ARRHENOBLASTOMA OF OVARY

by

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Arrhenoblastoma is a rare ovarian tumour, and majority of these are defeminising and virilising neoplasms. Their naked eye appearances are identical with those of feminising tumours. The somatic changes that are produced by the biologically active tumour are due to the hormonal activity of the neoplasm. The histologic characteristics of tubular structures that mimic the sex cords of the testes were definitely described by Meyer. More than 240 cases have been reported in the literature in the past 60 years. Thirty-one more arrhenoblastomas have been reviewed by Thomas N. O'Hern and Neubecker, Robert D.

The present case is reported because of comparative rarity and because of its quick response to the treatment, that is reversion to feminisation and ensuing pregnancy within a few months of removal of the tumour.

Case Report

J. aged 30 years, was admitted to M. R. Rangur Hospital, Tollygunge, on the 6th June 1963 with the following complaints: Amenorrhoea for 5 years & 3 months, swelling and pain in abdomen for 5 years, atrophy of the breasts for 1 year, excessive growth of hair, with beard and moustache for 3-4 months and change of voice for 3-4 months.

Past menstrual history—Menarche at 13 years, cycles regular, every 28 days, menstruation lasting for 3-4 days, flow moderate with slight dysmenorrhoea.

Present illness—Patient gave birth normally to a male baby at term 6 years ago. Six months later she had three consecutive periods lasting for 3 days and the flow was normal. Since then she had amenorrhoea. Two months after the stoppage of menstruation she noticed a small swelling in the right iliac region which gradually increased and attained the present size in 5 years. She nursed her child for 5 years. For the last one year i.e. after the stoppage of breast feeding, she noticed atrophy of both breasts. There was also change of voice and growth of excessive hair on the body, including a beard and moustache. The patient attended our O.P.D. on 4-6-63 and was admitted on 6-6-63.

On examination, build and nutrition of an average masculine, no anaemia and oedema. Hair and moustache present. Excessive growth of hair on limbs axillae beard and lower part of the abdomen. Hair on pubis, change of voice present. Atrophy of both breasts but the nipples and areolae were normal. On squeezing, milk came out.

Heart and lungs—normal. Liver and spleen—not palpable.

There was a mass in the lower abdomen in the right iliac region, size about 4″ x 3½″. Feel was firm and at places cystic, mobile and not tender; no free fluid in the peritoneal cavity was detected.

Per vaginam, external genitalia, enlargement of clitoris; labia majora and minora were normal on both sides. Uterus was found separate from the mass, mobile and normal in size. The firm cystic mass felt on the right side corresponded with the

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abdominal swelling. Vaginal walls were normal. Rectal examination revealed the same findings.

Provisional diagnosis of virilising ovarian tumour of right side was made.

Investigations: Hb—65%, B.P.—110/70; urine and stool—nil abnormal. Blood sugar—70 mgm. per 100 c.c. plasma, cholesterol—168'3 mgm per cent, urea—25 mgm per cent. N.P.N.—28 mgm per cent. 17-Ketosteroid 10'9 mgm. in 24 hours of urine. Ketogenic hormone—23'9 mgm. in 24 hours of urine. X-ray of pituitary fossa showed thickening of the posterior clinoid process with small pituitary fossa.

Vaginal smear—major portion of the cells was precornified. There were a good number of cornified cells as well. An approximate count of maturity index 2-60-38. The smear was interpreted as having good hormone effect, particularly progesterone. Endometrial curettage was done but no endometrium was obtained. Plain x-ray of the chest revealed no metastases in the chest. Peritoneal insufflation could not be done due to lack of facilities. Pyelography was done—no abnormality detected.

On 20-6-63 a laparotomy was done. On opening the abdomen the tumour was found on the right side, the uterus and the left appendages were found normal in size and position. There was no peritoneal fluid or no adhesions in the abdomen. The tumour was well capsulated and not potentially malignant, and as the patient was young with only one child, right-sided ovariotomy was done.

Histopathological report: The tumour was globular and 12 cm in diameter, the surface was smooth, and heterogenous in consistency. Cut section showed multiple locular cysts containing yellowish, thin glairy mucoid material. The solid areas showed multiple coloured areas, and were suggestive of solid mucoid material. This was a classical case of arrhenoblastoma of mixed type. (intermediate type). Fig. I, II, III.

