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Lymphangiomas Involving the Ovary

Report of a case and review of the literature

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Summary

Lymphangioma involving the ovary is an extremely rare tumor. There are only 8 cases documented in the literature. This report describes a case of peritoneal lymphangiomatosis which presented as an apparent ovarian tumor and rapidly recurred and spread into adjacent and distant peritoneal organs following oophorectomy. We will comment on its complete response to irradiation treatment and review the literature.

Key-words: Lymphangioma – Ovary – Peritoneal Lymphangiomatosis – Irradiation

Introduction

Lymphangiomas are benign and uncommon tumors of the lymphatic system, composed of multiple cystic spaces lined by endothelium and separated by collagenous septa. They are classified as capillary, cavernous, or cystic. They may be unilocular or multilocular and contain serous or chylous fluid (1, 2, 3, 4, 5, 6, 7). Lymphangiomas involving the female genital organs usually are noted as incidental findings at surgery or autopsy, but occasionally may attain a size sufficient to cause symptoms by displacement, compression or involvement of adjacent structures (8, 9).

Lymphangiomas of the ovary are among the rarest tumors encountered in the field of gynecologic oncology. In most reference texts the condition is either excluded or dismissed briefly (11, 12). From a careful search of the medical journals, it is evident that only isolated case reports are available. The literature contains only 8 cases wherein both the gross and microscopic description will allow such a diagnosis (13, 14, 15, 16, 17, 18, 6). We report a case of peritoneal lymphangiomatosis which presented initially as an ovarian tumor.

Case report

The patient is a 27 year old white female gravida I para I who was first admitted to the hospital on March 25, 1972, because of recurrent episodes of severe right lower quadrant pain. Physical examination including pelvis and adnexal evaluation was within normal limits. Urinalysis was normal; the white cell count was elevated with a shift to the left. With presumptive diagnosis of chronic appendicitis, the patient underwent laparotomy and exploration with the following findings: the right ovary adherent to the appendix was enlarged and studded with small cysts. The adjacent omental peritoneum and serosal surface of cecum and were also irregularly studded with similar cysts. Appendectomy, right oophorectomy, and distal right salpingectomy were performed. The resected right ovary measured 4 x 2 cm; the surface was covered by small cysts which were smooth walled and contained a clear fluid; cut sections at the cortical surface showed the presence of what looked like dilated lymphatic spaces lined with endothelium.

One and one half years later, October 1973, the patient had a pelvic mass considered to be probable ovarian carcinoma and underwent repeated exploratory laparotomy. Upon entering the peritoneal cavity at the level of the pelvis, there was a large thick mass composed of hundreds of small cysts adherent to the parietal peritoneum from the level of the um-

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bilicus to the bottom of the pelvis. The small cystic masses also covered various pelvic organs including the dome of the bladder and the remaining left tube and ovary. A total hysterectomy and left salpingo-oophorectomy and omentectomy were performed. The last consisted of a 22 x 12 cm mass of mesenteric fat diffusely infiltrated with multiple confluent small cysts containing a clear yellow fluid. Attached was a 9 x 7 cm ovary with a ragged, coarsely granular multicystic surface.

The patient did well again for several months until April, 1974 (two years after the initial diagnosis), when she developed a sensation of fulness in the bladder, right-sided pain, and noticed the presence of a mass in the lower abdomen. On admission to the hospital, she was found to have a subumbilical firm, tender, movable mass. Chest x-rays, I.V.P., upper G.I. series and barium enema were normal. Pelvic ultrasound scan revealed bilateral irregular multiloculated masses. Abdominal exploration on May 6 showed a large soft cystic grape-like tumor filling the pelvic basin and implanted in the parietal peritoneum laterally at the level of the bladder. The cystic lesions could also be observed along the ascending colon to the level of the hepatic flexure, at the level of the line of excision of the omentum, in the posterior peritoneum, in front of both kidneys, at the splenic hilum, and on the peritoneum at the level of the pancreas. Only a biopsy was obtained and the procedure was terminated.

Histopathology: All three surgical procedures produced similar lesions consisting of 1–6 mm cysts reminiscent of hydatids. They were situated in proximity to serosal surfaces. The cysts had an inner smooth wall and contained granular pink proteinaceous precipitate. The lining was a single layer of uniform, mature endothelial cells without mitoses, pleomorphism or atypical features. No smooth muscle occurred in cysts wall. Background stroma was loose fibrovascular tissue with a variety of small lymphatic and blood vascular structures (Figures 1 and 2). Small lymphoid tissue aggregates were widely disposed. The pathologic diagnosis of each sample was "lymphangioma"; no evidence of sarcoma was present.

