RETROPERITONEAL TUMOR

case report with a special comment on the technique of surgical removal

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Two cases of neurofibromas, one fibrolipoma and one fibroma of the retroperitoneal origin have been presented. Primary retroperitoneal tumors are uncommon, the majority being sarcomatous. Benign tumors as presented here are very rare. Clinically, it may be asymptomatic or may be responsible for obstructive dystocia or may simulate an uterine fibroid or an adnexal tumor, thus, presents problems in differential diagnosis. Technically, surgical excision is not always easy and even benign tumors tend to recur after apparent complete excision, hence, nearly all retroperitoneal tumors should be considered either actually or potentially malignant. We found it better to enucleate the tumor from its capsule and then resect the capsule.

Retroperitoneal tumors are usually defined as tumors that arise from retroperitoneal tissues such as connective tissue, adipose tissue, muscle fascia or nerve tissue[7,10,16]. Though Lobstein (1829)[10] is credited with applying the term, the first patient with a retroperitoneal lipoma was reported by Morgagni in 1761[15]. Some authors[5] have applied the term "Middeldorpf" tumors to those tumors of many pathologic varieties in the region ventral to sacrum, though what Middeldorpf[14] described in 1885 only indicated sacrococcygeal teratomas originated from the postnatal hingut.

Pathologically, various entities are encountered; fibroma, fibrosarcoma, myxoma, myxosarcoma, ganglioneuroma, haemangioma, lymphangioma, lymphoblastoma, reticulosarcoma, leiomyoma, leiomyosarcoma or dermoid cyst has been described. The most common types described in the literature are sarcoma, lymphoma, lipoma and neuroblastoma[9]. Benign tumors, such as neurofibroma, fibrolipoma or fibroma, are rather uncommon. Two cases of neurofibromas. one fibrolipoma and one fibroma are presented in the present paper.

CASE PRESENTATION

Case 1 Neurofibroma

A 38 year old woman, gravida 1, para 1, was admitted on May 12, 1962, because of a tumor mass in the lower abdomen. She has had urinary frequency, difficulty in urination since November 1960. Shortly after the episode, she has found by herself that a tumor mass was palpable in the lower abdomen. In May 1961, she had had an exploratory laparotomy for it; however, the retroperitoneal location deterred the surgeon from dissection of the tumor. As the tumor got increasing in its size, difficulty in urination became worse and obstipation developed in addition.

Physically she was a moderately developed and poorly nourished woman with chronically ill appearance. B. P. was 132/80 mm Hg. Pulse rate was regular and 72 per min. Lung field was free from râles and apical systolic murmur was audible. Breasts were normal. A lower median longitudinal scar was visible on the abdominal wall. A child head sized elastic firm, nontender mass was palpable in the lower abdomen. Pelvic examination revealed that the tumor had trespassed on the pelvic basin and pushed the slightly enlarged uterus with its adnexae anteriorly.

Laboratory examination showed nothing abnormal except for the increased ESR.

Laparotomy was carried out under general anesthesia on May 15, 1962. A huge retroperitoneal tumor was found to push the uterus and the sigmoid anteriorly toward pubis and was densely adherent to the posterior wall of the sigmoid. The capsule of the tumor was highly vascular and parts of which were made up of mesenterium and of the sigmoid. Enucleation of the tumor accompanied with partial resection of the sigmoid, then resection of the capsule was done. Her postoperative course was uneventful and she made a fair recovery.

Pathological examination of the specimen revealed a whitish pink, well encapsulated ovoid solid tumor, measuring 16×13×8 cm in size, weighing 960 gm. On cutting, it was fibrous homogenous with tiny areas of mucoid degeneration. The tumor had a fibrous capsule, of which inner surface was uneven with tiny polypoid masses. Microscopic section showed a picture of neurofibroma. At the peripheral part, several cells with hyperchromatic large nucei were seen, but no definite sign of malignancy was noted. The polypoid masses on the inner surface of the capsule were found to be lymphnodes.

