PRIMARY RETROPERITONEAL TUMOURS

A REVIEW OF TEN CASES*

HYMIE KATZ, CH.M.
Cape Town

Retroperitoneal tumours in general form an interesting group which, because of their anatomical position, often give rise to difficulty in exact diagnosis. The group includes a large variety of pathological conditions, but in this review only the primary retroperitoneal tumours will be considered. These are tumours which are found in the retroperitoneal space, but do not arise from organs in that area. The definition would therefore exclude renal, adrenal and pancreatic tumours. They are of interest to the general surgeon, gynaecologist and urologist alike, since they may may occur at any level between the floor of the pelvis and the diaphragm.

* A paper presented at the South African Medical Congress, Pretoria, October 1955.

Because of the appearance of this type of tumour over a wide area, the symptoms are variable and may simulate those caused by disease of the chest, kidneys or stomach and may lead even to a diagnosis of obstruction of the small bowel. As a rule the symptoms are caused by pressure on neighbouring organs, but often symptoms are minimal and the condition presents itself merely as a large abdominal mass.

These tumours are highly malignant and present a bad prognosis on account of their wide infiltration in the retroperitoneal space and their close relationship to vital organs. Wherever possible the tumour should be removed, for surgery offers the only chance of a cure.

In only 2 of my 10 cases (see below) was the tumour considered resectable.

The incidence of these tumours as reported by the Radcliffe Infirmary, Oxford,[2] is 1 in 12,000 admissions; 14 cases were described there in a period of 10 years. My own series, personally treated, comprises 10 cases in the past 7 years. Of these 10 cases 6 were male and 4 female. Most authors report a preponderance of females, except Donelly[1] whose figures correspond with mine—a proportion of 3 males to 2 females. The ages of my 10 cases varied from 7 to 73 years. The condition is found most commonly after the age of 50.

CLINICAL FEATURES

The vast majority of cases present as an abdominal swelling without any symptoms. As pain is not a predominant feature, the tumour has already attained a large size by the time the patient reports the condition. Only when neighbouring organs are involved, will pain be a prominent feature, and the symptoms, which are produced by pressure and depend on the situation of the tumour, are variable and simulate many abdominal conditions. They include:

(*a*) Symptoms referable to the *stomach*, e.g. nausea and vomiting, are seen with upper abdominal swellings.

(*b*) *Chest symptoms.* Praecordial pain is caused by a tumour pressing on the diaphragm (case 1).

(*c*) *Obstruction of the small bowel* is caused by adhesion to tumours in the region of the mesentery of the small bowel (case 3).

(*d*) *Renal symptoms.* Uraemia will follow blockage of the ureters from infiltration by the tumour (case 10).

(*e*) *Urinary bladder.* Frequency and dysuria may occur (case 4).

(*f*) *Referred pain* is caused by infiltration of a nerve plexus (case 10).

(*g*) *Oedema of legs* may result from occlusion of large veins.

(*h*) *Distant metastases* may produce symptoms (case 6).

The presenting signs are those of large tumours, either solid or cystic. The majority are immobile owing to fixation to the posterior abdominal wall. Occasionally mobility may be a feature, in tumours which have invaded the root of the mesentery of either the small bowel or transverse colon.

Special Investigations. Retrograde pyelograms are of the greatest diagnostic value and should never be omitted. The ureters being attached to the posterior parietal peritoneum can readily be displaced by a retroperitoneal tumour. A barium enema is also of great value and may reveal either displacement or indentation of the colon. For tumours of the upper abdomen, a barium meal may show pressure on the stomach.

Histology

The classification of these tumours is made difficult by their diverse histological appearances. According to Herdman,[2] it seems possible to recognise 3 main groups of tumours which can be identified by their histological appearances:

Group 1. *Tumours of adreno-genito-urinary origin.* These tumours are embryological remnants of the urogenital apparatus and their origin can be explained on the basis of foetal rests or displaced embryonic tissue. They may therefore resemble ovarian, kidney or adrenal tissue.

Group 2. *Tumours of connective-tissue origin.* To this group belong chiefly the lipomata or liposarcomata. As we know, lipomata very rarely undergo malignant changes except in the retroperitoneal space, where the incidence of malignancy is very high indeed. Some authors go as far as to state that lipomata in the retroperitoneal region are nearly always malignant. Pemberton[4] showed that 10 out of 30 cases of retroperitoneal lipomata showed malignant changes. More rarely leiomyomata and lymphangiomata have been described.

Group 3. In this the origin of the cyst is unknown and the wall has a simple epithelial lining or is composed of fibrous tissue.

Progress

The majority of patients are dead within 1 year. In the present series of 10 cases, 2 are living, one 5 years and the other 18 months after operation. They were both localized tumours and were resectable, the first case being a lipoma and the other a cystadenocarcinoma.