In view of the progressive nature of the growth and extensiveness of involvement, and the futility of any further surgical approach, it was decided to treat her with roentgentherapy. The whole abdomen from the bottom of the pelvis to the dome of the diaphragm received a total of 2600 rads measured at the mid-plane using the strip technique (19). Both kidneys were blocked posteriorly and the liver anteriorly for part of the treatment doses. The pelvis received an additional 1400 rads mid-plane dose in 7 daily equal fractions. The irradiation course

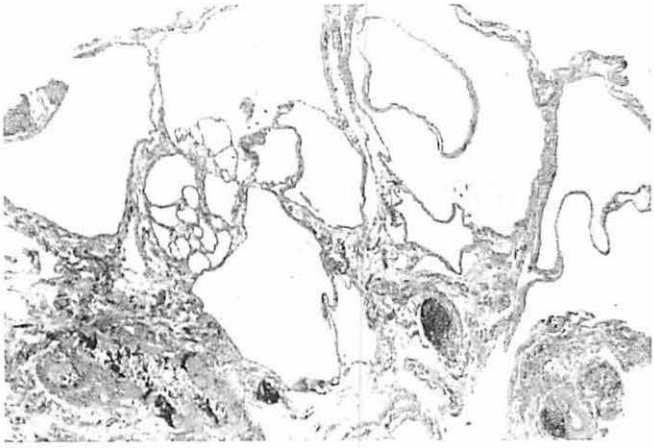

Fig. 1 Specimen from third laparotomy showing congery of cystic spaces and dark areas of lympoid tissue. H & E; 37X.

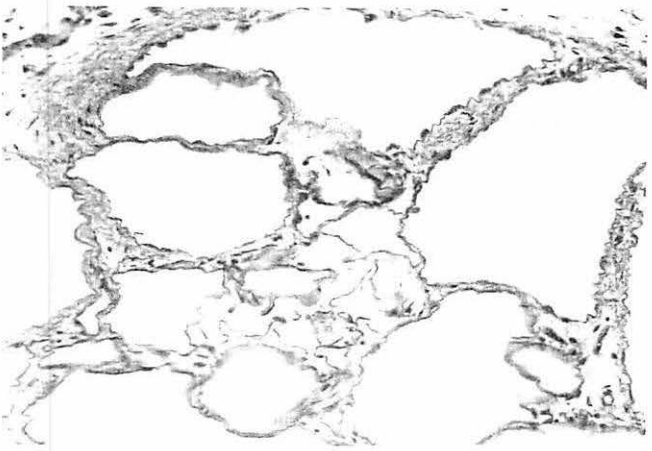

Fig. 2 Same as Figure 1, showing uniform character of the lining endothelial cells. H & E; 252X.

was delivered with a 4 MeV Linear Accelerator 80 cm source skin distance. It was started on May 21 and completed on July 24, 1974.

It was our impression that during the first 4 weeks of treatment, the pelvic component of the tumor increased and then steadily decreased in size and was barely palpable 3 weeks after completion of the irradiation course. However, 6 weeks later, there was again an ill-defined palpable right adnexal mass 8 x 10 cm in size. There was no enthusiasm on the part of the gynecologist or radiation oncologist for any further surgical or irradiation treatment; the patient refused chemotherapy.

Over the ensuing weeks, the palpable mass disappeared, and the patient returned to her full-time job. She has since remarried, is asymptomatic and enjoying a normal active life as of February of 1978, 46 months after the last operation. This has been the longest interval free of symptoms and findings since her initial exploration in 1972.

Discussion

Lymphangiomas in young patients are more commonly seen in the peritoneum and mesentery than pelvic organs. The precise origin of lympangioma in this patient is not certain. At initial exploration the ovary was most severely involved although adjacent peritoneum (appendix omentum) also had small lesions.

The infrequency of lymphangioma of the ovary is reflected in the small number of published reports. The first documented cases were 2 patients of *Kroemer* (16) in 1908. Additional single cases were recorded by *Fleisher* (14) in 1923 and by *Siddall* and *Clinton* (6) in 1937. A case in which malignant changes were present was documented by *Rice* et al. in 1943 (18). *Hartz* (15) described one more benign case in 1945, and another was reported by *Ferrary* and *Angelis* (13) in 1953. In a recent study on the relative frequency of the various ovarian tumors in children and adolescents, *Norris* and *Jensen* foung only one lymphangioma of the ovary among their cases (17).

Kroemer (16) doubted their extreme rarity and expressed the belief that many had been overlooked because of the lack of distinctive gross appearance. But on the other hand, *Meigs* (11) states that they had never seen on lymphangioma of the ovary in the pathological material from the Massachusetts General Hospital.

