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Endolymphatic Stromal Myosis

Report of a Case Treated Surgically and with Hormones

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Summary. Stromal endolymphatic myosis is regarded as a mixed homologous tumour with a prominent proliferation of the stroma and a varying degree of proliferation of gland-like structures and vessels.

A case is presented in which the patient was treated periodically with progestine. This therapy seems to have been of benefit in controlling the frequency of recurrence.

Endolymphatic stromal myosis is often used in Anglo-American literature for a rare, tumorlike proliferation of the uterine endometrium [15]. Origin from endometrial elements outside the uterus has been described in a very few cases [9, 13, 16]. Microscopically the tumour is characterized by infiltrative growth in the uterine wall of strands or masses of an endometrical stroma-like tissue which shows a tendency to proliferate around or within vascular channels, some of which are large [15].

Although the growth may represent a sarcoma of the endometrium with a low grade of malignancy, most authors seem to regard it as a distinct and specific entity with a characteristic microscopical appearance, a "benign" cytology and relatively benign clinical behaviour [4, 7, 18, 23]. Others, however, consider it a simple variant of endometrial sarcoma [11].

Different views on the histogenesis have also led to the use of many synonyms. Doran and Lockeyer who described the disease for the first time in the English literature in 1908 called it a perithelioma [6]. Similar uterine tumours seem to have been described by Hansen in 1903 and Bechaus in 1907 as hemangio-endotheliomas [3, 12]. A vascular origin has also been proposed by many later authors [10, 24, 25, 27].

Casler who reported the first case in the American literature, described it as an adenomyoma without glands [5].

Some have regarded the disease as a variant of either adenomyosis or endometriosis with a predominance of stromal elements, and with minimal or absent glandular components [8, 14, 30].

It has recently been suggested that the disease may be hormone dependent [2, 19, 21, 26].

The case presented here has been followed for more than 7 years and demonstrates pathological, clinical and therapeutic aspects of the disease.

Case Report

A woman aged 35 yrs, was admitted to hospital in June 1967. She had 4 children. The last was born in 1965. She had abnormal vaginal bleeding for 2 months prior to admission. This was treated with Lynestrol-ethinyloestradiol ((Lyndiol)®, but she had become worse.

The uterus was enlarged, firm and rough. The clinical diagnosis was myomata. A rich curetage was histologically classified as endometrial polyps. Two months later supravaginal hysterectomy and removal of a slight enlarged cystic ovary was carried out because of further bleeding. The histological diagnosis was now endolymphatic stromal myosis.

In June and December 1968 she had a recurrence in the form of vaginal polyps. These were removed surgically. In 1969 2 polyps were detected in the vagina and removed together with residual tumour in the pelvis and her remaining ovary. The patient was then treated with progesterone for 6 months. Depo-Provera® (Medroxiprogesteroni acetas) was given in doses of 750 mg weekly for 2 months, later 250 mg weekly for 3 months, thereafter 500 mg monthly. She then stopped medication due to increasing weight and discomfort. In January 1972 there were 2 polyps on the anterior vaginal wall. These were removed, but not radically. Since that time she has again been treated with high doses of progesterone and there have been no further signs of recurrence.