Treatment

Surgical removal will give the best results. Unfortunately the majority of tumours by the time of their discovery have a very wide extension, are fixed to the posterior abdominal wall and involve vital organs. In these cases surgery may be extremely hazardous and unjustifiable. Where the tumours are localized they should be removed, even at the expense of a kidney or a portion of the bowel. Sound judgment will be required in these cases, for the surgery is of a considerable magnitude. The transperitoneal route is usually preferred. A large number of tumours are radiosensitive and X-rays should be given in those cases where the tumour has been left *in situ*.

CASE REPORTS

Particulars of my series of 10 cases are set out below:

Case 1. February 1953: A.L., a well-built European male aged 55 years, weighing 215 lb, complained of praecordial pain for the past 4 months. He was thought to be suffering from coronary thrombosis, but the electrocardiogram did not support this diagnosis. Soon afterwards a large lobulated mass appeared in the left hypochondrium. It was of rubbery consistency and moved freely from side to side. He then developed dyspeptic symptoms, with poor appetite and loss of weight. Barium meal, however, showed no involvement of the stomach and later X-rays of the colon also proved negative. A laparotomy through a left-upper-rectus splitting incision revealed a large lobulated retroperitoneal tumour invading the root of the mesentery. This tumour was quite inoperable and the abdomen was closed after removal of gland for biopsy. Section showed the tumour to be lymphosarcomatous. After deep X-ray therapy the mass diminished considerably in size and pressure symptoms were relieved. Nevertheless the condition deteriorated and the patient died 9 months later with involvement of the glands in the mediastinum and root of the neck.

Case 2. November 1950: B.K., a European girl aged 11 years, complained of increasing adiposity specially confined to the abdomen. She felt perfectly well and weighed 128 lb. The abdomen was very prominent and the limbs were well developed. The skin

was normal in appearance with no hirsutes. A large tumour, filling the right loin and extending to the brim of the pelvis, could be felt. A barium enema showed the ascending and transverse colon displaced to the left. An intravenous pyelogram demonstrated upward displacement of the right kidney. Through a right paramedian incision, a large retroperitoneal lipoma was removed. It measured 8½ inches in its largest diameter and weighed 6 lb. Microscopy did not reveal any sarcomatous changes and the patient is alive and well 5 years later. This case has previously been reported, with illustrations.[3]

Case 3. August 1950: C. v.d. M., a European male aged 45 years, was admitted to hospital with symptoms simulating a small-bowel obstruction. A mass was not palpable clinically, but at operation a well-developed retroperitoneal tumour was found in the left iliac fossa. A loop of small bowel, adherent to the tumour was responsible for the intestinal symptoms. After freeing the adhesions, an attempt was made to remove the growth but was abandoned on account of the marked vascularity and friability of the growth. The tumour resembled a sarcoma and responded well to deep X-ray therapy. The patient was still alive when seen 2 years later.

Case 4. September 1945: A European soldier aged 43 years complained of marked frequency of micturition. A mass the size of a golf ball could be felt in the hypogastrium. Rectal examination revealed a normal prostate but on cystoscopy an extra-vesical mass could be seen indenting the bladder anteriorly. At operation, through a suprapubic abdominal incision, the tumour was found to be occupying the cave of Retzius. The tumour was not removed and its histology was that of a cellular sarcoma. It responded readily to deep X-ray therapy. The patient was still alive 10 months later.

Case 5. November 1954: Mrs. D.F., a European female aged 45 years, presented with a large abdominal swelling resembling a full-term pregnancy. She complained of abdominal discomfort from increased intra-abdominal pressure. She looked and felt well. A plain X-ray of the abdomen excluded pregnancy. At operation a large cystic tumour, the size of a rugby football, was found. At first it was deemed to be resectable but, owing to extensive retroperitoneal spread in all directions, the tumour was left *in situ*. It resembled a cystadenoma of the ovary with multiple loculations and extreme vascularity. Great difficulty was experienced in closing the abdomen. There was no response to deep X-ray therapy and the patient died 6 months later.

Case 6. February 1952: F.C., a European male aged 65 years, complained of a painful swelling of the costochondral junction of the 4th rib on the left side for 4 months. Apart from this swelling he had no symptoms. The swelling was the size of a golf ball, bony-hard and attached to the chest wall. He was thoroughly investigated and his lungs and gastro-intestinal and renal tracts were X-rayed for evidence of a primary malignant tumour. An X-ray of the ribs did not reveal any bony destruction or periosteal reaction. He did not respond to antibiotics or deep X-ray therapy. Four months later he developed symptoms of intestinal obstruction. His general condition had deteriorated but there was no abdominal swelling present. A barium enema revealed indentation of the sigmoid colon due to pressure from without. At operation a retroperitoneal tumour filling the left iliac fossa and pressing on the colon was found. The tumour was not removed owing to its fixity and wide infiltration. The patient died 6 weeks later.

