and firm.

Investigations gave the following results: Hgb-100%, W.B.C. 9,900. Serum alk. phosphatase 8.7 K-A units, acid phosphatase 0.5 K.A. units. Blood urea 35 mgh%.

Barium enema showed a forward displacement of the rectum with a rounded indentation at the posterior aspect, suggesting a presacral mass.

Operation under general anaesthesia (Dr. F. X. Micallef) was performed on 13.10.70.

The tumour was explored through a sacro-perineal approach with excision of the coccyx and last piece of sacrum. It was found to be filling the true pelvis and to extend upwards to beyond the promontory of the sacrum. It was quite fixed to this latter bone and seemed to be arising from it. It was enveloped in a pseudocapsule which, on opening, revealed the tumour to consist of a gelatinous chondro-osseous mass. Most of this material was scooped out in the face of fairly brisk bleeding, the resulting cavity being packed.

A lateral left iliac colostomy was performed. His post-operative course was punctuated by bouts of severe hypogastric pain and retention. This was to some extent relieved by catheterization. He is now passing urine per urethram and relieved of pain.

The histological diagnosis was as follows "Several hemorrhagic portions of lipomatous tissue that measure $9 \times 7 \times 6$ cms. Section shows a myxomatous sarcoma infiltrating fat. Cellular pleomorphism is a prominent feature and mitotic activity is considerable."

He was referred to Dr. Sultana for radiotheraphy; but was however turned down because: "size of tumour, its histological nature, and the obesity of the patient contraindicated even an attempt at palliative radiotherapy."

Case 5. J.A.B. aged 3½ years.

He was referred to the Paediatric Wards under the care of Dr. T. Agius Ferrante on 29.1.69 with a 2 month history of abdominal enlargement accompanied by cyanosis. Clinical examination revealed ascites. There was no hepatosplenomegaly.

Paracentesis was performed on 5th February, 1969 and some 500 c.c. of chylous fluid was obtained.

Laboratory and X-Ray findings were as follows:

1. Urinalysis repeatedly normal.
2. Blood count and picture.

Hb: 13.4/100 ml: 91%. P.C.V.: 43%. 64%. Easinophils 4%. Basophils, Lymphocytes 24% Monocytes: 8%. Stained films: No abnormal features.

3. Bl. urea: 19 mg./100 ml.
4. Occult blood in stools: negative.
5. Agglutinins titration — negative.
7. The chylous fluid was examined

a) bacteriologically: no pathogens were detected.

b) biochemically and histologically:
Total fluid cholesterol: 85 mg./10 ml.
Total extractable fats: 2.4 g./100 ml.
Total fluid proteins: 4.3 gr./100 ml.
Fluid albumen: 3.2 gr./100 ml.

Electrophoresis revealed practically the same absorption of protein fractions in the fluid as in normal serum with only a marked lower concentration in the γ fraction.

The cytological appearance was homogeneous with the presence only of mononuclear cells belonging to the 'lymphocyte' series; scattered between the more numerous, larger and less mature lymphoid elements were many small mature lymphocytes, with homogeneous deeply staining nuclei suggesting the possibility of giant follicular lymphoma.

The child was discharged to out patients on 19.3.69 and readmitted on 21.4.71 with recurrent abdominal disturbance.

On 16.5.64, he was transferred to the Isolation Hospital because he had measles. He also had gross ascites which was embarassing his respiration. On 5.5.69, 500 c.c. of white chylous fluid were removed. He continued to leak for 3 days. On 25.5.69, he had recovered from his measles and was transferred back to the paediatric ward.

The patient was transferred to the Hospital for Sick Children at Great Ormond Street, London on 17.6.69 The possibility of a blockage of the lymphatic duct system was entertained. There it was found that the chylous effusion was not an ascites but a large cystic mass. The child was operated on by Mr. H. Nixon on 2.7.69. A large unilocular cyst about 8 × 6" was found situated in the transverse mesocolon and extending up posteriorly behind the pancreas. The cyst was between the leaves of the mesocolon. Its own wall was delicate and thin like the wall of a lymphatic vessel, so that it was impracticable to excise the cyst wall completely. The cyst was therefore marsupialised and two large tubes placed within it. The drains were left in for 14 days post-operatively, using suction for the first 4 days. There was no evidence of re-collection of fluid so the drains were removed.

He returned to Malta on the 20.7.70. Part of the operation wound was infected and kept discharging pus for some time. He was discharged well on the 29.7.70.

