Introduction

Sertoli-Leydig cell tumours (SLCTs) are rare, unilateral neoplasms of the stroma of the ovary constituting less than 1% of all ovarian neoplasms. Within SLCTs we may differentiate a separate group of neoplasms that secrete α-fetoproteins (AFP). Histopathogenesis and further prognosis in case of...
α-fetoprotein-producing SLCTs were not conclusively investigated, but one has to take into account the potential role of gene mutations (e.g. DICER1 on chromosome Y) in the diagnostics of this inhomogeneous group of neoplasms. To make a correct diagnosis, many factors have to be considered, including clinical signs and symptoms, immunohistochemical analysis and imaging results. The aim of this quiz was to present a case of a young female patient, who presented with hyperandrogenism and secondary amenorrhoea.

Case report

A 24-year old female patient reported to the Clinic of Endocrine Gynaecology at the University Hospital in Krakow with symptoms of secondary amenorrhoea, hirsutism (31 points in Ferriman-Gallwey score) and increasing lower abdominal pains of several months’ duration. The last spontaneous menstruation occurred over 3 years ago.

During the first hospitalization, a set of basic diagnostic tests was performed. Detailed analyses of hormonal levels as well as tumour markers pointed to significantly elevated testosterone levels (8.16 nmol/l – with norms between 0.29-1.67 nmol/l) and α-fetoprotein levels (10.02 IU/ml – with norms 0.0-5.8 IU/ml). Values of other parameters such as: cancer antigen 125 (CA-125), carbohydrate antigen 19-9 (CA 19-9) and carcinoembryonic antigen (CEA) were normal.

Furthermore, the extent of hirsutism was evaluated using the 36 degree Ferriman-Gallwey scoring system, where the value below 8 points is correct. The woman self-assessed her symptoms of hirsutism with a score of 31 points and this value confirmed the androgen excess (Fig. 1).

Imaging study results

During hospitalization an ultrasound examination of the abdomen was performed. An echogenically changed structure of the right ovary was noted, measuring 42 × 27 mm (Fig. 2), with no clear demarcation of the tumour, showing signs of increased peripheral vascularisation (Fig. 3).

Owing to low sensitivity of the ultrasound, pelvic magnetic resonance imaging (MRI) was performed. The MRI showed a well demarcated solid-cystic tumour measuring 44 × 32 × 40 mm, with small cystic spaces of 5 mm in diameter, localized in the posterior part of the right ovary. The lesion was adherent to the uterine cervix, bulging into the Douglas pouch and partially abutting the rectal wall. Considering the imaging study results and the clinical data, there was a high probability for the lesion to be a Sertoli-Leydig cell tumour or thecoma or fibroma or granulosa cell tumour.

Surgical procedure

The patient was qualified to undergo a laparotomy and unilateral removal of the right ovary (Fig. 4). Owing to the young age and nulliparity, it was of highest concern to preserve patient’s fertility – so it was necessary to perform ovary reserve testing, which before the procedure was 6.0 ng/ml. On the day of the procedure, the control value of testosterone was 10.81 nmol/l (normal values: 0.29-1.67 nmol/l),
which meant an increase of about 2.65 nmol/l over 30 days, since the first hospital admission.

During the procedure, the right ovary was removed using a classical approach and then a frag-
ment of the left ovary and peritoneum were collected for further pathological examination.

The procedure and postoperative period were tolerated well and the patient was discharged home on the fifth day of hospitalization.

Results of histopathological studies

Histopathological and immunohistochemical studies confirmed the initial diagnosis of Sertoli-Leydig cell tumour (Figs. 5 and 6). Such neoplasms may be divided basing on histopathological examination into three main subtypes: well, intermediately and weakly differentiated. Two last categories may additionally include ~20% of heterologous elements, most of them being glands and cysts lined with intermediately or well differentiated gastric or intestinal epithelium. In the presented case, next to Sertoli-Leydig cells, the structure of the tumour included glandular cells from of the colonic epithelium (Figs. 7 and 8). The fragment from the left ovary did not show any pathological finding.

Follow-up visits

During the first follow-up 10-days after the surgical procedure, biochemical laboratory test results normalized, with testosterone 0.50 nmol/l (norms: 0.29-1.67 nmol/l) and AFP 3.12 IU/ml (norms: 0.0-5.8 IU/ml). The patient was in good general condition and had no complaints. The last follow-up visit was 90 days after the procedure. Laboratory test results were still normal for her age and sex, with testosterone value of 0.53 nmol/l and AFP at 0.98 IU/ml. Additionally the patient reported spontaneous menses. During the last follow-up visit, hirsutism was again evaluated using Ferriman-Gallwey score reaching 13 points with results shown in Figure 9 and anti-Müllerian hormone (AMH) level after removal of the right ovary was 4.7 ng/ml.

Discussion and conclusions

We presented a case of Sertoli-Leydig cell tumour with heterogeneous elements of glandular colonic cells producing α-fetoprotein. So far the medical literature lists approximately 30 similar cases [1, 2, 3, 4, 5, 6, 7, 8, 9, 10, 11, 12, 13, 14, 15, 16, 17, 18, 19, 20, 21, 22, 23, 24, 25]. Sertoli-Leydig cell tumours most commonly (75%) occur in young females up to 30 years of age [26, 27]. The significant majority of them are unilateral and at the time of diagnosis they are limited to the stroma of the ovary [26, 27, 28]. 30-50% of SLCs produce androgens (testosterone and other androgen precursors), which are responsible for signs and symptoms of androgen excess, such as oligo or amenorrhea, hirsutism
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References


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