Case 7. June 1955: Mrs. E.P., a European female aged 75, presented in a very similar manner to Mrs. D.F. (case 5). The swelling was situated mainly in the lower abdomen. She had lost 50 lb in weight and her general condition was poor. A large retroperitoneal mass involving the lower abdomen and pelvis was found at operation. It strongly resembled the tumour in case 5 and it was difficult to decide whether it was primarily ovarian in origin. The tumour was considered not resectable and the patient died 3 weeks after the operation.

Case 8. August 1954: G.H., a European female aged 54 years, presented with a swelling in the left iliac fossa of 4 months' duration. The swelling was the size of an orange, of rubbery consistency and movable from side to side only. It was difficult to decide whether the tumour was of pelvic origin or had arisen from the large bowel. A barium enema, however, suggested that there was no connection with the colon. As she was experiencing a fair degree of pain and as the diagnosis had not been established, an exploratory operation was performed. The abdomen was opened through a muscle-cutting incision as used for appendicectomy. A tumour was palpable in the left flank extending retroperitoneally. The posterior parietal peritoneum was incised laterally to the descending colon in order to open the retroperitoneal space. A greyish tumour, rather vascular, presented and was found to be attached by a broad pedicle to the posterior aspect of the descending colon. As it was virtually impossible to dissect this tumour from the bowel, a partial resection of 6 inches of large bowel was carried out and an end-to-end anastamosis performed. Drainage tubes were placed both intra- and extra-peritoneally, down to the resected portion of bowel. The patient made an uneventful recovery. The histology of the tumour showed features of a serous capillary cystadenocarcinoma resembling ovarian tissue.

Case 9. March 1955: H.P., a European male aged 72 years, was admitted with pyrexia of unknown origin which had lasted for 2 months. Examination revealed marked tenderness in the left loin but there was no evidence to suggest that this was of renal origin. A plain X-ray of abdomen and chest revealed that the diaphragm was markedly raised, suggesting a subphrenic collection. A blood count showed marked anaemia but no polymorphonuclear leucocytosis. At operation, through the bed of the 12th rib, a large retroperitoneal haematoma consisting of old blood-clot was found. The left kidney showed no signs of trauma or other pathology. The patient died 2 weeks later and at autopsy was found to have a large retroperitoneal tumour with degenerative changes. The haematoma had been caused by a large haemorrhage into the tumour.

Case 10. July 1955: R.B., a European boy aged 7 years, was admitted to the Victoria Hospital, Wynberg, complaining of pain down the right lower limb and severe constipation. He looked ill and was pale and undernourished. No pathological condition could be detected in the limb but a hard fixed mass was palpable in the right iliac fossa. In addition, a soft cystic intra-abdominal swelling presented in the umbilical region. Except for a mild degree of anaemia the blood picture was normal. On rectal examination a rock-like mass attached to the side wall of the pelvis was palpable. At laparotomy it was found that the mass practically filled the pelvic cavity, causing marked pressure on the rectum and displacing the bladder upwards towards the abdominal cavity, thus explaining the cystic mass which was present on examination. Similar retroperitoneal masses were found in the right iliac fossa and in the region of the 3rd lumbar vertebra. The tumour masses were deemed to be irresectable on account of their fixity and close proximity to the rectum and ureters. A biopsy report revealed a leiomyosarcoma, and deep X-ray therapy was recommended. The pain down left limb could be explained on the basis of sciatic nerve involvement. There was no response to deep X-ray therapy to the primary masses and the child died 4 months later.

SUMMARY

Retroperitoneal tumours are an interesting group of tumours occurring in the retroperitoneal space. Only the primary tumours, which have no relationship to organs in the retroperitoneal region, have been described. They are of interest to the surgeon, urologist and gynaecologist on account of their wide distribution. A series of 10 cases of primary tumours is described. They are highly malignant and offer a poor prognosis. Only 2 cases out of the 10 have lived longer than 18 months. The histology is extremely difficult and an attempt has been made to classify the tumours on histological grounds. These tumours present in a protean fashion with a wide variety of signs and symptoms, and they are facinating subjects for the clinician's diagnostic acumen.

REFERENCES

1. Donelly, B.A. (1946): Surg. Gynec. Obstet., **83,** 705.
2. Herdman, J. P. (1953): Brit. J. Surg., **40,** 331.
3. Katz, H. (1950): S. Afri. Med. J., **24,** 917.
4. Pemberton, J. de J. and McCaughan, J. M. (1933): Surg. Gynec. Obstet., **56,** 110.