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# LIPID-STORING SARCOMATOUS LYMPHOCYSTIC FIBROMA IN THE CORPUS UTERI

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The clinical and morphological definition of lymphocystic fibroma, as distinct from other infrequent forms of heterogeneous uterine cystic growths such as uterine cysts, cystomyomas and cystic adenomyomas, originates from Robert Meyer [7] who classified the tumor with the group of *hamartomas*. In his review of 4 own cases and 3 cases reported in the literature he described an up to then unidentified form of tumor. In an earlier report [1], dealing with the surgical removal of a sarcomatous tumor similar to that described by Meyer, we gave a detailed account of its histogenetical and clinical aspects. Neither Meyer's work nor the subsequent reports arose due attention; the recently published comprehensive textbook of Seitz & Amreich fails to take notice of this specific form of tumor except for a brief reference in the chapter entitled: "Genese und Anatomie der Uterusmyome" written by Mittelstrass [9], and other manuals of authority, *e. g.* Novak & Novak's [13] Gynecologic and Obstetric Pathology (1958), or Rubin & Novak's [15] Integrated Gynecology (1956), ignore it entirely. By all appearance, lymphocystic fibroma of the uterus, as a pathologic process of rare occurrence, either escaped clinical observation altogether or has been identified and recorded under a different name. This is all the more regrettable as from the point of view of tumor genesis deep importance apparently attaches to the study of hamartomas, as has been emphasized by Haranghy [3].

The case to be discussed furnishes clear proof that lymphocystic fibroma of the uterus, far from being a mere theoretical term of classification, designates a distinct though rare form of tumor which requires different treatment in dependence on its malignant or benign nature. The unusual diffuse accumulation of lipids in it points to a histogenetic deficiency and furnishes an entirely new basis for interpretation.

K. E. a female patient 51 years of age (No. 720/1954), was admitted October 11, 1954, for more than usually vehement abdominal pains accompanying some months her otherwise still normal menstruation. At admission there were found a vagina of medium width; a conical compact, non-relaxed portio with round orifice, leading to the compact, tumorous uterus of a size

simulating pregnancy in the fifth month. No pathological changes were palpable in the environment. Pregnancy was ruled out by a negative frog test. Myoma was diagnosed and after due preoperative treatment the patient was subjected to laparotomy.

Exposure of the uniformly enlarged uterus with softened posterior wall revealed a tumor which discharged 200 to 300 ml of clear liquid. The shrunken uterus and the adnexa were removed by Chrobak's amputation. The removed organ showed the large cystic cavity in the posterior uterine wall — not communicating with the uterine cavity — and to harbour tuberous growths of small walnut to plum size which bulged into it from every direction and made its inner surface uneven (Fig. 1). These tumorous structures displayed yellowish cut surfaces. For suspicion of malignancy, based on the translucent homogeneous appearance of the cut surface, the cervix too was removed. After an uneventful postoperative period the asymptomatic patient was discharged on the 12th day after surgery.

The excised organ without the adnexa weighed 460 g. The uterus deprived of the cyst's fluid content still outmeasured a newborn's head. The anterior uterine wall was compact, the cavity displayed a polypous area of cornel-stone size in the thick mucous membrane lining it. A large cyst was seen in the posterior wall with protruding tumor nodes from hazelnut to walnut to plum size. The nodes, devoid of distinct capsules, were marked off from the muscular layer of the uterine wall by a faintly coloured zone. The cystic surface of the growths was smooth and glistening, with a delicate network of interstices in the deeper parts. The cut surface was homogeneous, pale yellow in colour, with some darker parts. In the left adnexum there was a small tuboovarian cyst replete with clear fluid; the right tube was of little-finger width, with hydrosalpinx. The right ovary was undersized and atrophied. Endothelium of the portio was smooth, the cervical mucosa pale.

Microscopic examination revealed marked hyperplasia in both the thickened uterine mucous membrane and its polypous portion. The growths bulging into the cyst in various places were excised together with adjacent parts of the uterine musculature. They all agreed in histological structure. Differently thick layers of connective tissue, in some places oedematous and loosely fibrous, separated the muscular from the tumorous parts in which two structural types occurred. In one there were interlacing close bundles of tumor cells which displayed spherical or spindle-shaped nuclei with little cytoplasm and a moderate amount of chromatin and contained one or more large dark-staining nucleoli. An abundant fibrous network was seen to connect the cells and cell group with the intrafascicular blood vessels. Many of these presented wide lumina, some with a thick muscular layer. The cells, generally immature in appearance (Fig. 2), showed certain variations in nuclear size and chromatin content. This was especially marked wherever the fascicular structure gave way to extensive

