Picture story

Granulosa cell tumour in a 9 month old girl

T P J Gunawardena[1], J A P Dhammika[2], Ajith Lamahewa[3]

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Introduction

Ovarian tumors are a rare cause of precocious pseudopuberty, accounting for around 1% of all cases. Least uncommon is the rare granulosa-theca cell tumor and presentation is rare before 4 years[1].

Case report

A previously healthy 9-month-old girl with a normal birth history and development presented with a history of significant per vaginal bleeding for 4 days duration (Figure 1).

Figure 1 Per vaginal bleeding

Her anthropometric measurements were as follows. Length 70 cm (50th centile), occipito-frontal circumference 42 cm (3rd centile) and weight 10.5 kg (97th centile). She had mild pallor (haemoglobin 9.0 g%) and her breast and genital development were prepubertal. On palpation of abdomen a lump arising from the pelvis was observed. This was later confirmed by ultrasound and magnetic resonance imaging (MRI) scan of the pelvis as a solid lump arising from right ovary with enlarged uterus containing a greatly thickened endometrium.

Oestradiol levels were greatly elevated at 608.0 pg/ml (less than 40 in prepubertal girls) with normal levels of human chorionic gonadotrophin (HCG) and alpha fetoprotein (AFP) levels. Gonadotrophin levels were unmeasurable with the available assays. Her bone age was slightly advanced to 18 months.

At surgery a well encapsulated tumour arising from right ovary of 7 x 6cm was removed (stage Ia) There were no tumor deposits in the peritoneal cavity (Figure 2).

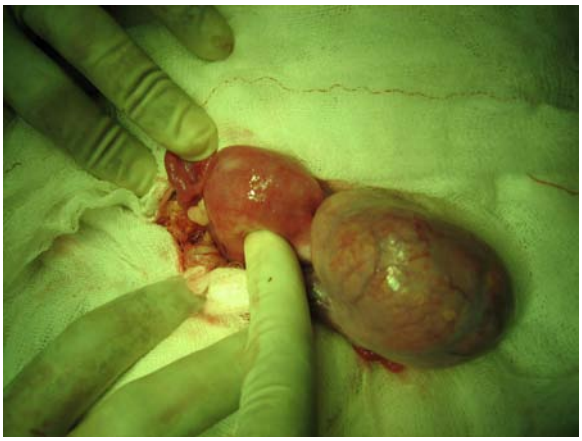

Figure 2 Well encapsulated tumour

Microscopic appearance was that of a juvenile granulosa cell tumor with cells containing rounded moderately hyperchromatic nuclei and abundant eosinophilic cytoplasm.

Discussion

At presentation this girl had bleeding per vagina for 4 days duration in the absence of breast development and genital hair growth. The differential diagnoses considered were isosexual precocious pseudopuberty, foreign body and genital rhabdomyosarcoma.

[1]*Senior lecturer, Department of Paediatrics,* [2]*Senior Lecturer, Department of Obstetrics and Gynaecology,* [3]*Senior Lecturer, Department of Pathology, Faculty of Medicine, Galle.*

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However, clinical finding of a pelvic lump made the latter two possibilities unlikely.

Following surgical treatment bleeding quickly improved and she was referred to the oncologist for a decision regarding further treatment.

Granulosa-theca cell tumors, more commonly known as granulosa cell tumors (GCTs), belong to sex cord-stromal group origin. GCTs account for approximately 2% of all ovarian tumors and can be divided in to adult (95%) and juvenile (5%) types[2]. GCTs are thought to be tumors of low malignant potential, most following a benign course with only a small percentage showing aggressive behaviour[3]. More than 90% are diagnosed before spread occurs outside ovary. Five-year survival rates usually are 90-95% for stage I tumors.

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