The diagnosis was of a large unilocular lymphatic mesenteric cyst and there was no evidence of any other abnormality of the lymphatic system.

Comment

The rarity of primary retroperitoneal tumours is generally accepted. The incidence may be gauged by the fact that over a 30 year period between 1930 and 1960, 101 cases were treated at the Lahey Clinic while at the Memorial Hospital in New York between the years 1926 and 1951 there were 120 verified cases. 30 cases were left unverified. This latter series which is the largest reported from any

one institution represents an incidence of 0.2% when one takes into account that throughout this 26 year period 60,000 patients with tumours were seen. Incidence seemed equal in both sexes.

The evil reputation which they have is on the whole well deserved. 85% of the New York series were malignant, as were 88 cases of the 101 from the Lahey Clinic. In Donnelly's series 91% were malignant.

The lymphomas constitute the largest group amongst the malignant neoplasms. Fully a third of the tumours reported from the Lahney Centre were of lymphatic node origin, while 24 of the 120 from the Memorial Centre were classified as lymphomas.

However of these, the reticulsarcomas form the least common type, the lymphosarcomas being the most frequent.

Most workers agree that the diagnosis is not easy. Enlargements of the kidney, adrenal, pancreas, spleen and liver have to be excluded, as also such lesions as aortic aneurysms. The tumours may arise from anywhere from the diaphragm to the pelvic floor, and from a wide veriety of tissue such as fat, areolar tissue, connective tissue, fascia, muscle, vascular tissue, nerve tissue, somatic and autonomic, lymphatic vessels and lymphatic nodes. Only a minority are hormonally active tumours. These include the extra- may here be made of the hypoglycaemia and its associated symptoms occasionally seen with retroperitoneal sarcomas. This was not seen in the two large series quoted above. (47 Fibro Sarcomas; 4 Neurofihomas).

Up to 1966, 145 patients were reported showing this phenomenon.

The most common *clinical* finding is an abdominal mass. Pain is generally present; it is ill localised, it may be felt in the back but is not often severe when the patient presents. Gastro-intestinal symptoms may be remarkably inconspicuous even in the patient with a huge abdominal mass as was seen in our second case. Anorexia, weight loss, and constipation are commonly complained of, less frequently vomiting and haematemesis. This latter may be the result of portal congestion from extrinsic compression. It is rarely due to direct involvement of stomach or intestine. Haematuria is a very rare early symptom; it was present in profuse degree in our first case.

With regard to special investigations all agree that the most useful radiological study is an intravenous pyelogram. AP and lateral views often show displacement of kidney and/or of the ureter.

A gastro-intestinal series is also very useful to show displacements and thus afford evidence as to precise location of the tumour. Newman & Pindi reckoned they were "sufficient to establish a diagnosis".

In our cases, I.V.P. did point to involvement of the left ureter in the first case and in the second the man had displacement of the left ureter and the diffuse spotty calcification afforded clues as to the nature of the mass in question.

Other X-Ray procedures which may provide highly useful information are aortography and lymphangiography. Perirenal insufflation is not without its dangers and pneumoperitoneum is not helpful. A preoperative diagnosis may be arrived at in about 30 to 40% of cases. However, there is no substitute for exploratory laparatomy to reach a definitive histological diagnosis in order to determine resectability.

Knowledge of the state of the renal tract, the previous lavage and sterilization of the intestinal tract and the availability of considerable quantities of blood are useful prerequisites to successful surgical treatment.

Our second case required to be transfused with 6 pints of blood over the operation and also necessitated a caecostomy as a safeguard following inadvertent contusion of the mesocolon.

The mortality rates reported in the larger series in the literature is not inconsiderable. A 10% mortality and a 22% morbidity rate was present in the Lahey Clinic series, among the malignant group.

Radiotherapy plays a primary rôle in the treatment of lymphomas. With other tumours, it has a subsidiary rôle. According to Pack and Tabak, Myxosarcomas are only slightly radio-sentitive. However, it

is generally considered that radiotherapy should be offered to all cases of tumours not amenable to resection and to those that are not for technical difficulties excised completely.

To conclude: primary retro-peritoneal tumours, though uncommon, are not great rarities and should not be neglected. They merit more notice than is generally given them in the odd half page of the standard surgical text book. They pose a difficult and fascinating problem to the pathologist and to the physician. To the practising surgeon they are a challenge that may tax his resource. In dealing with them, though he can often only bring to his patient temporary relief, he is occasionally rewarded with a gratifying outcome.