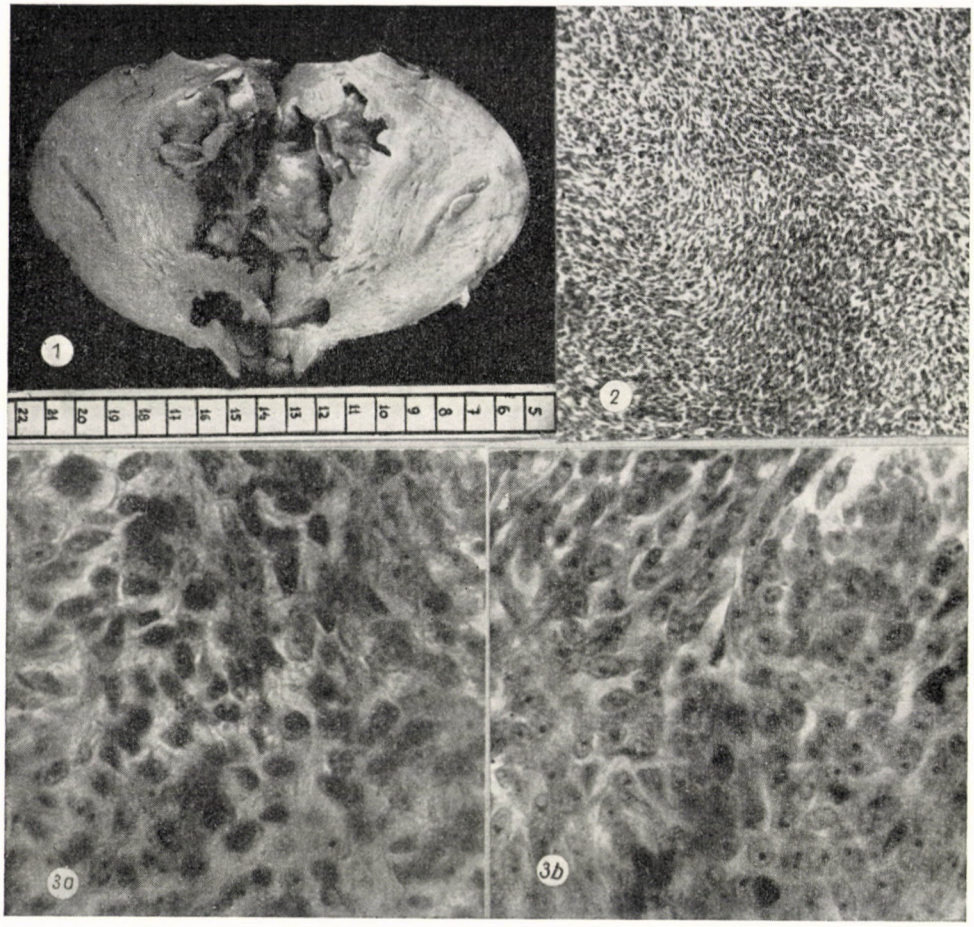


*Fig. 1.* Sagittal section of uterus with tumorous growths bulging from every direction into the cystic cavity of the posterior wall. The uterine cavity appears in the form of oblong gaps on the left and right sides

*Fig. 2.* Portion of tumor with the fasciculate structure in some places clearly distinguishable, in others about to disappear. Atypical nuclei (Haematoxylin-eosin, ×225)

*Fig. 3a—b.* Area rich in cells with frequent nuclear divisions (Haematoxylin-eosin, ×540)

cell aggregation, reminiscent of vast germinal centres and presenting frequent cellular divisions with nuclei varying in size and in chromatin staining (Figs. 3a, 3b). In several places multipolar divisions gave rise to groups of undersized cells. A number of binucleated cells produced very little of the delicate fibrous substance. Fibrous structure enclosing the capillaries formed the framework the tumor, with a very delicate reticulum interconnecting the cell groups (Figs. 4a, 4b). In certain areas the basic structure appeared explicitly reticular (Fig. 5). Frozen sections obtained from every part of the tumor abounded in

