Ovarian mucinous cystadenoma with extended calcification in an 11-year-old girl: case report and review of the literature

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Summary

The majority of ovarian masses in childhood and adolescence are non-epithelial in origin, with a predominance of germ cell tumors, while epithelial neoplasms comprise a small proportion of the total (approximately 15-20%). Mucinous cystadenomas in particular are only sporadically reported in this age group. We present a case of an ovarian mucinous cystadenoma with extended calcification in a premenarchal 11-year-old girl. Pediatric mucinous cystadenomas of the ovary may on rare occasions display extended calcification. Careful evaluation of the remaining pathological features of the tumor is needed in order to avoid misinterpreting this relatively non-specific finding as a feature of malignancy.

Key words: Calcification; Cystadenomas; Mucinous; Ovarian.

Introduction

Surface epithelial neoplasms of the ovary are commonly found in adult women and account for 60-70% of all ovarian masses. However, in childhood and adolescence the majority of ovarian masses are non-epithelial in origin, with a predominance of germ cell tumors, while epithelial neoplasms comprise a small proportion of the total (approximately 15-20%) [1]. We present a case of a mucinous ovarian cystadenoma with an extensive deposit of calcifications in an 11-year-old premenarchal girl, and discuss the histogenesis and pathological significance of calcifications in epithelial ovarian neoplasms.

Case report

A previously healthy premenarchal 11-year-old girl presented to the Outpatient Department of our hospital with acute abdominal symptomatology. Pelvic examination was suggestive of a tumor in the left adnexa. Ultrasonography of the lower abdomen showed a large multiloculated mass in the left ovary measuring 6.8 x 4 x 6.2 cm. The right ovary was normal and there was no sign of ascites. Blood tests, including serum levels of estradiol, progesterone, total testosterone, free testosterone and sex hormone binding globulin, as well as carcinoembryonic antigen (CEA), CA125, CA15-3 and alpha-fetoprotein (AFP) were all within normal limits. The patient was submitted to laparotomy and the cyst was removed intact and sent to our pathology laboratory for histological examination. On gross examination the specimen appeared as a cystic multilocular mass measuring 5.5 x 3 x 0.5 cm. The surface was smooth and the thickness of the walls ranged from 0.3-0.5 cm. Frozen section biopsy of the tumor showed a neoplasm consistent with a mucinous cystadenoma. The entire remaining specimen was subsequently serially sectioned and studied histopathologically. Microscopic examination showed that the cyst was lined by a single layer of uniform small columnar epithelial cells of endocervical type invaginating into the stroma to form clefts. No multilayering, necrosis, atypia or mitotic activity were noted. Large deposits of calcifications were seen covering the cyst walls to a remarkable extent (approximately 50% of the surface), as well as within the epithelium, thus resembling psammoma bodies (Figures 1 and 2). The final diagnosis was mucinous ovarian cystadenoma with extended calcification.

Menarche occurred one year later and has remained regular ever since. The patient is well without any recurrence, gynecological complication or any relative disorder two years following the operation.

Discussion

Less than 20% of ovarian masses in childhood are epithelium-derived, and most previous studies and literature reviews have indicated a preponderance of serous cystadenomas among epithelial ovarian tumors in this age group [2]. Mucinous ovarian cystadenoma is therefore an extremely rare entity in childhood, especially before menarche.

Dystrophic calcification represents a local disturbance of calcium homeostasis, triggered by various initiators and encouraged by aging [3, 4]. Certain tumors, especially in the breast and ovary tend to calcify more often, and this dystrophic calcification is traditionally considered to develop within areas of necrosis or as a result of epithelium degenerative changes associated with inflammation or ischemia [3, 5]. A secretory mechanism has also been proposed on the basis of the absence of stainable DNA and RNA in calcified areas [5, 6]. Silva *et al.* recently presented both experimental and clinicopathologic data, suggesting that ovarian calcifications may be due to metabolic changes related to hormones [5].

Revised manuscript accepted for publication April 23, 2006

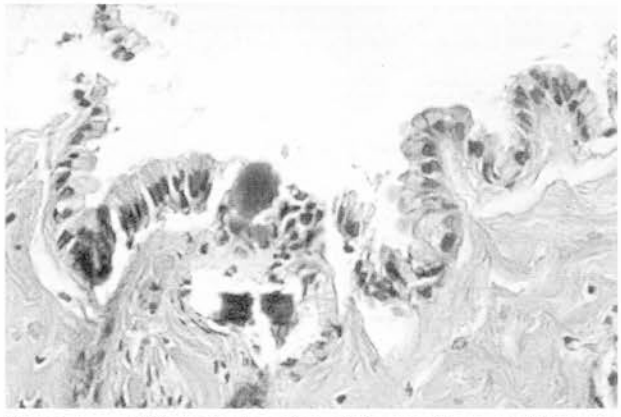

Figure 1. — Histological section of the mucinous cystadenoma showing intraepithelial calcifications resembling psammoma bodies (H-E x 250).

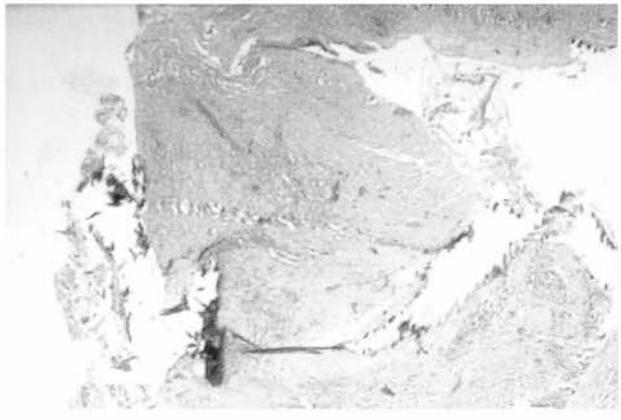

Figure 2. — Histological section of the cyst wall showing extensive stromal calcification (H-E x 120).

Several authors have previously reported that dystrophic calcifications are relatively commonly found in simple mucinous ovarian cysts and cystadenomas, as well as in borderline or carcinoma ovarian specimens [7, 8]. Calcifications have also been noted in normal ovaries and are associated with stromal hyperplasia, as well as in other neoplasms, such as teratomas, sex-cord stromal tumors and gonadoblastomas [5, 7, 9]. Their extent may be variable, ranging from microscopic findings or even diffuse impregnations found by chemical analysis only, to macroscopically visible masses [3]. In the former case they may be missed, unless the entire specimen is studied. Extended calcification has been described in neonates and children, in association with ovarian torsion and subsequent autoamputation and necrosis [10, 11]. However, there is no previous report – to our knowledge – describing a mucinous ovarian cystadenoma with extended calcification in a pediatric patient.

Relatively few studies have focused on the clinicopathologic significance of calcifications in mucinous ovarian tumors. Okada *et al.* recently reported that those calcifications located in the wall or the septum tend to be more frequent in benign ovarian tumors of serous type, while those located in the necrotic material within cystic structures (psammoma-body like calcifications) tend to be more frequent in malignant or borderline mucinous tumors [8]. Silva *et al.*, who also assessed the location and incidence of calcifications in ovarian neoplasms (including adenofibromas, serous borderline tumors and carcinomas), reported that the incidence of calcifications was in all cases higher in the stroma than in the epithelium [5]. In our case, calcifications were found predominantly in the stroma but also within the epithelium. The resemblance of the latter with psammoma bodies raised suspicion of a malignancy. However, thorough examination of the entire specimen failed to reveal any multilayering, atypia, necrosis or mitotic activity.

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