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Virilizing Ovarian Stromal Tumor in a Young Woman With Carney Complex

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Abstract

A woman with Carney complex presented at age 22 years with abdominal pain and hirsutism. As a baby, she had excision of a right eyelid lesion and at age 14 years removal of a left lower eyelid nodule that subsequently recurred. Investigation revealed an elevated level of serum testosterone and a 2-cm left ovarian tumor. A left salpingo-oophorectomy was performed. Postoperatively, the abdominal pain was relieved, the serum level of testosterone was normalized, and the hirsutism was ameliorated. The tumor featured sheets of eosinophilic cells with lipochrome pigment, myeloid metaplasia, stromal metaplasia, and markedly abnormal blood vessels. Immunocytochemically, the tumor cells were positive for vimentin, synaptophysin, inhibin A and calrenin. Because of the clinical setting in which the neoplasm occurred, it is unlikely that the occurrence was unrelated to Carney complex..

Keywords

Carney complex; ovarian tumor; virilizing tumor; myeloid metaplasia; *PRKARIA* gene

Introduction

Carney complex (CNC) (3, 20) comprises spotty skin pigmentation, myxomas, endocrine overactivity, and other conditions. The disorder is inherited as an autosomal dominant trait (5, 10) and is caused by mutation of the *PRKARIA* gene in two-thirds of the cases. The endocrine lesions include primary pigmented nodular adrenocortical disease, which causes Cushing syndrome; growth hormone--producing pituitary adenomas, which causes acromegaly and gigantism; and testicular tumors, which may cause gynecomastia (3, 20).

The testicular neoplasms, including large-cell calcifying Sertoli cell tumor, Leydig cell tumor, and adrenocortical rest tumor, are of stromal origin and affect most of the male patients who have been screened by ultrasonography (20). Tumors of ovarian stromal origin have not been described in CNC. The surface epithelial tumor (cystadenoma) and the germ cell tumor (benign cystic teratoma) described in 2 of the 24 women in the original description CNC (3) were thought to be coincidental events. In a subsequent study (21),

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ovarian cysts were found to be common in patients with CNC and it was suggested that CNC might also predispose to ovarian carcinoma.

Another genetic condition, the Peutz-Jeghers syndrome, has been confused with CNC because of the similarity of the facial, labial and oral cavity pigmentation in both conditions. Peutz-Jeghers syndrome includes ovarian stromal tumors (gonadoblastomas (16) and testicular stromal tumors (large-cell calcifying Sertoli cell tumors and distinctive Sertoli cell tumors) (23, 25). These findings raised suspicion that an ovarian stromal tumor might eventually be found in a woman with CNC.

Herein, we describe such a tumor, an ovarian hilus testosterone-secreting stromal tumor in a 22-year-old woman with familial CNC. Because of the rarity of the tumor and the special clinical circumstances in which it occurred, it is unlikely that the lesion was unrelated to CNC.

Case Report

The patient was born in September 1978. She had a right upper eyelid lesion excised as a baby. Menarche occurred at approximately age 10 years. Periods were regular, occurring at 21-day intervals and lasting 4 to 5 days. At age 14 years, she had a nodule excised from the left lower eyelid. This nodule recurred and was treated by laser. No further information about these lesions is available, but they occurred in locations typical for the cutaneous myxomas in CNC (4). At age 22 years, left lower abdominal pain developed, menstruation became irregular and short, and facial hirsutism including a mustache necessitated shaving.

The family history revealed CNC in the patient's mother (22), the patient's brother, a maternal uncle, and a maternal aunt (11). Genetic testing showed that the patient did not have a mutation of the *PRKARIA* gene.

The patient was 157 cm tall and weighed 70 kg. On examination, she had facial hirsutism (including a mustache), a male-pattern escutcheon, and clitoromegaly. Blood pressure was normal. There was a pigmented spot at the left vaginal introitus and a similar spot on the conjunctiva of the left lower eye near the medial canthus—both locations are typical for spotty pigmentation in CNC (3). An echocardiogram was normal. The pelvic examination was normal except as noted above.

Abdominal and pelvic computed tomography showed normal adrenal glands and a left ovarian mass. Transvaginal ultrasonography revealed a hyperechoic, exophytic mass, 2 cm in diameter, with bright flecks (suggesting the presence of foci of fat) at the inferior aspect of the left ovary. The serum testosterone level was 277 ng/dL (reference range, 18-73 ng/dL), and the serum androstenedione level was 630 ng/dL (reference range, 85-275 ng/dL). The clinical impression was virilization caused by a testosterone-secreting left ovarian tumor.