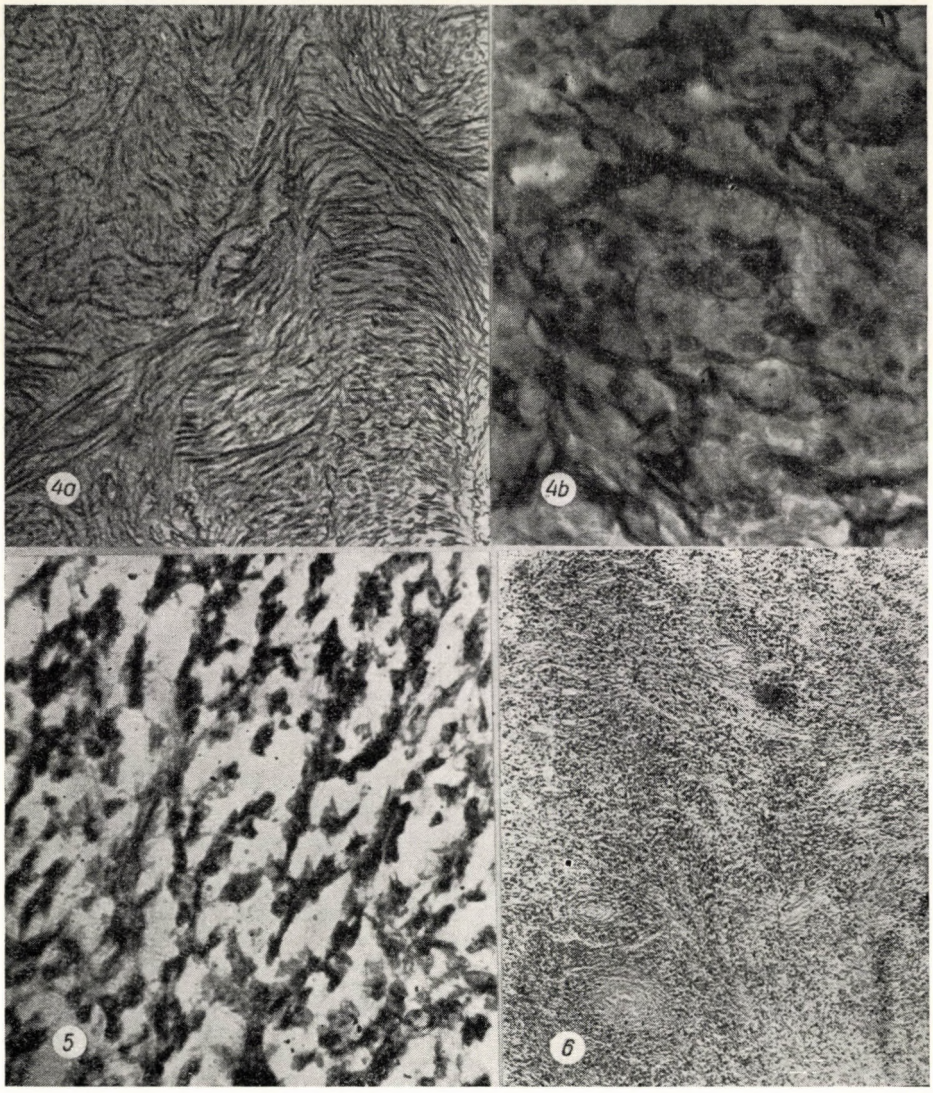


*Fig. 4a.* Argyrophilic and collagenous tumor fibres (Bielschowsky's silver impregnation, ×225)
*Fig. 4b.* Non-fasciculate tumor portion characterized by cell abundance and fibre scarcity (Bielschowsky's silver impregnation, ×540)

*Fig. 5.* Reticular tumor structure (van Gieson's, ×540)

*Fig. 6.* Frozen section of tumor area abounding in lipids (Sudan III, ×120)

lipids staining readily with Sudan III. The fatty substance in the cytoplasm appeared either as fine granules or as larger clods (Fig. 6). Some of these sections were stained with haematoxylin-eosin, some according to van Gieson and some were impregnated with silver. It was found that in the areas rich in

fibres the cells contained much more fat than those situated in areas poor in fibres and characteristic of sarcomatous degeneration. The latter areas contained very few and delicately fine fat granules.

Histological examination revealed accordingly multicentric sarcomatous degeneration associated with lipid deposition in the intracystic fibroma of the uterine wall, with considerably more lipid in the more mature and quiet areas than in the degenerated parts characterized by cell abundance and fibre scarcity.

The tumor abounded in wide blood vessels with independent muscular layer. The intracystic superficial tumor portions were covered with a distinct layer of endothelium. Smaller widened lymphspaces were occasionally seen in the tumorous growth itself and in the loose connective tissue between the tumor and the uterine musculature. The scanty cells in this loose connective tissue also showed evidence of incipient malignancy.