Post-operative period was uneventful. 30-6-63—Ten days after operation the muscles of her body were relaxed and the patient said that she felt her body had become softer.

30-7-63—Voice was slightly changed. Menstruation started on 20-7-63, that is just one month after removal of the tumour, the flow was normal lasting for 3 days. Hair over the face was much less and the clitoris was smaller than before. 11-9-63, voice changed but still hoarse, beard and moustashe had fallen. Body was more rounded. The clitoris was still slightly enlarged.

8-10-63—Voice still hoarse. Clitoris was almost normal in size.

7-4-64—She was 2½ months pregnant. Voice was feminine now. She attended pre-natal clinic regularly, had a normal course and on 26-10-64 delivered a female child weighing 5 lbs. 4 ozs. Puerperium was normal in all respects.

10-9-66—She was examined and there was no recurrence of any other abnormality.

Comments

Review of literature was done on the basis of 240 cases. This is a defeminising and virilising ovarian neoplasm. The tumour was unilateral in 96% of cases, usually 6-15 cm. in diameter but may attain much bigger size than that. The over-all malignancy was 22 per cent in 119 cases of Meyer, Javert and Finn. It occurs most frequently in the 2nd to 4th decade of life. Hirsutism is the most common virilising symptom (in 79% of cases). A male voice and enlarged clitoris developed in 50% of cases. The symptoms including amenorrhoea, oligomenorrhoea, breast atrophy, loss of subcutaneous fat occurred in 70% of cases. All the symptoms mentioned above were present in this case. Meyer was the first to divide arrhenoblastomas into 3 histologic types.

Type I—a well differentiated form also known as adenoma tubularae, composed of uniform tubules the

cells of which have the appearance of mature sertoli cells.

Type II—as an intermediate form presenting tubules like testicular tubules in all stages of gonado-genesis.

Type III—an atypical sarcomatoid type. The present case was of intermediate type.

Endocrinologic aspect

Virilising changes were associated with 50 per cent of the well differentiated tumours (Type I), in about 70% of the intermediate type (Type II) and all those of the sarcomatoid type (Type III). In 31 patients of Thomas N. O'Hern *et al,* 4 patients had no evidence of endocrinological effects, 3 showed no virilisation but rather, evidence of hyperoestrogenism. Previously reported arrhenoblastomas produced evidence of hyperoestrogenism and have been reviewed in detail and tabulated by Telium. Plate has presented evidence suggesting that interstitial cells may produce both androgenic and oestrogenic hormone. In most patients with arrhenoblastomas reported in the literature urinary 17-ketosteroids level was normal. In the present case the urinary 17-ketosteroid was not elevated. Sometimes 17-ketosteroid level in the urine is slightly elevated in case of arrhenoblastoma. Despite the marked virilisation 17-ketosteroids level was increased preoperatively in only 13 of 35 cases. Among the 51 malignant cases, metastases were noted at operation in 23. In 28 cases malignancy was evidenced by recurrence of the neoplasm usually within 2 years after the initial operation. The true incidence of malignancy was 25 to 30%. Recurrence may occur 10-12 years after extirpation of the tumour.

Relationship of the tumour with pregnancy

The simultaneous occurrence of arrhenoblastoma and pregnancy is exceedingly rare, only five such cases having been reported. Pregnancy prior to the appearance of tumour has been recorded at least a hundred times in 38 patients. Pregnancy after surgical removal is known to have occurred at least thirty times in twenty-five cases. Presumably the simultaneous existance of arrhenoblastoma and amenorrhoea accounts for some of the relative infertility. Removal of the tumour is often followed by a return of menstruation and subsequent pregnancy as happened in this case. The opportunities for pregnancy prior to the development of arrhenoblestomas are virtually the same as for normal women if there are no associated congenital malformations and amenorrhoea. Pregnancy after conservative treatment of arrhenoblastoma is apparently not different from that in any woman who had an unilateral salpingo-oophorectomy. Conservative extirpation is desirable for apparently benign unilateral tumours in young women with normal genital organs.

Summary

A case of unilateral arrhenoblastoma of the right ovary in a woman aged 30 is described where all virilising and defeminising symptoms were present. 17-ketosteroid was within normal limit; conservative treatment

i.e., right-sided ovariotomy was performed followed by immediate reappearance of menstruation and reversion from virilisation to feminisation. She became pregnant within 7-8 months after the operation and there was no recurrence during follow-up for over three years.

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Figs. on Art Paper XI