Case 2 Neurofibroma

A 28 year old housewife, gravida 1, para 1, was admitted on Dec. 12, 1962, because of a pelvic retroperitoneal tumor which was found occasionally at the time of cesarean section by an obstetrician 2 years ago. She had been healthy and free from complaints since the operation, though the tumor was not removed at that time. Several days prior to admission, she had consulted the same obstetrician for surgical sterilization, but the doctor had considered that it was better to remove the tumor since it has been growing in size, thereby, he referred the patient to us.

Physically, no particular abnormal signs were found except for the pelvic findings. B. P. was 120/70 in mmHg. Bimanual examination revealed an anteverted, normal sized uterus and negative adnexae. An over fist sized hard tumor was palpable at the right latero-posterior aspect of the uterus.

Laboratory data were within normal limit except for the increased ESR.

Laparotomy was performed under general anesthesia on Dec. 25, 1962. An over fist sized tumor locating in presacral retroperitoneum was extirpated. Postoperative course was uneventful.

Pathological examination showed a solid globular mass, measuring 8 × 7 × 6.5 cm in size, weighing 220 gm, with fibrous adhesion. On cut surface, it was solid, fibrous and pinkish white in color. Microscopically the tumor was found to be a neurofibroma.

Case 3 Fibrolipoma

This is a 51 year old nulliparous, menopausal woman, admitted on July 20, 1968 because of lower abdominal distension and edema of lower extremities for about past 3 months. Family history and past history revealed nothing contributory.

Physically, the patient was an emaciated, moderately developed woman with clear consciousness. Her B. P. was 110/70 mm Hg. Breathing sound was vesicular without râles. Soft systolic murmur was audible at the apex. The abdomen was distended but no definite signs of ascites were noted. Margin of the

liver was palpable. By pelvic examination, a child head sized mass was palpable at the right aspect of the uterine body which is retroverted and normal in size. Both adnexae were negative.

Laboratory data were within normal limit. Barium meal and enema study of gastrointestinal tract revealed displacement of stomach, duodenum entire small intestines, ascending colon to the left lateral aspect of the abdomen by an extrinsic lesion. E. K. G. was within normal limit.

Laparotomy was done under general anesthesia. on Aug. 23, 1968 A huge lobulated retroperitoneal tumor, occupying pelvic cavity and extending upward, reaching the upper abdomen was found. It was impossible to remove the tumor en masse, thereby, it was divided into several pieces and excised step by step.

Pathologic examination of the specimen showed six large tumor masses, measuring up to 24×17×8 cm in size and weighing 4450 gm in total. The tumor was yellowish and greasy. On cutting, it was made up of entirely yellowish fat tissue but rather rubbery firm than ordinary fat tissue. No sarcomatous area was found grossely on the serial cutting. The uterus was rather distorted by two protruding myomatous nodules on its anterior wall. Microscopically, the tumor revealed a picture of fibrolipoma without evidence of liposarcoma.

She had been healthy until Summer of 1970 when she had found that her abdomen was increasing in size again. She had consulted another university hospital and had a second laparotomy on July 23, 1970. A palliative surgery with biopsy revealed a recurrence of lipoma. She was readmitted into our hospital on Nov. 24, 1970 with complaints of dyspnea and distended abdomen. Physically, the patient was markedly emaciated and dyspneic because of accumulation of ascites. Laboratory ex. revealed; R. B. C. 215×10^4, W. B. C. 15,600, N. P. N. 32, B. U. N. 14, Protein 6.29, A/G ratio 0.31, Takata T 4, CCF 3+, GOT 21, GPT 25. E. K. G. showed sinus tachycardia, low voltage QRS complex with nonspecific ST-T changes. Chest × ray film showed marked elevation of right hemidiaphragm. The cachectic codition remained unimproved in spite of our supportive therapy and she expired on Feb. 14, 1971.

Case 4 Fibroma

This was a 39 year old housewife, gravida 5, para 5, admitted on May 4, 1971, because of a lower abdominal tumor. She had noticed a tumor mass in the lower abdomen for years, however, no particular attention was paid, since it causes little discomfortness, until several months ago when she noticed that the tumor has been growing rapidly in size and consulted a surgeon. External irradiation with Co^{60} was instituted under a diagnosis of malignancy. The patient was referred to us because the irradiation showed no effect.