At surgery, the left ovary was double the normal size and adherent to the left pelvic sidewall and the sigmoid colon. Left salpingo-oophorectomy was performed. The tumor was initially interpreted as a teratoma with immature elements. Postoperatively, the abdominal pain was relieved, the serum level of testosterone returned to normal, and menses became more regular, but the hirsutism persisted. The patient subsequently had 3 normal pregnancies.

At age 28 years, pain and tenderness developed in both breasts; the discomfort was worse in the right breast and was worse at menstruation. Physical examination revealed a mass behind the right nipple. The mass was excised. It was composed of 2 myxoid fibroadenomas (1.5 cm and 0.5 cm in diameter) respectively with multifocal intralobular myxoid change in

the extratumoral breast parenchyma. These findings are characteristic of CNC (6). At age 33 years, the patient is well but still has some hirsutism. The bilateral breast pain and tenderness persist.

Materials And Methods

Material for the study included 1) the pathology report of the salpingo-oophorectomy, 2) 4 hematoxylin--eosin (H&E)-stained slides, and 3) 4 formalin- fixed paraffin-embedded blocks.

The following histochemical stains were used: H&E, Masson trichrome, periodic acid-Schiff (PAS), reticulin, Mallory phosphotungstic acid--hematoxylin (PTAH), sulfate alcian blue, Gomori iron, Verhoeff--elastic van Gieson, Congo red, and Fontana-Masson.

Immunostaining was performed with antibodies directed to the following: vimentin (Dual Env; PT link: 1/500 BRD; Dako; V9), synaptophysin (Ventana; CC1 mild; 1/50 BRD; Leica [Novocastro]); 27G12), inhibin-A (Advance; PT link: 1/60 BRD; AbD Serotec; R1), melan A (Advance; PT link: 1/500 BRD; Dako; A103), CD56 (Ventana; CC1 mild; 1/100 BRD; Dako; 123C3), calretinin (Ventana; CC1 mild; 1/50 BRD; Leica [Novocastro; %A%]), S100 (Ventana; CC1 mild; 1/4000; Dako; polyclonal), AE1/AE3 (Ventana; CC1 mild; 1/100; Invitrogen; AE1 and AE3), EMA (Dako; none; 1/50; Dako E29), and MIB1 Ki-67 (Ventana; CC1 mild; 1/20BRD; Dako; MIB-1). A polymer--based detection system was used.

Additional Investigation

Formalin-fixed, paraffin-embedded, H&E-stained sections of a group of ovarian, adrenal and testicular tumors were examined for myeloid metaplasia, hyalinized blood vessels with fibrinoid material, and stromal metaplasia (coarse, branching, largely hyalinized and acellular fibrous bands) similar to that in the tumor being reported.

Results

Gross Pathology

The ovarian tumor was 2.5 cm in diameter, dark brown, and rubbery with a peripheral rim of gray tissue 0.2 cm thick.

Light Microscopy

The tumor was solid and circumscribed and was partly in direct contact with the ovarian stroma, partly separated from it by a fibrous pseudocapsule, and partly in direct contact with the ovarian hilus where it surrounded large blood vessels and groups of fat cells.

Composed of a patternless sheet of epithelioid cells, the lesion featured areas of myeloid metaplasia, stromal metaplasia, and abnormal vasculature. Reticulin staining showed that the cells were arranged in small clusters, and individual cells were surrounded by reticulin. There was no degeneration or necrosis. The extratumoral ovary was normal, including its blood vessels.

The cells were regular, poorly outlined, and arranged in a patternless sheet (Figure 1). Their size varied considerably, from large to huge with correspondingly sized nuclei. The cytoplasm was eosinophilic and very finely granular (almost glassy), and it often contained randomly dispersed punctate basophilic material that was sometimes peripherally located. Many of the cells contained fine to very fine, PAS-positive, granular brown pigment that did not stain for iron or melanin and had an appearance consistent with lipochrome. The

pigment granules were sometimes situated peripherally in the cytoplasm, leaving a paranuclear or central zone of cytoplasm clear of pigment. Very occasionally the cytoplasm was finely vacuolated. Reinke crystals were not seen despite a careful search.

The nuclei were round, vesicular, normochromatic, and regular, each with a single nucleolus (Figure 1). The nuclear size corresponded to the size of the cells--consequently some nuclei were very large. The nuclear to cytoplasmic ratio was high. The nuclei were located centrally in the cytoplasm, less frequently, eccentrically. Some very large cells appeared anucleated. Many nuclei had 1 or more vacuoles with homogeneous or granular eosinophilic content without pigment granules. Nuclear grooving was very rare. Hyperchromatism was evident in 30% to 40% of the nuclei which were presumed to be undergoing degeneration. Mitotic figures were not seen.

Irregularly shaped zones of myeloid metaplasia (composed of hematopoietic cells, megakaryocytes, and fat) occupied about 25% of the area of the tumor (Figure 1). At its periphery, the metaplasia commonly extended among individual tumor cells.