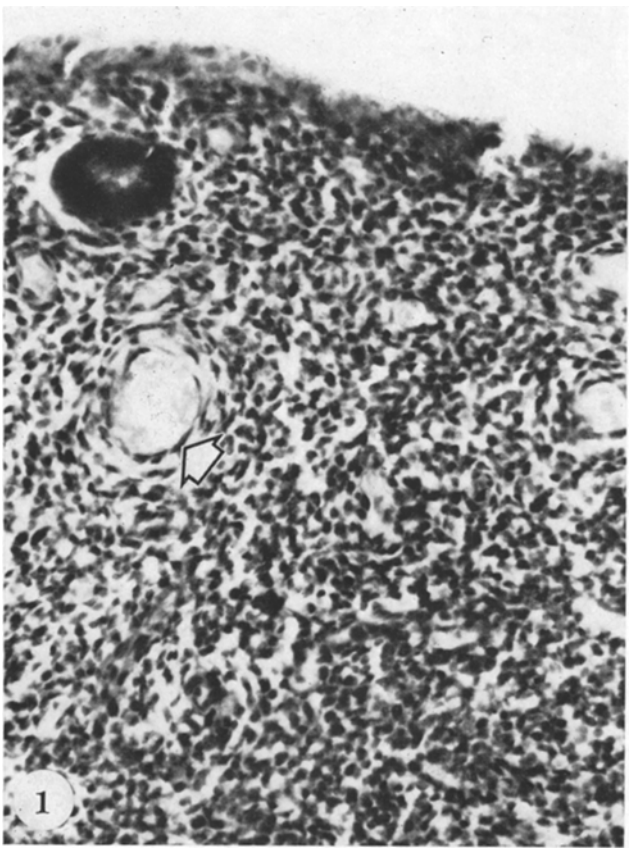

Fig. 1. (P. 5322/67) Endometrial polyp with a single small gland and distinctive arteries (arrow) in a proliferative stroma (HE × 110)

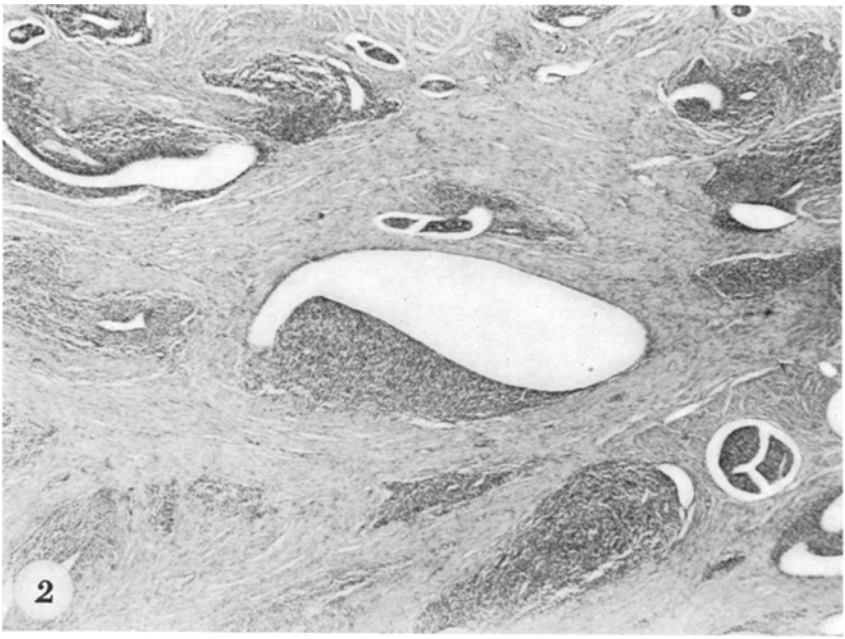

Fig. 2. (P. 7880/67) Histological specimen showing the characteristic growth pattern of endolymphatic stromal myosis. (HE × 30)

Pathological Examination

The material investigated consisted of: Endometrial curettings (P 5322/67), the amputated uterus (P 7880/67), the recidual tumour in the pelvis and different metastases to the vagina (P 5080/68, 10403/69, 2107/69, 6351/69, 826/72 and 1462/72). The histological specimens were all stained with haematoxylin-eosin and many of them with Azan-Heidenhain, Weigert's Elastin-van Gieson, Wilder's Reticulum Stain, Mallory's P. T. A. H., PAS and Alcian Blue.

The endometrial material consisted of "polyps" microscopically characterized by a proliferative endometrial stroma, very few rounded glands, and comparatively many vessels resembling arteries (Fig. 1).