The ovaries showed many fibrous bodies deep underneath a thin cortical zone. There was no histological sign suggestive of thecal hyperactivity.

REUTER [14] regarded lymphocystic fibroma as sarcoma; WOLFF [16] described one case as explicitly fibrosarcomatous in structure. R. MEYER [7], though admitting the possibility of a sarcomatous degeneration, recorded neither metastases nor a fatal outcome. In his view the tissue-dissolving action of the tumor parenchyma was not more intensive than that seen in adenomyosis. NEUMANN [12], NETZBAND [11], LIMBURG [6], KAUFMANN [5] and MUSCHIK [10] all failed to encounter malignancy. Because of the sarcomatous degeneration in our earlier case, that patient was subjected to X-ray irradiation; now, seven years later, the patient is still symptomless.

As to the case under review, fear of malignancy based on examples in the literature and on the finding of tumorous growths invading the cystic cavity but sparing the uterine musculature, induced us to remove the tumor radically and to abstain from post-operative X-ray treatment. There were no further complaints after a five-year term of close observation.

Lipid deposits, a change that has not been mentioned in the literature on lymphocystic fibroma, were observed in every part of the tumor. There was more lipid in the areas with fasciculate structure than in those marked by immaturity and cell abundance. The loose connective tissue between the tumor and the uterine musculature abounded in lipids whereas beyond that zone no lipid was found in the uterine wall.

As to the question, whether accumulation of fat inside the tumor cells was due to some peculiar cellular activity or to lipid precipitation in consequence of a protoplasmic lesion, BÜCHNER [2] in agreement with HARANGHY [3] suggested the intracellular accumulation of lipids to be due to "a change in the molecular network of the cytoplasm", brought about either by defective cellular function or by assimilation of an excessive amount of fat. HARANGHY

suggests three alternatives to explain the intercellular accumulation of fatty substance in loose bondage: (I) fat production in excess to the amount readily used up by the cells under normal physiological conditions; (II) reduction of fat metabolism due to pathological cellular functioning; (III) circulatory or histogenetical disorders unfavourably influencing fat metabolism.

In accordance with MEYER's view, it seems legitimate to claim that the described tumor qualifies as one form of *hamartoblastomas* due to primary disorders in the lymphatic circulation accompanied by cyst formation, with ensuing impairment of blood circulation and tissue oxidation. The significance of lipid accumulation will have to be clarified by future investigations.

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## НАКОПЛЕНИЕ ЛИПОИДОВ В САРКОМАТОЗНОЙ ЛИМФОЦИСТОФИБРОМЕ ТЕЛА МАТКИ

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Найденная в теле матки радикально оперированной 51 летней женщины опухоль величиной в голову грудного ребенка показала морфологические характеристики лимфоцистофибраны матки, но ее диффузный желтый цвет отклонялся от описанных до сих пор в литературе случаев. Под микроскопом наблюдалось многоцентрическое саркоматозное перерождение, и во всех частях опухоли хорошо окрашиваемое Суданом III обильное накопление липоидов. В полном согласии с мнением *Р. Мейера* опухоль можно рассматривать гамартобластомой, а накопление жира как возвращение эмбрионального свойства клетки. — Больная не получила послеоперационного лучевого лечения, так как бесспорно было выявлено, что опухоль не показала инфильтративного роста в направлении мышц матки. По истечении 5 лет больная была свободной от жалоб и в здоровом состоянии.

## LIPOIDANHÄUFENDES LYMPHOCYSTOFIBROMA SARCOMATOSUM CORPORIS UTERI

K. KOVÁTS

Die im Gebärmutterkörper einer radikal operierten 51 Jahre alten Patientin gefundene Geschwulst erschien makroskopisch als Lymphcystofibrom, doch wich sie mit ihrer diffus ockergelben Farbe von den bisher in der Literatur beschriebenen Fällen ab. Mikroskopisch

konnte eine multizentrische sarkomatöse Degeneration und reichliche diffuse Lipoidanhäufung beobachtet werden. Im Einklang mit R. MEYERs Auffassung kann der Tumor als *Hamartoblastom* angesehen und die Fettanhäufung mit der Wiederkehr der embryonalen Zelleigenschaft erklärt werden. Die Patientin erhielt keine postoperative Strahlenbehandlung, da ein infiltratives Wachstum des Tumors in die Gebärmuttermuskulatur ausgeschlossen werden konnte. Nach 5 Jahren war die Kranke noch immer beschwerdefrei.

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