Occasional small normal arteries and veins were present in the tumor but they were not conspicuous. Abnormal vessels, however, were very conspicuous. They were large, with greatly thickened hypocellular to acellular walls, and many lacked obvious media. Some lumens were obliterated. In some instances, the thickening was due to subendothelial accumulation of amorphous, nonfibrillar (rarely fibrillar), eosinophilic and amphophilic material that stained deep blue with PTAH, sky blue with sulfated alcian blue, and dull green with Verhoeff--van Gieson, and did not stain with Congo red (Figure 2). In other vessels, the intima and inner media were intact, and acellular fibrous stroma with dot-like basophilic particles (nuclear debris, presumably) was present external to them. Dystrophic calcification was common; rarely, it was coarsely laminated and, exceptionally, occurred as fine guttate droplets. Large, foamy cells (histiocytes presumably) were occasionally present in the vessel wall. Rare vessels had a lateral, very hypocellular, hemispherical, aneurysmal-like protrusion displacing the adjacent tumor cells. A large vein in the mesovarium adjacent to the ovarian hilus showed marked intimal hypocellular thickening and focal calcification. Extratumoral vessels in the ovarian theca were normal, including thin-walled dilated sinusoids.

Coarse, brightly eosinophilic (tinctorial quality of erythrocytes), branching and anastomosing, largely hyalinized and acellular fibrous bands with focal calcification were scattered in the tumor (Figure 3). The few nuclei present were thin and wavy. The bands showed prominent "cracking" due to tissue shrinkage. They projected among tumor cells and surrounded groups of them and individual tumor cells. There was a 7-mm-long band of stromal metaplasia along the periphery of the tumor. This focally calcified band contained eosinophilic tumor cells and compressed spindle cells (the latter probably entrapped normal theca cells). The stromal metaplasia occupied approximately 5% of the area of the sections.

The extratumoral ovary, including the blood vessels, was normal histologically.

Immunocytochemical results are shown in Figure 4 and detailed in Table 1.

Additional Investigation

The findings from the additional investigation are presented in Table 2.

Discussion

We describe the case of a 22-year-old woman who had CNC, abdominal pain, and virilization. Investigation revealed elevated serum levels of testosterone and a left ovarian tumor. Resection of the tumor cured the abdominal pain, normalized the serum level of testosterone, and ameliorated the hirsutism.

Gonadal stromal tumors are common in male patients with CNC (3, 20). A few affected women have had surface-derived epithelial tumors (21), but none, to our knowledge, has had an ovarian stromal tumor. While it is possible that the stromal tumor we describe was a random occurrence, this is unlikely for the following reasons: the tumor was found in a patient with a rare syndrome and the tumor was unusual histologically, featuring myeloid metaplasia, abnormal blood vessels with fibrinoid and hyalinization, and stromal metaplasia. We did not encounter these findings in a series of steroid cell tumors (Table 3).

The argument for its syndromic nature would be stronger if the tumor we report had been multifocal in the left ovary or bilateral (or both). This deficit notwithstanding, we think that circumstantial evidence favors its association with CNC. Another rare component of CNC, osteochondromyxoma of bone, has thus far occurred only as a single lesion (2).

Myeloid metaplasia, a common finding in cortisol-secreting adrenocortical adenomas (12), has not been reported in ovarian steroid-producing tumors (9). In testicular Leydig cell tumors steroid-producing tumor cells have transformed into fat cells (24), but we did not see this phenomenon in the tumor we report.

Scully and colleagues (9) described fibrinoid replacement of the walls of moderate-sized vessels in ovarian Leydig cell tumors but did not mention vascular hyalinization and calcification. In previous studies, we did not find these vascular changes in nongonadal lesions in CNC (eg myxomas, primary pigmented nodular adrenocortical disease or psammomatous melanotic schwannoma). Therefore, they are unlikely to be related to CNC per se but to a peculiarity of the tumor itself. A somewhat similar vascular abnormality is present in pleomorphic hyalinizing angiectatic tumor (8, 19) and schwannoma (15), but in both of these tumors the vessels are ectatic.

Stromal metaplasia has not been mentioned in ovarian steroid-producing tumors, to our knowledge. Our finding resembled so-called ovarian thecal metaplasia found occasionally in the normal adrenal cortex of elderly women (1, 7, 14). Transformation of steroid cells in testicular Leydig cell tumors into spindle cells does occur rarely (24), but we did not observe this.

In summary, we describe a testosterone-secreting ovarian steroid-cell tumor in a young woman with CNC—a lesion that was anticipated. The tumor featured myeloid metaplasia, vascular abnormality, and stromal metaplasia. To our knowledge, the totality of the findings make the tumor unique.

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Figure 1.

