ENDOLYMPHATIC STROMAL MYOSIS

(A Case Report)

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Endolymphatic stromal myosis which has got several synonyms like—Endometrioma, Fibromyosis, Haemangiopericytoma of the uterus, Perithelioma, Stromal endometriosis, Stromatoid new growths of the uterine wall, and stromatous endometriosis is characterised by the presence of strands and masses of infiltrating non-collagenous connective tissue within the myometrium. These infiltrating masses are usually regarded as being of endometrial in origin and associated with endothelial lined channels presumably lymphatics.

This is a very rare disease which as the name indicates stands between uterine adenomyosis and endometrial stromal sarcoma. It is progressive and infiltrates the supporting tissues of the pelvis by way of vascular spaces. Till 1949 when Park reviewed the subject he found that only 50 cases were reported. After that Hunter in 1953 reported 10 cases apart from few others reported in the literature.

The case is reported due to rarity of the disease itself.

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CASE REPORT

Mrs. K. R., aged 42 years, married, para 3, gravida 3, was admitted on 17-10-1975 with complaints of irregular and excessive menstrual flow for the last 2 years and gradual enlargement of abdomen for the last one year.

Patient had normal menses till 2 years back (4-5/28-30 days, moderate flow). In the last 2 years the flow as well as the frequency increased (5-15/15-20 days, excessive flow).

Past history, family history and personal history were not of importance. Birth of last child was 15 years ago.

The patient was fairly built and nourished, her B.P. was 136/80 m.m. Hg. and systemic examination did not reveal any abnormality. On abdominal examination a midline, well defined suprapubic firm mass of 14-16 weeks size was palpated which was non-tender. Speculum examination did not show any abnormality. On bimanual examination uterus was uniformly enlarged to 14-16 weeks of size, firm in consistency and fornices were clear.

Her haemoglobin was 10.5 gm%, fasting blood sugar was normal and routine urine examination did not reveal any abnormality.

She was diagnosed clinically as a case of fibromyoma uterus and total abdominal hysterectomy with right sided salpingo-oophorectomy was done. Postoperative period was uneventful and patient was discharged on the 10th postoperative day in good condition.

Histology-Gross: Specimen received in formal saline consists of uterus with cervix and right sided adenexa, measuring 10 x 8 x 8 c.ms, greyish pink, soft to firm in consistency. Cut

surface shows a well circumscribed tumour mass measuring 5 c.ms. in maximum diameter and presenting a variegated appearance with area of haemorrhage and necrosis. The viable tumour tissue presented a lobulated appearance. The ovary measured 2 x 1 x 1 c.ms., greyish white firm without any specific feature on external or cut surfaces. The tube was patent.

Microscopic: Microscopically non-collagenous connective tissue resembling endometrial stroma is seen extending in between the myometrium. The individual cells were slightly oval to rounded in appearance with large nuclei and rather scanty cytoplasm. The tumour mass showed moderate degree of vascularity. No definite vascular or lymphatic permeation was observed.

Discussion

Most of the cases of stromal myosis are found in patients between 31 and 50 years but the age range is from third through the seventh decades. In our case the age was 42 years.

There is no characteristic clinical picture. Menorrhagia, metrorrhagia, "Vaginal bleeding" and post menopausal bleeding are said by Park (1949) to be the common symptoms. The commonest physical sign is enlargement of the uterus.

There is no agreement as to the anatomic origin of the component tissue in the endolymphatic stromal myosis. Three main theories have been advanced.

1. Direct extension or invasion from the basal endometrium.

2. Unifocal or multifocal development from undifferentiated tissue already present within the myometrium.

3. Embolism by way of lymphatics or blood vessels or both from the endometrial stroma.

Recently origin from vasoformative cells has been suggested (Pedowitz *et al* 1954) and the tumour named "hemangiopericytoma". It is interesting that late recurrences have a vascular pattern similar to the original tumour.

The treatment is difficult to evaluate as there are so few cases and such a long time may elapse before recurrence. As long as the uterus is removed it does not seem to be important whether the tubes and ovaries are removed unless there is extension into the muscular wall of the tube. It seems indicated that any pelvic extension should be removed, although some authors such as Handerson (1946) consider that the tumour left behind will die since its blood supply presumably has been destroyed. However, if it is truly a vascular tumour as suggested by Pedowitz *et al* (1954) this would not be logical reasoning. Whether or not roentgenotherapy should be given is also open to question. Handerson (1946) considers this condition well differentiated and not radio-sensitive.

Summary

A case of endolymphatic stromal myosis has been reported and its clinical features, pathogenesis, treatment and prognosis has been discussed.

References

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See Fig. on Art Paper I