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Case study

Juvenile granulosa cell tumor arising in ovarian adenosarcoma: an unusual form of sarcomatous overgrowth

Claire Carleton MD, Oisín P. Houghton FRCPath, W. Glenn McCluggage FRCPath*

Dept of Pathology, Belfast Health and Social Care Trust, Belfast, Northern Ireland BT126BA

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Summary We report 2 ovarian neoplasms in women aged 58 and 69 years composed of an admixture of adenosarcoma and a predominant stromal component morphologically and immunohistochemically in keeping with juvenile granulosa cell tumor. As far as we are aware, this association has not been reported previously. We speculate that, in both cases, the juvenile granulosa cell tumor component arose from the adenosarcoma as an unusual form of sarcomatous overgrowth of sex cord elements.

1. Introduction

Mullerian adenosarcomas are uncommon mixed epithelial and mesenchymal neoplasms containing a benign epithelial element and a malignant, usually low-grade, stromal component [1,2]. They most commonly arise in the uterus, especially the corpus [3,4], but rarely occur as primary neoplasms in extrauterine sites, especially the ovary [5]. Occasionally, the stromal component of both uterine and extrauterine adenosarcomas contains so-called sex cord–like elements [1,5,6]. Although referred to as sex cord–like, these may exhibit a true sex cord immunophenotype with positive staining with inhibin, calretinin, and other sex cord markers [5,6]. We report 2 unusual ovarian neoplasms composed of an admixture of adenosarcoma and a predominant stromal component morphologically and immunohistochemically in keeping with juvenile granulosa cell tumor (JGCT). As far as we are aware, this combination of neoplasms has not been previously reported, and we speculate that the JGCT arose from the adenosarcoma, probably from sex cord elements, this representing an unusual form of sarcomatous overgrowth. In reporting these cases, we review the phenomenon of sex cord elements occurring within ovarian neoplasms, which are not intrinsically of sex cord lineage.

2. Case reports

Case 1 was an in-house case from the Department of Pathology, Belfast Health and Social Care Trust, Belfast, and case 2 was from the referral practice of one of the authors (W.G.M.). The hematoxylin and eosin–stained slides (40 in case 1 and 10 in case 2) and immunohistochemistry slides were reviewed.

* Corresponding author. W. Glenn McCluggage, Department of Pathology, Royal Group of Hospitals Trust, Grosvenor Road, Belfast BT12 6BA, United Kingdom.
E-mail address: glenn.mccluggage@belfasttrust.hscni.net (W. G. McCluggage).

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2.1. Case 1

2.1.1. Clinical details

A 69-year-old woman presented with a 6-month history of abdominal discomfort, decreased appetite and, nausea and was found to have a pelvic mass. Ultrasound and computed tomographic scans showed a large pelvic mass probably arising from the left ovary with a small amount of ascites. No extraovarian spread was seen. The serum CA125 was elevated at 551 U/mL (normal, <35 U/mL). She had undergone a hysterectomy for uterine fibroids 34 years previously. At laparotomy, there were ascites and a large solid left ovarian mass that was stuck to the sigmoid mesentery. Tumor was also present in the Pouch of Douglas. The right ovary, upper abdomen, and omentum appeared normal. She underwent bilateral salpingo-oophorectomy and omentectomy. Pelvic peritoneal tissue from the Pouch of Douglas containing tumor nodules was also removed. After the pathology result and discussion at the multidisciplinary gynecologic oncology meeting, it was decided to follow the patient up, and no adjuvant therapy was administered. The case is recent, and there is no follow-up.

2.1.2. Pathologic findings

The left ovary measured 20.5 cm in maximum dimension and weighed 1200 g. The capsule was disrupted in several places. On sectioning, the ovary was composed entirely of a predominantly solid tumor with occasional small cystic areas. The solid areas had a slightly nodular, rubbery, pale yellow cut surface with occasional areas of hemorrhage. The left fallopian tube was not identified grossly. The peritoneal tissue from the Pouch of Douglas contained grossly visible tumor nodules. The right ovary (1.5 cm in maximum dimension), right fallopian tube (5 cm in length), and omentum (30 cm in maximum dimension) were grossly unremarkable. Fifteen milliliters of ascitic fluid was also submitted.

