Recurrence of Sertoli-Leydig cell tumour in contralateral ovary. Case report and review of literature

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Key words: Sertoli-Leydig cell tumour.

Case Report

An 18-year-old "A" level student presented with a history of primary amenorrhoea in October 1997. She had no abnormalities of note on examination. The initial ultrasound report suggested a right ovarian mass. Her FSH, LH, AFP & hCG levels were normal. The serum estradiol level was 73.4 (normal range 91-1148) and CA125 was 69ku/l (normal range 0-65). She had normal female karyotype (46XX) on chromosomal analysis with no evidence of numerical or structural abnormality. Laparoscopy followed by laparotomy, right salpingo-oophorectomy and biopsy of the left ovary was performed for a 14x10x10 cm complex mass emanating from the right ovary. Bowel, omentum, liver, gall bladder and left ovary appeared normal. Macroscopically, it was a partly cystic but mainly solid ovarian neoplasm composed of lobules of fawn coloured tissue with areas of haemorrhage and mucoid degeneration. Histopathology revealed a poorly-differentiated sex cord stromal tumour of Sertoli-Leydig cell type with heterologous mucinous epithelium and focal retiform glandular differentiation.

High mitotic activity was noted. It was strongly positive for immunohistochemical staining for alpha-inhibin. The biopsy from the left ovary showed no evidence of malignancy. The patient had her first menstrual period four weeks after surgery.

MRI done in February 1998 revealed an abnormal left ovarian cyst with a few septations (3.5x2.5 cm) and although this could have been due to a large follicle a transvaginal ultrasound scan (USS) was suggested to confirm this possibility. There was no evidence of pelvic lymphadenopathy or ascites. The following USS did not confirm this and she was followed-up regularly with USS tumour markers and clinical examination. In January 1999 the ultrasound revealed a left adnexal mass. Laparoscopy was performed which showed a suspicious left ovary about 12 cm in diameter. There was no obvious capsular rupture. No obvious peritoneal or hepatic disease was visualised. The fluid aspirated from the Pouch of Douglas contained isolated suspicious cells with irregular nuclei and coarse chromatin but unequivocally malignant cells could not be identified. The ovarian aspirate showed poorly-differentiated sex cord tumour-like cells. The ovarian biopsy showed evidence of malignancys under to that previously identified on the right side.

In late January 1999, when she was referred to our centre, her hormonal profile was within normal limits and CA125 was 91ku/L. The MRI confirmed a mass within the pelvis which displaced the rectum and also abutted onto the bladder wall, but with no obvious evidence of invasion. Chest CT scan was normal. In February 1999, laparotomy undertaking total hysterectomy, left salpingo-oophorectomy, para-aortic and pelvic lymph node biopsies and infracolic omentectomy was performed.

The histopathology revealed a poorly-differentiated Sertoli-Leydig cell tumour (SLCT) with probable rhabdomyoblastic heterologous elements, with no evidence of metastatic disease to the uterus, pelvic and para-aortic lymph nodes or omentum. The patient underwent adjuvant chemotherapy with bleomycin, etoposide and cisplatin for 4 cycles and is currently in remission.

Discussion

Sertoli-Leydig cell tumours are sex cord-stromal tumours which exhibit testicular differentiation. They make up less than 0.2% of ovarian neoplasms in total but they account for 4% of ovarian tumours in patients under 20 years of age [1]. Seventy-five per cent of the patients are aged 30 years or less. Most SLCTs are unilateral (97-98%) and one-third manifest with signs of virilisation such as amenorrhea, hirsutism, and hoarseness and deepening of voice [2]. Sometimes they may present with a pelvic mass. According to the classification of the World Health Organisation, well-differentiated, intermediately-differentiated, poorly-differentiated forms, as well as those with heterologous elements or retiform patterns can be discriminated histologically [1]. It is based on morphology, implying neither potential hormonal activity nor a specific histogenesis.

Most of the sex cord stromal tumours are either benign or of low malignant potential. The morphological appearance of these tumours varies more widely than that of any other ovarian tumour except for the teratomas. Poorly-differentiated SLCTs may resemble early stages of testicular development. It is for this reason that terms like "arrhenoblastoma" or "androblastoma" have been used. Inspite of this misleading terminology some of the tumours may be endocrinologically inactive or oestrogenic [3]. Heterologous elements like intestinal epithelium, muscle or cartliage are present in about 20% of the neoplasms [4].