Physically, the patient was an emaciated woman with chronically ill appearance. Her B. P. was 130/80 mm Hg. Chest; negative. Lower abdomen was occupied by a child-head sized tumor with brownish pigmented skin due to external irradiation. By bimanual examination, the tumor was located behind the corpus uteri, connecting with the posterior abdominal wall, firm and fixed.

Laboratory data were within normal limit. Upper G I study revealed nothing abnormal but barium enema study suggested an extrinsic lesion compressing the colon. Chest X ray film, negative. E. K. G., normal with negative exercise test.

Laparotomy was carried out under spinal anesthesia on May 6, 1971. A large tumor was located behind the uterus, simulating a pseudointraligamentary ovarian tumor, however, after a careful searching, an elongated ovary of the ~~left~~ side was found on the lateral surface. The tumor seemed to be

arising from the first sacral bone with a broad short pedicle. Removal of the tumor, simple hysterectomy and bilateral adnexectomy were done.

Pathological study of the surgical specimen showed that the tumor was 17×15×10 cm in size and weighed 1100 gm. The outer surface was moderately hemorrhagic with fibrous adhesion. On cutting, it was an well encapsulated solid tumor, made up of myxomatous fibrous tissue, with some focal calcification. Microscopically, the tumor showed a picture of fibroma with marked fibrosis of the peripheral portion of the main tumor probably due to irradiation. No evidence of malignancy was noted.

Postoperative course was uneventful except for a episode of cystitis which was overcome by antibiotic therapy.

COMMENTS

Primary retroperitoneal tumors are rare in incidence. Lovelady et al[11] have found about once in every 10,000 female admissions at Mayo Clinic; they have collected 127 cases during a period of 38 years. Pathologically, these tumors were divided into 4 categories; congenital (57%), neurogenic (9%), osseous (14%) and miscellaneous groups. However, the classification, as adopted by Johnson et al[9]; into malignant and benign groups is more practical since the former group is much more common than the latter group. Of 72 cases collected by Johnson et al during a period of 22 years, 56 or 78% were malignant and the majority being sarcomatus (45 cases or 62%). Sarcomas account for 37.6 of 138 cases reported by Schmid[23] and account for 68 or 34.5% of 197 cases reported by Ogihara et al[20]. On the other hand, presacral neurofibroma is very rare. In 1962, Nucci and Beyer[18] presented one case as well as nine cases collected from the literature, including 6 cases from the Lovelady's series[11]. Johnson et al[9] reported one in 72. Several cases have been described by Japanese authors[7,8,21] under a name of neurinoma. None of such instances were included in the large series reviewed by Melicow[12], North[17], Pack and Tanban[22] or Ogihara et al[20]. In spite of the fact that lipoma is one of the common benign retroperitoneal tumors, fibrolipoma is regarded as one of the rarest tumor. Only one case was noted in the series of 183 benign tumors collected by Ando[1]. Fibromas are also rare; one in 72 cases of Johnson's series and one in 127 cases of Lovelady's series[11].

Clinically, the retroperitoneal tumor presents no pathognomonic symptoms or signs. They may be symptomless for many years[8]. As the tumor increases in size, symptoms or signs due to compression of the neighboring structures by it will appear. For examples, compression to bladder and ureter induces frequency, dysuria, oliguria or even anuria; to intestines, nausea, vomiting, obstipation or frank intestinal obstruction; to blood vessels, edema, varicosity, of lower extremities or external genitalia; to nerve, neuralgia, numbness, or other motor and/or sensory disturbances. It may occasionally obstruct the birth canal at the time of delivery or it may simulate an utrine fibroid or an adnexal tumor, thus, presents problems in differential diagnosis. Occasionally, it may be responsible for menstrual aberration, hypermenorrhea, irregular bleeding, abortion, etc. in the first patient, her symptoms were mainly of urological, caused by the compression of urinary bladder or ureters. In the second patient, her numbness, sore pain and tingling in the right buttock, dorsum of the foot and lateral aspect of the right leg sugggested that the posterior division of the right first sacral nerve being inolved and compressd by the tumor[2]. The tumor was also responsible for her dystocia which necessitated a cesarean section.