The amputated uterus measured $10 \times 10 \times 6$ cm and weighed 550 grams. The uterine cavity was eccentric in position and with a somewhat thickened and polypous mucous membrane.

The uterine wall was diffusely thickened with a whorled appearance, grayish in colour with light yellow areas. Some distinct bleeding points were seen. The consistency alternated between firm and soft areas.

Microscopically the uterine wall was infiltrated in a rather characteristic manner by tumour tissue quite like proliferative endometrial stroma. The tissue seemed to split up the fibers of the uterine wall, without being continuous with them. There was a tendency to protrude into vessels, some of which were probably lymphatic and others blood vessels. Many of the vessels contained tumour tissue free in their lumina (Fig. 2).

Continuity between the endometrium and the tumour tissue was demonstrated (Fig. 3). The tissue was mainly built up of rounded to slightly polygonal cells with bright nuclei and a small amount of cytoplasm. The cells varied only a little in size and mitoses were few. Collagen fibers were not seen within the tumour tissue. Groups of cells with more hyperchromatic nuclei were present, especially in tumour tissue lying within vessels (Fig. 3).

Both in the main tumour and in the metastases characteristic small areas were seen that were built up of cells lying between and around small vessels. These vessels were not unlike arteries seen in the endometrium (Fig. 1).

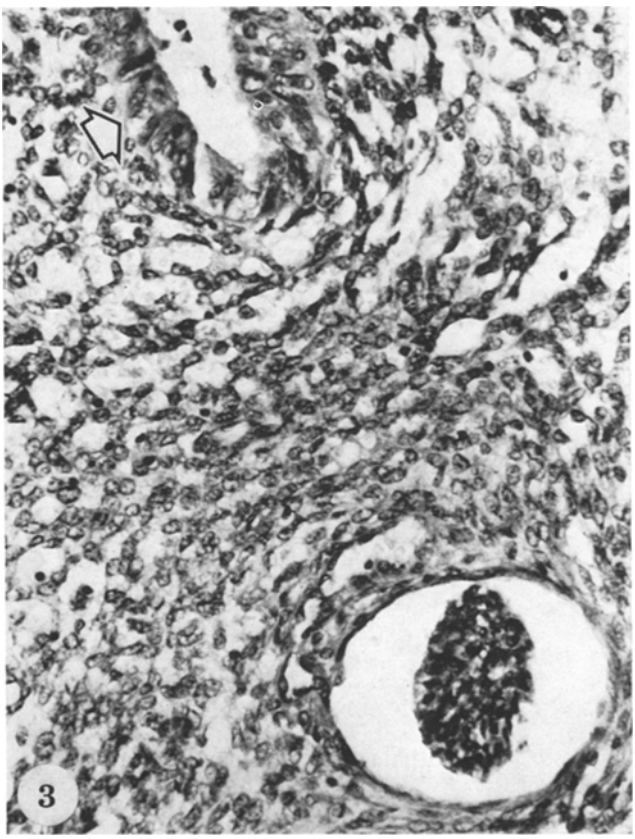

Fig. 3. (P. 7880/67) Tumor tissue, with covering endometrium (arrow), partly lying within a vessel. (HE×280)

In one of numerous specimens from the depth of the uterine wall, garland-like epithelial structures were found, probably abortive glands (Fig. 4).

The vaginal and the pelvic metastases varied in size, from that of a pea to a small tomato and the microscopical picture was similar to that of the main tumour.

Discussion

The behaviour of the tumour in our case is that of a tumour with low degree of malignancy, although within 2 yrs of the first operation 4 new tumours were detected in the vagina and 1 in the pelvic cavity. Surgery and x-ray therapy were previously used in treatment; but hormone therapy seems valuable in this disease as well as in cases of carcinoma of the endometrium [20]. Our case was treated surgically and was not given x-ray treatment.

