

Case Report

A THECOMA OF OVARY CAUSING PRECOCIOUS PUBERTY IN A BABY AGED 6 MONTHS

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Although many cases of precocious puberty due to functioning ovarian neoplasms have been reported, they remain rare. From the point of view of frequency they are probably the least common cause of sexual precocity. In the majority of cases of precocious puberty the aetiology is unknown.¹ Compared with ovarian tumours in general, functional neoplasms of the ovary are relatively infrequent, and only 5% occur before puberty,² so that exploratory laparotomy, in the absence of a palpable adnexal mass, is not warranted in these children. Gross³ states that all granulosa-cell tumours which have produced secondary sexual characteristics in children, have been readily palpable on abdominal examination.

Ovarian tumours producing the female sex hormones, and therefore capable of causing precocious puberty, are feminizing mesenchymomas (granulosa-cell tumours, thecomas and luteomas); teratoma; chorionepithelioma and dysgerminoma. The feminizing mesenchymomas form the most important group. These tumours contain cells similar to the zona glomerulosa and theca interna of the normal ovary. Willis⁴ says they originate in normal follicular tissue or from the bipotential formative ovarian stroma. Novak's view⁵ is that they arise from cells of the early ovarian mesenchyme and that the tumours show either mainly granulosa cells or theca cells or lutein cells. Granulosa-cell tumours usually contain some theca cells and, though thecomas are more usually 'pure', they often contain granulosa cells in certain areas. A luteoma is probably a granulosa-cell tumour which has undergone luteinization.

USUAL HISTORY AND PRESENTATION

These tumours cause enlargement of the uterus, endometrial hyperplasia and bleeding, which may vary in amount and may be continuous or intermittent. In some it is cyclical, and in some it may only occur after removal of the tumour. The vulva and vagina develop and cornification of the vaginal epithelial cells occur as in the adult. Secondarily induced breast development and growth of sexual hair commonly occur, but are not invariable. Body growth is stimulated, and the child will be taller, and the bone age on X-ray examination greater, than the real age.

No long-term follow-up of cases with prepubertal mesenchymomas has been reported. It is, however, known that a second similar tumour may develop in the other ovary many years after a granulosa-cell tumour has been removed. The prepubertal sexual changes regress after removal of the tumour in children under 8 or 9 years, but above this age no very significant change occurs.

Thecomas are generally considered benign, although 10 instances of malignancy in adults have been reported. Morris and Scully⁶ consider that some of these tumours,

reported as malignant thecomas, could as well be labelled fibrosarcoma.

If the tumour is intracapsular in a child, only local removal of the tumour, preserving the normal ovarian tissue, if possible, should be carried out. If the pathological report is favourable, nothing further should be done and no radiotherapy given.⁷

Previously Reported Cases

Only 5 thecomas producing precocious puberty have been reported in the English literature.^{1,8-11} We wish to report a sixth case, which is also the youngest recorded case of a thecoma.

CASE REPORT

History

The patient, J.v.T., was a White female infant, aged 6 months, who was admitted to the Transvaal Memorial Hospital for Children, Johannesburg, on 13 September 1963.

The main complaint was vaginal bleeding 4 weeks before admission. It lasted for 5 days and no further bleeding was seen. The mother also noticed breast development at the age of 2 months. The baby was born after a normal full-term pregnancy, delivery was normal and all milestones were reached normally.

Findings

On examination the salient features were enlarged breasts with enlarged areolas and nipples (Fig. 1). The clitoris was not much enlarged but especially striking were the enlarged labia (Fig. 2). On abdominal examination a large suprapubic mass was easily palpable.

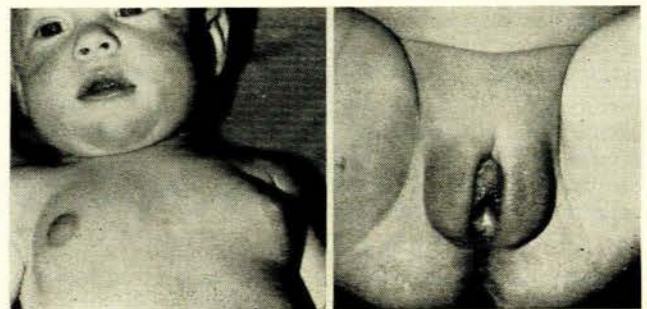


Fig. 1

Fig. 1. Patient, aged 6 months, before operation, showing development of breasts, areolae and nipples.

Fig. 2

Fig. 2. The external genitalia are shown. Note especially well-developed labia majora.

A provisional diagnosis of a feminizing ovarian tumour was made.

Investigations. The following results were obtained from examining the urine and radiographs of the wrist:

1. Urine—24-hour specimen—for oestrogens (volume of specimen—85 ml.): less than 2 µg. of oestriol; less than 2 µg. of oestrone; less than 2 µg. of oestradiol.

