

Extraovarian granulosa cell tumor: A case report

Ekstraovaryen granulosa hücreli tümör: Olgu sunumu

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Abstract

Granulosa cell tumor (GCT) of the ovary is a rare tumor representing 2 to 5% of all ovarian neoplasms. It can reproduce or metastasize several years after the initial treatment. In rare cases, CGT can develop on an extra-ovarian site, which is thought to originate from ectopic gonadal tissue along the embryonic genital ridge. We herein present a case of extraovarian granulosa tumor, the clinical and therapeutic aspects, and our approach.

Keywords: Extraovarian, Granulosa cell tumor, Histological diagnosis

Öz

Overin granuloza hücreli tümörü (GCT), tüm over neoplazmalarının %2-5'ini oluşturan nadir bir tümördür. Tedaviden birkaç yıl sonra yeniden ortaya çıkabilir veya metastaz yapabilir. Sık olmamakla birlikte, embriyonik genital katlantı üzerindeki ektopik gonadal dokudan köken alarak, ekstra-ovaryen bir bölgede de görülebilir. Burada bir ekstraovaryen granuloza tümörünü, klinik, terapötik yönleri ve kendi yaklaşımımızla birlikte sunuyoruz.

Anahtar kelimeler: Ekstraovaryen, Granulosa hücreli tümör, Histolojik tanı

Introduction

Tumors of adult granulosa cells are considered as sexual tumors of the cord and ovary [2]. There is considerable debate about the cellular origin of these tumors. In the literature, several cases of extraovarian tumors with granular cells have been reported [3].

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Case presentation

A 66-year-old female patient with no significant pathological history presented with severe abdominal pain. Physical examination revealed the presence of a large mass at the left flank. An abdominopelvic scan showed the presence of two large ganglionic clusters measured respectively at 13 and 11 cm, the first one extending from the subcarinal region to the renal hilum, and the second one extending along the left psoas muscle (Figure 1). Histopathological examination of the biopsy (confirmed by immunohistochemistry) revealed adult granulosa cell tumor characteristics (Figure 2). Patient was diagnosed with extra-ovarian GCT, and therapeutic strategy was based on chemotherapy.



Figure 1: CT image shows the location of the granular tumor

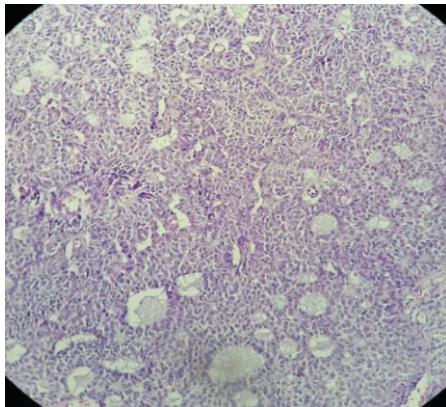


Figure 2: Microscopic image shows the histological appearance of the extraovarian granulosa tumor

Discussion

An uncommon ovarian cancer [1], adult and juvenile type GCTs exhibit different clinical and histological features. The more common adult type of GCT usually presents during the perimenopausal or the early menopausal period, the median age of diagnosis being 50-54 years [1]. GCT patients require long-term follow-ups with proper history-taking, physical examination, and tumor marker studies because 17% of relapses occur more than 10 years after diagnosis [3]. The most common site of recurrence is the pelvis. Rarely, extraovarian GCT can develop. Eight such cases have so far been reported in the English literature until 2001 [3], one of which was from India [5]. The tumors of four cases originated from the broad ligament, three from the retroperitoneum and one from the adrenal gland [3]. Recently, one case of GCT arising in a Müllerian cyst of the broad ligament has been reported [6].

The histogenetic origin of extraovarian sex-cord stromal tumors is considered as the ectopic gonadal stromal tissue, with the sex-cord originating from the mesonephros. A dual origin theory from both coelomic epithelium and mesonephros has also been proposed [7]. Mesonephros or its influence seems necessary for creating the sex-cord. This may explain why the sites of extraovarian sex-cord stromal tumors being limited to the broad ligament, the retroperitoneum, and the adrenal gland, all of which differentiate close to the mesonephros and the mesonephric duct [7]. The morphological differential diagnosis of GCT includes undifferentiated carcinoma, small cell carcinoma and endometrial stromal sarcoma [1]. However, characteristic histological findings and immunostaining are helpful in definitive diagnosis.

Several tumor markers are used for confirmation of histologic diagnosis of GCT, an important one being inhibin. The ovary is the only source of inhibin in non-pregnant woman whereas in pregnancy, it is also secreted from the placenta. It is reportedly a more reliable marker for GCT than estradiol. Although an elevated inhibin level may be observed in some epithelial ovarian cancers, confirmation is based on EMA positivity, which is negative in GCT [10].

Our patient presented with two separate intraabdominal ganglionic clusters, which is unusual for a primary tumor. GCT can recur or metastasize years after initial diagnosis and treatment, hence the strong possibility of metastatic deposit in intraabdominal sites. As slides of the excised ovaries were not available for thorough review, the possibility of late metastasis of a low-grade GCT could not be evaluated, and the diagnosis of extraovarian GCT was deemed most appropriate.

Conclusion

Tumors of adult granulosa cells are considered sexual tumors of the cord and ovary, which poses a diagnostic problem. A diagnosis of an extraovarian GCT can be made by excluding any history of ovarian GCT.

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