Postoperatively progestine was given at high doses, but the patient stopped the medication herself after 6 months. Within less than 2 years after that she developed 2 new tumours in the vagina. Since then progestine has been used for more than $3^1/_2$ years without sign of further spread. As the metastases occurred

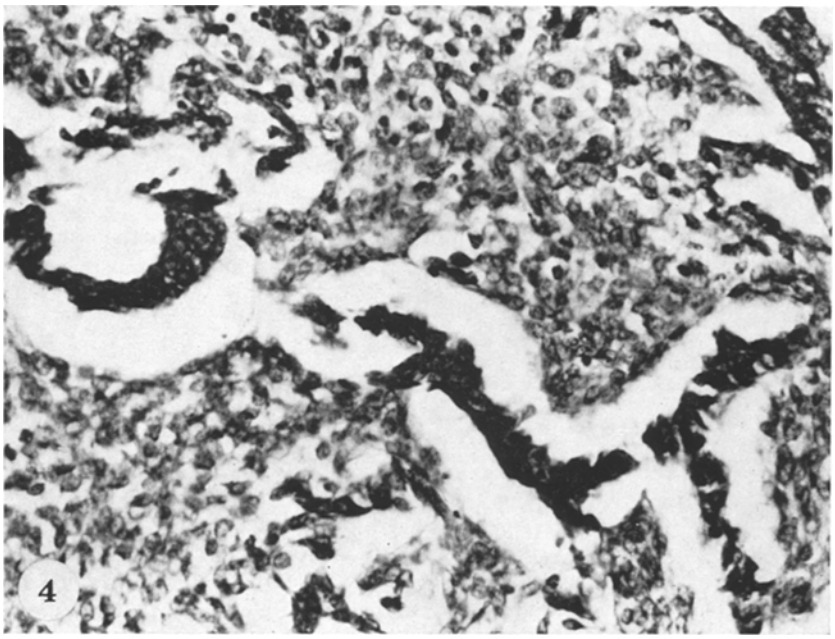

Fig. 4. (P. 7880/67) From the depth of the uterine wall. Epithelial-like structures within the tumor. (HE×280)

in the period without medication, the treatment may have been of benefit, in accordance with the effect described by others [2, 19, 26]. It must not be forgotten, however, that such tumours can be silent for years without treatment, up to 23 years having been reported [7, 18]. Baggish described gross regression of endolymphatic stromal myosis after bilateral oophorectomy probably due to reduced production of oestrogens [2]. Toker described development of malignant stromal elements in a patient with a hormone producing the coma in the ovary [34]. Our case gave the impression that the condition became worse prior to surgery, while she was given Lyndiol.

The tumour is thought to be of endometrial origin. Hormone therapy with progestine has been shown to induce decidual changes in the tumour tissue [2], and electron microscopic studies indicate compatibility with an endometrial stromal histogenesis and ultrastructure different from hemangiopericytoma [23, 29]. It can not be denied, however, that small parts of the present tumour closely resembled a hemangiopericytoma [1, 31, 32]. There is a similarity between the vessels of the tumour and the arteries of the uterine mucosa (Fig. 1), and these are interpretated as spiral arteries by some authors [17]. In one of numerous sections in the present material glandlike structures were seen deep in the uterine wall. Only a few authors have described such findings, in up to 25% of their cases [23]. This variation may be connected with the number of specimens investigated.

It is likely that we are dealing with a tumour of the endometrium with predominantly stromal proliferation and varying degree of proliferation of the glands and the arteries. The view that this tumour bears a close relationship to mixed mesodermal tumours of the endometrium, is interesting [33]. However, the prognosis in endolymphytic stromal myosis is generally much better, with up to 100% 5 year survival [23]. It is therefore suggested that endolymphatic stromal myosis should be regarded as a specific entity.

The histological diagnosis of endometrial sarcomas in curtage material is difficult [22]. In endolymphatic stromal myosis the primary diagnosis in curetage material seems to be a rarity, and the clinical behavior is usually of little help [18, 28].

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