Lymphangiomas usually are diffuse non-tumorous lesions, but may produce localized tumorous masses. The most common example of the tumorous lymphangioma is the cystic hygroma, generally located in the neck region. The growth history of lymphangiomas is unpredictable although usually slow in growth, and proliferation ceases after a

period of time; occasionally renewed growth recurs. Histologically, the lymphangioma is typically benign despite the seemingly infiltrative growth and multicentricity. Recurrences after incomplete excision and mechanical complications may simulate malignancy. Lymphangiosarcoma is not a common association, although two of the eight cited ovarian lymphangiomatous lesions may have had a sarcomatous transformation (*Rice* (18), and *Kroemer* (16)).

In relation to treatment, there seems to be a consensus that the primary treatment of lymphangioma in general and of intra-abdominal lymphangiomas in particular should be a surgical approach (20, 3, 21, 4, 5, 22, 7). Usually complete excision can be carried out with relative ease but occasionally neighboring vital structures are involved precluding total resection. When this is true, marsupialization or incomplete excision may be done (23, 24, 25, 26, 27, 22). Radiation therapy is not generally accepted as a primary therapeutic measure for these benign and supposedly radioresistant lesions (25, 20, 2, 3, 5, 7). *Kittredge* (20) expressed that "involvement of intestinal lymphatics must be treated supportively since no definite method of treatment is known". *Watson* (7) on the other hand, believed that postoperative irradiation was helpful in preventing recurrence which is not an uncommon event (5, 7). Also, roentgentherapy has been used in cases in which location of the tumor did not permit surgical excision. *Rauch* (4) in a review of retroperitoneal lymphangioma reported two instances of tumors involving the duodenum and the head of the pancreas in whom only aspiration and drainage of the cysts and x-ray therapy have resulted in complete disappearance of evidence of the lesions 4 and 7 years respectively, postoperatively.

Radiation therapy, in our case, was used because of the impossibility of any further resection. The tumor response despite the low dose of irradiation delivered to the upper abdomen and to known areas of tumor spread (kidneys and liver had to be blocked for part of the treatment to avoid radiation damage) was impressive and unexpected. Irradiation was followed by complete tumor regression as evidenced by dissolution of palpable tumor, total absence of symptoms and physical findings, and the return of the patient to a normal and productive life.

The cause of this dramatic result is somewhat speculative. In view of the natural growth cessation and involution characteristic of lymphangiomas, the effect of irradiation therapy is uncertain. However, considering the rapid clinical disappearance of extensive abdominal and pelvic tumor, the possibility of a beneficial effect of irradiation is plausible. Perhaps this treatment triggered and was complementary to an otherwise spontaneous tumor regression.

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BOOK REVIEWS

Steinhausen, D., K. Langer: **Cluster Analysis.** An Introduction to Methods and Applications of Automatic Classification. Walter de Gruyter, Berlin–New York 1977, 206 pages (in German)

Cluster analysis is a valuable mathematical way of classifying automatically a given set of data into smaller units with certain similarities. It is currently mainly used in economic and social studies, however, there is also a large number of publications applying this method to biological data. The authors have written a concise monograph which enables the mathematically advanced biological researcher to solve classifying problems. People with somewhat limited mathematical abilities will find an easy comprehensible representation of methods and examples of cluster analysis of biological data. Thus improving the communication between the biological researcher and the statistician for a cooperative solution of problems.

Peters, Münster

Sapin, M.R., E.I. Borziak: **Anatomie du système lymphatique de l'homme.** Oran University Press, Oran (Algerie), 1976, 286 pages, 69 figures, in French

A considerable body of valuable and new evidence on the structure and arrangement of the lymphatics in various human organs has been amassed in lymphology since the publication of *D.A. Zhdanov's* "General Anatomy and Physiology of Lymphatics" and "Physiology and Pathology of the Lymph Flow" by *Ruznyak, Földi* and *Szabo* in 1952 resp. 1957.

Recent years have seen new findings particularly concerning micro- and ultrastructure of lymph capillaries, vessels and nodes.

The monograph by *Sapin* and *Borziak* is an attempt to generalize all these present-day concepts on the structure of the lymphatic system of the human body. The authors used material of their own research and quoted various other sources published in the Soviet Union and elsewhere.

The first part treats the general anatomy of the lymphatic system. The authors provide a clear-cut idea of the structure of lymph capillaries which are an important link in the microcirculatory pathways of the organs.

The following section deals with texture and age related changes in lymph nodes. Some of the authors observations are here published for the first time.

The part of the monograph devoted to composition, formation, and movement of lymph outlines theoretical considerations of lymphopoiesis and lymph drainage.

The second half of the monograph is mainly concerned with the specific anatomy of the lymphatics,