PSEUDO-PRECOCIOUS PUBERTY DUE TO GRANULOSA CELL TUMOUR

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Granulosa-theca cell tumours of the ovary are not rare and comprise about 10% of all solid malignant ovarian neoplasms. (Novak and Woodruff, 1967). Ovarian tumours are rare in children and constitute only 1% of all the neoplasia Functional tumours constitute about 30% of the ovarian tumours in children and only 6% of these are found in girls below 10 years of age.

CASE REPORT

(R/73) A female child aged 5 years was transferred from the Kalavathi Saran Children Hospital to Lady Hardinge Medical College and Hospital on 9.10.1973 with a history of a gradual enlargement of both breasts for 3 months and vaginal bleeding for 22 days. There was no history of trauma, head injury or medication or headaches or visual disturbances.

The birth history was not significant and the mile-stones were normal. She had 2 sisters and 3 brothers all of whom were normal and had no endocrinal stigma.

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She was a young child of 5-6 years. Her height was 79 Cm and arm span 94 Cm. The pulse was 100 per minute and regular. There were no abnormalities of the Cardio-Vascular and respiratory systems and her thyroid was normal. There was development of both breasts. The nipples were prominent and the areole showed pigmentation. The axillary and pubic hair particularly over the mons were fine and downy.

The abdomen was soft. A mass 7 Cm x 5 Cm was found arising from the pelvis extending to the right side. It was freely movable, smooth and non-tender. The percussion note over it was dull. There was no fluid thrill or shifting dullness.

The external genitalia appeared to be that of a pubescent girl with well developed labia majora and minora.

Vaginal examination under anaesthesia showed an intact hymen. The cervix was small but the uterus a little bulky for her age. A mass was felt high up in the fight fornix.

A clinical diagnosis of pseudo precocious puberty with a feminising tumour probably a granulosa cell tumour was made.

Investigations

Haemogram was normal except for mild anaemia. Urine-normal. Blood urea was 27 Mg%. X-Ray skull and chest were normal and that of the abdomen showed no evidence of any dermoid.

Bone, dental and mental ages were that of a child of 5-6 years. 17 ketosteroid 0.2 Mg%.

Vaginal swab was sterile and haemagglutination test was n egative. Vaginal cytology showed a highly oestrogenic phase. Matuation index

on 12.10.1973 was 0/26/74 and on 19.10.1973 was 0/13/87.

Hormonal assays could not be done.

Operation Notes

At Laparotomy on 22.10.1973 a small amount of free non-haemorrhagic fluid was found in the peritoneal cavity. The uterus was 3.5 Cm in size and the left tube was healthy and the left ovary was 1½ Cm in size. On the right side there was an ovarian mass 10 x 7 x 3 Cm with a smooth capsule and small yellow irregular nodules on the posterior surface. It was free and encapsulated. No secondaries were found. Right salpingo-oophorectomy was done and a biopsy from the left ovary was taken. On the 5th post-operative day the patient had a fair amount of vaginal bleeding which lasted 3 days. Vaginal cytology on the 10th Day after the removal of the mass showed a shift to the left with a matuation index of 1/55/44 and on the 25th day it was 1.5/98/0.5.

Pathology Grossly, the ovarian mass was 10 x 7 Cm in size with the fallopion tube stretched over it. The surface of the mass was smooth shining and lobulated. It cut with a soft feel and the cut surface revealed several haemorrhagic cystic areas, the largest being 2.5 Cm in size. The intervening solid tissue was soft granular with yellowish tinge.

Microscopically. The tumour was composed of large masses of granular cells surrounded by theca cells. Call Exner bodies were observed.

Left Ovary: Fibro-fatty and fibro-collagenuous tissue. Ovarian pasenchyma cannot be made out.

Follow up: The child has been followed up regularly since and showed regression of the breasts. Vaginal cytology showed a shift to left with presence of intermediate cells-MI being 0/100/0. She was last seen on April 1975 when she was well and asymptomatic.

Discussion

Granulosa-theca cell tumours are rare and constituted 1.7 to 3% of all primary ovarian tumours while in children the incidence is lower still, as only 5% of these occur in the prepubertal age (Morris and Scully, 1958).

Busby (1954) found 2 cases below 10 years in 103 cases and Bland and Goldstein (1955) 8 out of 160.

In India Rajoo in 1945 and Jacob in 1949 have reported a case each and Mehta (1959) commented on 8 cases.

In children these tumours almost always cause sexual precocity. However, the reverse is not true since only 10% of sexual precocity in the female arise from ovarian tumours.

Bone and dental development in this child were around 6 years. Usually sexual precocity in these children does not cause psychological precocity. Ascites may be present but in this child the fluid was scanty and cytologically non-malignant. Exfoliative cytology showed a marked shift to the right.

The breasts particularly the nipples, hypertrophy and the areole gets pigmented. The genitalia both external and internal hypertrophy. Vaginal bleeding is often proceeded by a discharge. This bleeding is anovular in type and since the FSH is usually not increased these patients cannot conceive. Hence, these cases should be termed precocious-pseudo-puberty.

The conservation of the potential of the reproductive capacity in the young girl and the benign nature in the pediatric group prompts the surgeon to do a conservative operation. Usually salpingo-oophorectomy. Pedowitz (1955) found 8 deaths in 49 proved cases in this age group.

Post-Operative regression of the condition can be anticipated in children under 8 years. Withdrawal bleeding occurs after removal and vaginal cytology shows a regression of estrogenic hormone as was in this child.

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