There are no Sertoli cells in the normal adult ovary. In neoplasia, the ovarian cells of the sex cord origin manifest capacity for testicular differentiation. The Leydig cells which normally are seen in the hilus, move to the medullary and the cortical stroma in neoplasia [5].

Because of their relative rarity, the diversity of histologic patterns, associated confusing terminology and their variable biologic behaviour, there are no large series of

Revised manuscript accepted for publication September 30, 1999

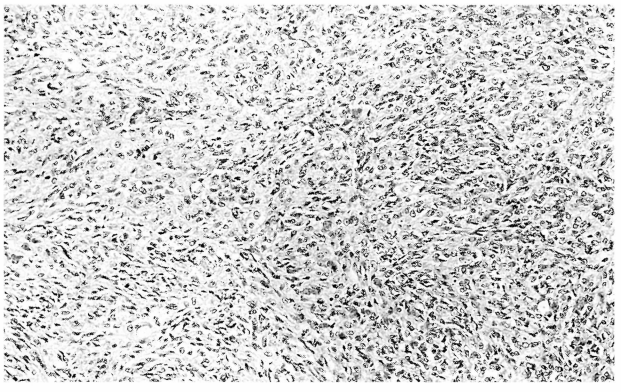

Figure 1. — Haematoxylin and eosin stained section of the tumour showing poorly-differentiated, sarcoma-like area of the tumour.

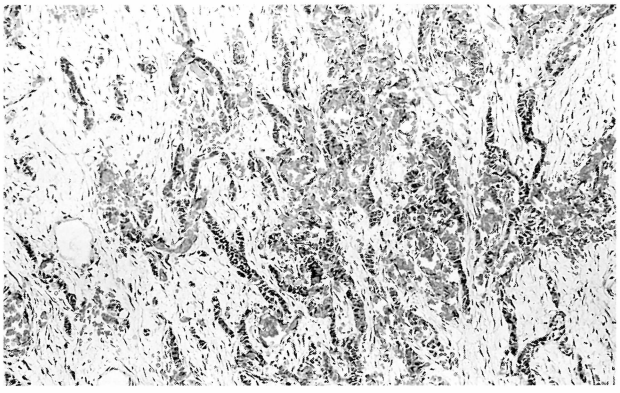

Figure 2. — A better differentiated area of the tumour stained by the Haematoxylin and eosin method showing sertoliform cords and tubules.

patients appropriately staged and managed uniformly and their optimal treatment remains undecided. Surgery remains the cornerstone of treatment. Since most of the patients are in stage IA, a favourable outcome can be achieved with conservative surgery. Unilateral adnaexectomy is the surgery recommended when conservation of fertility is important. When childbearing is complete, a total abdominal hysterectomy and bilateral salpingo-oophorectomy is performed. However management of patients postoperatively is confusing because of insufficient understanding of the associated prognostic factors. Poor prognostic factors are poor differentiation, retiform pattern or heterologous elements and metastases. Incidence of malignancy in these tumours is estimated to be between 10 and 30% [3, 6]. In such cases, late recurrences even in early stages can be detected which can prove to be fatal inspite of aggressive therapy [1]. The most reliable indication of malignancy is local extraovarian spread or metastatic deposits at the time of staging laparotomy. All such tumours follow an aggressive and generally fatal course. As many as 60% of these tumours with high mitotic activity pursue a malignant course. Alpha and beta inhibin produced by human sex cord stromal tumours might be a useful tumour marker [7]. Beta-hCG and carcinoembryonic antigen (CEA) are usually normal though rarely alpha-fetoprotein may be raised [8].

The overall response rate to the combination of bleomycin, etoposide and cisplatin in poor prognosis sex cord stromal tumours of the ovary is high. However, even after commencing chemotherapy in this group, the question whether a 50% or higher rate of relapse is reduced still is unanswered [9]. Although the best chemotherapy for SLCT is not known, a platinum-based one is generally favoured. The combination of Paclitaxel and a platinum drug is being tried at present.

The differential diagnoses which one has to consider with SLCT are granulosa cell tumour and sertoliform endometroid carcinoma. The pattern of recurrence of the granulosa cell tumour and SLCT is different from each other. Granulosa cell tumours may recur late. When SLCTs do recur, it usually happens within one year of initial diagnosis. The spread is characteristically to the omentum, abdominal lymph nodes or liver [10]. The recurrence just in the opposite ovary is quite rare. As the left ovary at the time of the original operation in our patient appeared normal, metastases to the left ovary was more likely than tumour bilaterality. We are not aware of a case report in the literature where metastases was confined only to the contralateral ovary. Trending: none.

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