Hilar Cell Tumor. A, Sheet of medium-sized and large tumor cells with nuclei of corresponding sizes. Some of the cells appear anucleate. A zone of myeloid metaplasia (bottom right) featured adipose tissue and hematopoietic cells. B, Reticulin surrounded clusters of tumor cells (upper left, upper right, and lower left) and individual cells (center). Reticulin was very abundant among small tumor cells at the interface with the zone of myeloid metaplasia (lower right). C, The patternless sheet of poorly outlined tumor cells with eosinophilic cytoplasm was traversed by a series of capillaries. The nuclei were regular and vesicular, and each had a well-defined nucleolus. There was dusty brown pigment in the cytoplasm of many of the cells. D, The size of the nuclei ranged from small to very large and the nucleoli were correspondingly sized. Periodic-acid Schiff stained the fine and coarse cytoplasmic lipochrome granules.

Figure 2. Hilar Cell Tumor. A, This area of tumor included a zone of stromal metaplasia (upper left) and a thick-walled blood vessel (lower right). B, The area of stromal metaplasia and the blood vessel in A are highlighted with PTAH staining.

Figure 3.

Hilar cell tumor. A, Two vessels (upper left and bottom) were thick - walled and featured homogeneous eosinophilic intramural material (fibrinoid) and clear cells in the upper left. The lumen in the lower vessels is almost obliterated. The vessel in the upper right is virtually replaced by fibrinoid material. The lymphocytic infiltrate among the tumor cells is likely a manifestation of myeloid metaplasia. B, A transversely cut vessel is shown (center) with a portion of the walls of 4 other vessels with fibrous replacement of their walls. The lumen of the transversely cut vessel was obliterated and showed intramural aggregates of laminated calcification in its wall (not visible at this magnification). C, Two vessels showed heavy deposition of fibrinoid material (black) (PTAH stain).

Fig. 4. Hilar Cell Tumor. Immunostaining showed positivity of various numbers of tumor cells with vimentin (A), synaptophysin (B), inhibin-A (C), melan A (D), calreninin (E), and CD56 (F). Most of the tumor cells stained with inhibin-A. The fewest stained with CD56.

Table 1

Results of Immunocytochemical Staining of Virilizing Ovarian Tumor Cells, Tumor Stromal Metaplasia, and Normal Ovarian Stroma

Immunostain	Epithelioid Cells	Stromal Metaplasia	Normal Ovarian Stroma
Vimentin	Positive, patchy; fine granularity; Smaller cells stained more heavily	Positive, minimal	Positive, strong
Synaptophysin	Positive, patchy; membrane and cytoplasmic body	Negative	Negative
Inhibin-A	Positive, strong; fine to medium diffuse granularity	Negative	Positive, fine granularity
Melan A	Positive, strong; fine to medium diffuse granularity	Negative	Negative
Calretinin	Positive; scattered cells stained strongly, remainder stained weakly with a more heavily stained periphery	Negative	Negative
CD56	Negative ^a	Negative	Positive, weak
AE1/AE3	Negative	Negative	Negative
EMA	Positive, weak; very fine granularity; focal	Negative	Positive, patchy
SI00	Positive, patchy	Negative	Negative ^a
Ki67	Rare nucleus stained	Negative	Negative

^aSmall nerves stained positively

Table 2
Selected Clinical and Pathological Features of Certain Steroid Cell Tumors

Syndrome	Tumor Site	Tumor Function	Cell Type	Tumor, No.	Myeloid Metaplasia	Hyalinized, Thick-Walled Vessels With Fibrinoid	Stromal Metaplasia
None	Ovary	Virilizing	Steroid	13	0 ^a	0	0
None	Ovary	None	Steroid	12	0	0	0
CNC	Testis	None	Steroid ^b	1	0	0	0
None	Testis	None	Steroid ^b	26	0	0	0
None	Adrenal	Virilizing	Steroid	6	1	0	0

Abbreviation: CNC, Carney complex

^aTwo had fatty metaplasia

^bLeydig cell tumor

Table 3
Comparison of Reported Histologic Features in Selected Steroid Cell Ovarian Tumors With Those in the Present Case

Tumor Type (Reference)	Tumor	Histological Features				Nuclear Pseudo- inclusions
	Function	Location	Myeloid Metaplasia	Fibrinoid Vascular Degeneration	Stromal Metaplasia	
Hilus cell (13)	Virilizing	Hilus	— ^a	+	—	+
Leydig cell (17)	Virilizing	Intraovarian	—	+	—	+
Steroid cell, NOS (9)	Virilizing or Cushing syndrome	Intraovarian	—	—	+?	—
Present tumor	Virilizing	Hilus	+	+	+	+

Abbreviations: NOS, not otherwise specified

^aMinus sign indicates absent ; plus sign, present.