Histology of the left ovary showed a low-power lobulated architecture with multiple cellular nodules separated by edematous stromal tissue. The nodules were composed predominantly of a diffuse population of cells with moderate to abundant, pale or eosinophilic cytoplasm. Many follicle-like spaces containing basophilic fluid were present within the otherwise diffuse arrangements. The cells were epithelioid with atypical nuclei, and there was marked mitotic activity (>20 per 10 high-power fields). There were areas of necrosis. These areas were morphologically in keeping with JGCT. The peritoneal tissue from the Pouch of Douglas contained similar tumor.

In 2 of the 40 slides, there was a different morphological appearance in the form of a polypoid "club-like" (phyllodes-like) architecture with benign ciliated epithelium lining stromal cores. The cells within the stromal cores contained spindle-shaped nuclei with mild to moderate nuclear atypia, and there was condensation around the epithelium forming a cambium layer within which mitotic figures were present, in excess of 20 per 10 high-power fields. The features in this component were in keeping with adenosarcoma. Overall, approximately 95% of the neoplasm was composed of the JGCT component. There was a relatively sharp transition between the 2 components with minimal intermingling. Fig. 1 shows representative images of the neoplasm.

Immunohistochemistry of the JGCT component showed diffuse positivity for calretinin (nuclear and cytoplasmic), CD56 (membranous), steroidogenic factor 1 (nuclear), and CD99 (membranous) (Fig. 2). There was focal cytoplasmic positivity with inhibin and focal nuclear staining with progesterone receptor. Melan A, estrogen receptor (ER), epithelial membrane antigen, and AE1/3 were negative. The immunophenotype was in keeping with an ovarian sex cord tumor. The epithelial element of the adenosarcoma component was diffusely ER positive, and the stromal element was negative.

Histology of the right ovary showed a focus of endometriosis. The right fallopian tube and the omentum exhibited no histologic abnormality. Cytologic examination of the ascitic fluid showed no malignant cells.

Because there was involvement of the pelvic peritoneal tissue, the tumor was staged as FIGO IIB.

2.2. Case 2

2.2.1. Clinical details

A 58-year-old woman was found at a routine health check to have an elevated serum CA125 of 72 U/mL. Ultrasound and computed tomographic scans showed a large right-sided pelvic mass probably arising from the right ovary. There was no extraovarian spread and no ascites. She had undergone a hysterectomy for benign reasons 23 years previously. She underwent a bilateral salpingo-oophorectomy, appendectomy, and omentectomy. After the pathology result and discussion at the multidisciplinary gynecologic oncology meeting, it was decided to follow the patient up, and no adjuvant therapy was administered. The patient has been followed up for 20 months with no evidence of tumor recurrence or metastasis.

2.2.2. Pathologic findings

The right ovary consisted of a cystic mass with a smooth capsule measuring 18 cm in maximum dimension. Sectioning revealed a partly cystic and partly solid appearance. The right fallopian tube was not identified. The left ovary (2.5 cm in maximum dimension), left fallopian tube (5 cm in length), omentum (13 cm in maximum dimension), and appendix (10 cm in length) were grossly unremarkable.

Histology of the right ovary showed a minor component with a polypoid club-like (phyllodes-like) architecture with benign ciliated and endometrioid-type epithelium lining stromal cores. The stromal component was morphologically bland with spindle-shaped nuclei, but there was increased cellularity surrounding the epithelial elements forming a cambium layer. Mitotic figures were identified within the cambium layer (several within a single high-power field in areas). This component was in keeping with adenosarcoma. In addition, there was a predominant component similar to the JGCT described in case 1 with diffuse areas, follicle-like