2. Urine—24-hour specimen—for 17-ketosteroids (volume of specimen—80 ml.): less than 1 mg. of 17-ketosteroids.

3. Radiographs of wrist showed a bone age of approximately 11-12 months.

Operation. Laparotomy was carried out on 23 September 1963 by Mr. J. Wolfowitz.



Fig. 3. Large ovarian tumour on left, well-developed uterus and right-sided broad ligament cyst are shown at operation.

The abdomen was entered through a transverse lower abdominal incision. A large well-encapsulated left-sided ovarian tumour measuring $7\frac{1}{2} \times 6 \times 4$ cm. approximately was

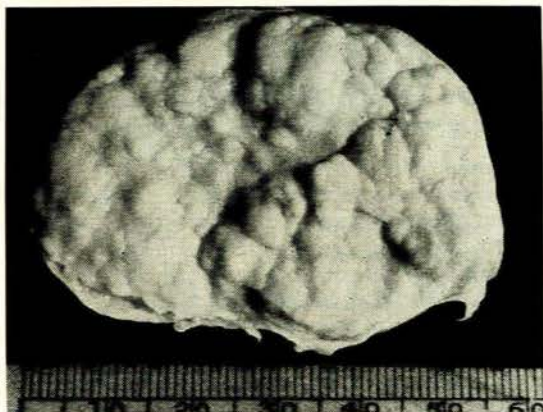


Fig. 4. Macroscopic appearance of ovarian tumour, cut surface showing lobulated appearance and rim of normal ovarian tissue.

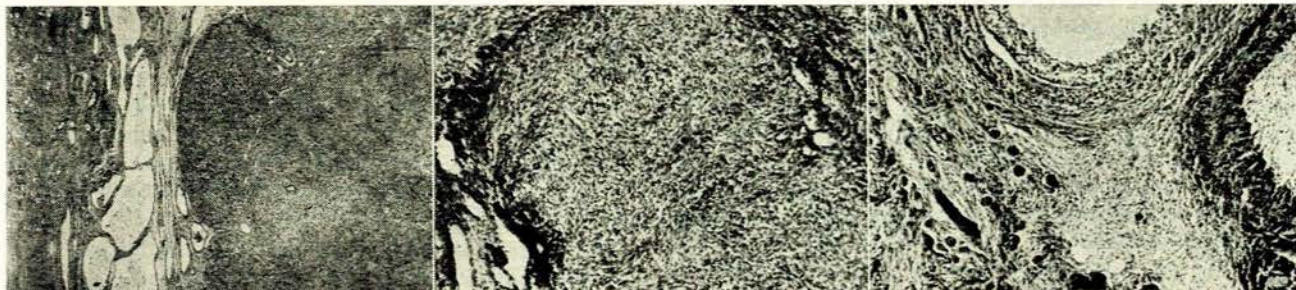


Fig. 5

Fig. 6

Fig. 7

Fig. 5. Photomicrograph (magnification x30) showing lobule of tumour and rim of normal ovarian tissue. Fig. 6. Photomicrograph (magnification x120). Tumour lobule showing interlacing spindle cells. Fig. 7. Photomicrograph (magnification x120) showing ovarian tissue, developing follicles and theca-cell hyperplasia in right ovary.

found, on a pedicle (Fig. 3). On the right side a broad ligament cyst ± 2 cm. in diameter was found. The uterus was markedly enlarged. The mass was removed by transfixing the vascular pedicle. The broad ligament cyst was also removed and a biopsy taken from the right ovary.

Pathology

1. *Tumour of left ovary.* The specimen consisted of an encapsulated oval mass measuring approximately $7\frac{1}{2} \times 6 \times 4$ cm. The outer surface was smooth, and the cut surface showed a firm, yellowish lobulated appearance (Fig. 4).

Sections show the histological features of a thecoma; an outer rim of normal ovarian tissue is present in some sections (Figs. 5, 6, 7).

2. *Right ovary.* Sections of this ovarian biopsy show the presence of infantile ovarian tissue with occasional developing follicles. There is some theca-cell hyperplasia surrounding one of the follicles.

3. *Cyst from right broad ligament.* The specimen consisted of a round thin-walled cyst measuring approximately $2\frac{1}{2}$ cm. in diameter, and containing watery fluid.

Sections show the features of a simple cyst lined by columnar and cuboidal epithelium. This cyst is probably derived from Wolffian vestiges.

Follow-up Examination

When seen 4 months postoperatively, the signs of puberty were receding. In a communication with the mother 11 months later she stated that the child was normal.

Summary

A case of ovarian thecoma occurring in a 6-months-old child is reported.

We should like to thank Mr. J. Wolfowitz under whose care this case was admitted for his advice. Thanks are also due to the Superintendent of the Johannesburg Hospital for permission to publish this case report. We are also grateful to Mr. Max Ulrich for the photographs.

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