Bilateral Sertoli cell adenoma in gonads, associated with serous cystadenoma

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Complete androgen insensitivity syndrome is an extremely infrequent disease. The patients exhibit female phenotype because of insensitivity to the androgen receptor and may develop tumors, especially in their undescended gonads. We report a case of bilateral Sertoli cell adenoma in gonads with unilateral serous cystadenoma, in an elderly phenotypic woman with primary amenorrhea. We also provide radiological and pathological studies.

Key words: androgen insensitivity syndrome, gonadal cyst, Sertoli cell tumor.
Fig. 1. Different imaging studies performed on the abdominal mass. A) CT scan shows the bilateral solid tumor with a cystic part on the right side (arrowed). B) Gross features of the tumor. The inset shows some pieces of the right-sided tumor with some fragments of the cystic part. C) Microscopic image of typical tubules from a well-differentiated Sertoli cell tumor. The inset displays a monolayered epithelium composed of columnar cells lining the cystic cavity (HE staining). D) Immunohistochemistry for inhibin shows strong cytoplasmic positivity in tumoral cells. E) The tumor cells display irregular and weak to moderate staining for calretinin. All immunostaining was performed by immunoperoxidase technique on paraffin-embedded tissue sections.
ical and pathological findings [1]. The patient is still being followed up regularly and she is healthy.

Although the risk of developing tumors in women with CAIS increases with age, Sertoli cell adenomas are not the most frequent [2]. However, many cases of CAIS developing pure bilateral Sertoli cell adenoma have been previously described, but only a few with unilateral serous cystadenoma associated [3, 4].

In conclusion, the integration of clinical symptoms, physical examination, CT body scan and pathology studies is essential to confirm an accurate diagnosis of this uncommon syndrome.

The authors declare no conflict of interests.

References

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