Diagnosis is not easy; the tumor may simulate more common lesions[4]. Melody[18] gives some excellent diagnostic points in an article on presacral tumors by soft tissue technique. Rectoabdominal examination is essential. Roentgenographic examination always give more valuable information. The plain X ray film and lateral soft tissue X ray series are advisable in all pelvic masses. G I series determine possible connection or displacement of intestinal coils in a way to show deep relations of the mass. Intravenous and retrograde urography and presacral retroperitoneal pneumograms[6] provide better visualization of extragenital pelvic tumors and help in differential diagnosis from urologic conditions. Most retroperitoneal tumors located ventral to the sacrum give no definite signs by which they may be diagnosed correctly. They are usually diagnosed as retroperitoneal tumors on operation. In a series of 267 cases, Schmid[23] reported an accuracy of preoperative diagnosis was only 17%. Ando[1] indicated that 37 cases or 42% of 88 cases were diagnosed correctly prior to operation. In the present 4 cases, no definite diagnosis was made prior to the operation; the diagnosis was established by the surgeon during the first laparotomy in the first and second patient. As to the third and fourth patient, we have suspected the retropertoneal origin of the tumor, however, we were not sure about it prior to the laparotomy. The final diagnosis of tumor type depends on pathological examination. All surgical specimen of the present 4 cases were studied pathologically at the Department of Pathology, Taipei Medical College.

Treatment of the retroperitoneal tumor is mainly of surgical; however, it is often technically impossible or difficult to accomplish a total excision without giving injuries to adjacent structures, such as, urinary tract, blood vessels, intestines or nerves. As some of these tumors have a highly vascular capsule, alarming bleeding, even fatal, at the time of removal may occur[18]. We found it better to enucleate the tumor from its capsule and then resect the capsule. Sometimes, a part of intestines or vessels should be sacrificed. Irradiation or chemotherapy is suggested for those inoperable or incomplete surgery, or for prevention of recurrence in case of malignancy[1].

The prognosis is very grave for those malignant categories; in Johnson's series, only 21 of the 56 malignant tumors were resectable and there was only 3 five year survivals. The remaining 35 cases had palliative therapy, including radiation or chemotherapy, and there were no five year survivals in this group. The prognosis is better for those benign tumors, however, it has been emphasized by Johnson et al[8] that recurrence of benign tumors after apparent complete excision are not uncommon. The unfortunate clinical course of the third patient with fibrolipoma presented here reminds us to consider that nearly all retroperitoneal tumors are either actually or potentially malignant, as suggested by Johnson et al[9].

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後腹膜腫瘤

病例報告及手術方法之介紹

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後腹膜腫瘤係指發生于後腹膜腔內之各種腫瘤，以淋巴腫，肉腫，脂肪瘤爲常見。本文報告2例神經纖維瘤，1例纖維脂肪瘤及1例纖維瘤。第一1例係38歲一次經產婦訴有下腹部腫瘤，頻尿，排尿困難，經某一外科醫開腹，但不能切除，轉送到本院再開刀切除神經纖維瘤，第2例係28歲一次經產婦，困難產而作剖腹產時發現後腹膜腫瘤，但沒有切除，轉到本院後切除，係神經纖維瘤。第3例係51歲未產婦有一巨大脂肪瘤，開刀後2年又再發，由另一醫院再開刀，以後情況不佳，腹水積留，陷入惡液質狀態終于死亡。第4例39歲5次經產婦在腹部惡性瘤診斷之下，接受鈷60照射18次後轉入本院，開刀後發現係一復後膜瘤而加以切除，是爲纖維瘤。後腹膜瘤大部分爲惡性，良性者甚少。縱係良性，雖經切除亦常有再發之趨勢，不可大意。由於其特殊部位，外科切除常呈困難，但如先作 enucleation 然後再切除其 capsule 則可達目的。

（上述4例皆由徐千田教授開刀，而病理檢查係煩請臺北醫學院病理科陳定堯教授所做，在茲謹表謝意。）