A case of Stein-Leventhal Syndrome complicated by large multiple fibromyomas

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Introduction

ALTHOUGH STEIN-LEVENTHAL syndrome[15] has been extensively documented since 1935, it often remains a diagnostic problem. That such confusion exists is made clear by a study of the literature on the subject.[6, 9]

In recent years, there have been reports of cases of Stein-Leventhal syndrome complicated by such pelvic lesions as ovarian neoplasms,[2, 7] endometrial carcinoma [1, 3, 8] and endometrial hyperplasia.[11, 14] So far, the author has been unable to find in the literature any report of large fibromyomas complicating Stein-Leventhal syndrome. The purpose of this case report is to draw attention to just such a case erroneously diagnosed as ovarian tumour.

Case Report

A 26-year-old nulliparous, unmarried Chinese woman was first seen on 26.7.69 because of secondary amenorrhoea of two years' duration. She also complained of swelling of the lower abdomen for three months, and swelling of both legs for six days.

Menarche had occurred at 14 years of age but menstruation was always irregular, occurring at two- to six-month intervals, with a three-day moderate flow. From the age of 22 years, she began consulting several doctors about this, and many attempts had been made to regulate her periods with oral progestogens with no success. There was no history of obesity or voice changes. She was the seventh child in a family of nine siblings. Her two sisters had normal menstrual histories.

Physical examination revealed a healthy looking young woman with no acne or hirsutism. The breasts were well developed. Abdominal examination showed a smooth, tense firm swelling arising out of the pelvis to the level of the umbilicus. No shifting dullness was detected. On rectal examination, the lower pole of the tumour could be felt in the Pouch of Douglas. The uterus could not be identified. No other masses were felt. The external genitalia were normal.

Gravindex test was negative. Nuclear sexing showed a female chromatin pattern. Straight X-ray of the abdomen showed a large soft tissue pelvic mass rising to the level of the 4th lumbar vertebra. A provisional diagnosis of ovarian tumour was made.

On 30.7.69, a laparotomy was carried out. The uterus was found to be enlarged by a fundal fibromyoma 6 cm in diameter, a posterior intramural

fibromyoma 12 cm in diameter and 4 anterior subserous fibromyomas. Both ovaries were cystically enlarged, each to 6 cm in diameter and covered by thick glistening opaque yellow capsules. The Fallopian tubes and the pelvic peritoneum were normal. Bilateral wedge resection of the polycystic ovaries and myomectomy were performed.

The postoperative period was complicated by the development of persistent pyrexia which resisted antibiotic therapy and which only subsided following the drainage of an unexpected pyometra on 21.8.69.

Twenty-eight days after she left hospital, menstruation occurred spontaneously; this lasted five days. Since that time, she has had two more normal periods, at monthly intervals.

Pathology Report

(1) Posterior fibromyoma. Gross: Specimen consisted of an encapsulated circular mass of tissue 12 cm in diameter. It weighed 350 grams. Cut section showed circular whorls.

Fig. 1: Photomicrograph of a section of polycystic ovary showing numerous cysts.

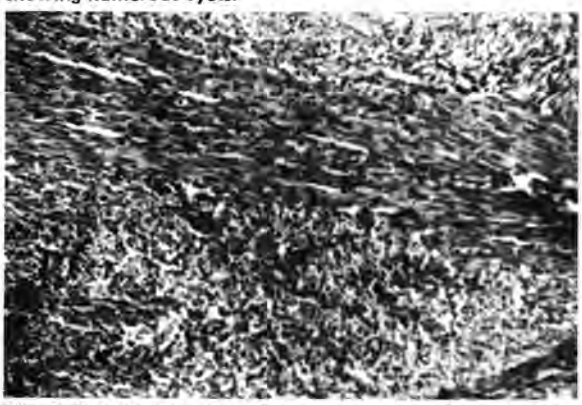

Fig. 2: Photomicrograph of fibromyoma showing hypercellularity.

Microscopic: Sections showed a tumour composed of dense aggregates of cells of varying shapes, from spindle to round. Some areas of hypercellularity were noted.

(2) Ovaries. Gross: Each section showed several cysts up to 1 cm in diameter.

Microscopic: Moderately thickened and fibrotic tunica albuginea; below this were several small cysts lined either by granulosa or theca cells. No corpora lutea seen.

Comment

The crucial point causing diagnostic difficulty in the young woman is the history of amenorrhoea. The occurence of amenorrhoea in Stein-Leventhal Syndrome is to be expected. But a woman with fibromyomas seldom has amenorrhoea, even of short duration, unless she is pregnant or past the menopause. Retrospectively, in this case amenorrhoea may be explained by the abnormal hormonal influences arising from a disturbed hypothalamic-pituitary-ovarian relationship postulated for Stein-Leventhal syndrome.[6, 9, 13] Whether the adrenal cortex is overactive in this syndrome remains a mystery.[12]

It is not known how often Stein-Leventhal syndrome and fibromyomas co-exist. This may be due to the following factors:-

(i) The difficulty in diagnosing Stein-Leventhal syndrome per se. Although this condition is characterised by menstrual irregularities (amenorrhoea and oligomenorrhoea), infertility, obesity, hirsutism and polycystic ovaries, most investigators agree that none of them is present consistently. There are many cases in which obesity and/or hirsutism may be totally absent.[5, 9] In the single woman, even infertility is only potential. Indeed, some authors would dispute the existence of Stein-Leventhal syndrome as a clinical entity.[9, 11] It has been stated that the sine qua non in the diagnosis of Stein-Leventhal syndrome is the presence of palpably enlarged polycystic ovaries.[10] Adopting such an arbitrary standard has occasionally resulted in the lax acceptance of cases with mere enlarged ovaries as Stein-Leventhal syndrome, unsupported by any consistent ovarian histopathologic picture or biochemical findings.[4]

(ii) A survey of the literature found little description of the state of the uterus in Stein-Leventhal syndrome. The few who did so, remarked that three-quarters of such uteri were hypoplastic; the rest were normal in size.[10, 15]

None reported a case of Stein-Leventhal syndrome complicated by large fibromyomas. Considering the relative frequency of fibromyomas, this discrepancy is all the more surprising. Perhaps it may partly be explained by the fact that the majority of small fibromyomas, and some large ones, are symptomless. Obesity may interfere with an accurate estimation of uterine size. It is also suggested that the silent presence of fibromyomas in patients with Stein-Leventhal syndrome could conceivably be overlooked, even by operators performing wedge resection on the polycystic ovaries.

Summary

1. A patient with the Stein-Leventhal syndrome who was found to have large multiple fibromyomas has been described. The fibromyomas were discovered at laparotomy.
2. Fibromyomas complicating Stein-Leventhal syndrome should be included in one's differential diagnosis, whenever a young woman with an unaccountably long history of secondary amenorrhoea, is found to have a large pelvic tumour.
3. It is recommended that more attention be paid to the state of uteri in cases of Stein-Leventhal syndrome.

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