

Primary trabecular carcinoid in the ovarian dermoid

Surang Triratanachat*
Niphon Praditphol** Vira Kasantikul*

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A case of primary trabecular carcinoid arising within an ovarian dermoid is described in a 40-year-old woman who was presented with a pelvic mass. An ultrasonogram disclosed a large cystic ovarian tumor. At laparotomy a 30x20 cm. cystic mass on the right ovary was observed. It contained hairs, sebaceous gland tissue, and sebrum. Additionally, ribbons of neoplastic epithelial cells were noted. The nature of the tumor cells was verified by immunohistochemical and electron microscopic studies.

Key words : Carcinoid, Ovarian dermoid.

Reprintrequest: Kasantikul V, Department of Pathology, Faculty of Medicine, Chulalongkorn University, Bangkok10330, Thailand.

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ผู้ป่วยหญิงไทยอายุ 40 ปี มีก้อนในอุ้งเชิงกรานด้านขวา อัลตราซาวด์พบถุงน้ำขนาดใหญ่ที่
รังไข่ด้านขวาจากการผ่าตัดพบก้อนเนื้องอกของรังไข่ขนาด 30x20 ซม. ภายในประกอบด้วยเส้นผม,
ต่อมไขมันและเซลล์เนื้องอกที่เรียงตัวเป็นแถบ เซลล์เนื้องอกดังกล่าวพบว่าเป็นคาร์ซินอยด์
โดยกรรมวิธีอิมมิวโนฮิสโตเคมี และการตรวจด้วยกล้องจุลทรรศน์อิเล็กตรอน

Carcinoids may occur in various organs such as the lung, gastrointestinal tract, testis, and in the ovary.⁽¹⁾ Carcinoids in the latter structure, however, are uncommon and account for less than 1 per cent of all carcinoids.⁽²⁾ Most ovarian carcinoids are associated with ovarian dermoids either as two separated lesions or they may arise within a cystic teratoma.^(2,3) Histologically, primary ovarian carcinoids have been classified into two main types. Most are of the insular pattern while less common is the trabecular form. Occasionally, this tumor may display other unusual features such as mucin production, spindle cell type, or in combination with thyroid tissue.⁽⁴⁻⁶⁾ We encountered a case of trabecular carcinoid which arose as part of a benign ovarian dermoid cyst. The nature of the tumor cells was verified by immunohistochemical and electron microscopic findings.

Case Report

A 40-year-old woman, gravida 3, para 3, was admitted to Chulalongkorn Hospital because of a progressively enlarging abdominal mass present for 2 years. Examination revealed a firm mass in the right adnexa. The heart, lungs, and other viscera were unremarkable. Routine laboratory data were within normal limits. An abdominal sonogram disclosed a large ovarian mass with areas of hyperechogenicity. Laparotomy disclosed a 30×20 cm cystic ovarian mass on the right side. A total hysterectomy with bilateral salphingo-oophorectomy was performed. The postoperative course was uneventful. The patient was sent home 5 weeks after surgery.

The cut surface of the tumor showed trilobulated cysts containing dark red fluid, sebum, and hairs. A small solid area projected from the cyst wall into the lumen showing a yellow soft cut surface. The uterus, fallopian tubes, and left ovary were unremarkable. The surgical specimens were fixed in 10 per cent formalin, embedded in paraffin and stained with hematoxylin and eosin (H & E), and Mayer's mucicarmine. Sections of the paraffin embedded tissue were further processed by the peroxidase-antiperoxidase (PAP) indirect immunohistochemical method using unlabelled antibodies to S-100 protein, neuronal specific enolase (NSE), and chromogranin. Moreover, a portion of solid mass was washed, fixed in buffered 2.5 per cent glutaraldehyde solution, embedded in epoxy resin and prepared for electron microscopy (JEOL 1210).

Microscopically, part of the cyst was lined by stratified squamous epithelium and contained abundant sebaceous glands, hairs, and keratin (Fig 1A). The solid mass was composed of long wavy ribbons of epithelial cells widely separated by a dense fibrous connective tissue. The ribbons were usually one to two cells thick, with the long axes of the cells parallel to one another (Fig 1B). The nuclei were oblong and located basally. The tumor cells had abundant granular eosinophilic cytoplasm. Mitoses were rarely observed. They were negative for mucicarmine. These cells showed reactivity to chromogranin and NSE (Fig 2A,B) but were negative to S-100 protein. Under electron microscopy, two types of cells were present, light and dark cells. The light cells contained many small round dense core granules (Fig 3A) the nuclei were oval. The dark cells often showed deep invagination of the nuclear membrane and contained lesser amounts of similar granules (Fig 3B).

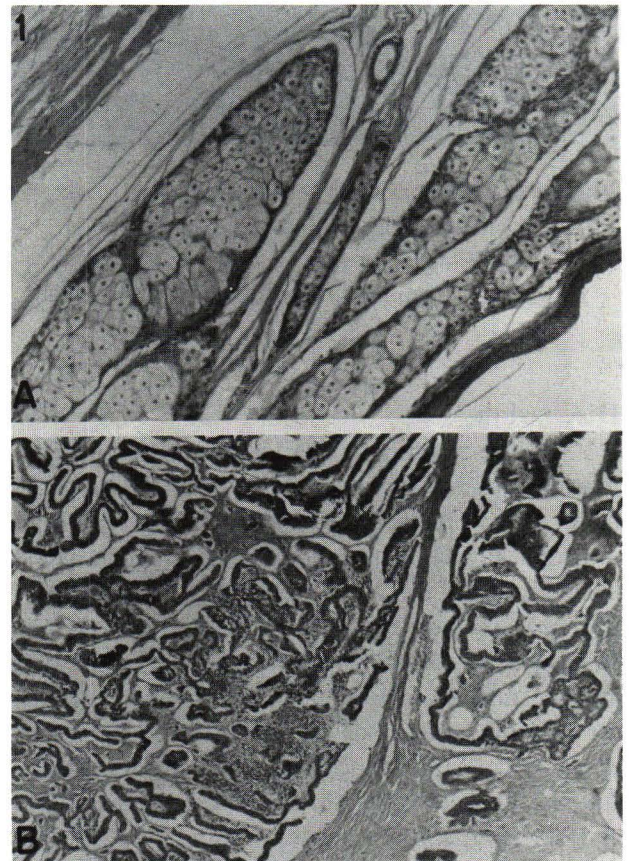


Figure 1. Microscopic findings of the mass.

- A. Abundant sebaceous gland and the squamous lining of the cyst. (H&E×200)
- B. Long wavy ribbons of neoplastic epithelial cells widely separated by dense fibrous connective tissue. (H&E×100)

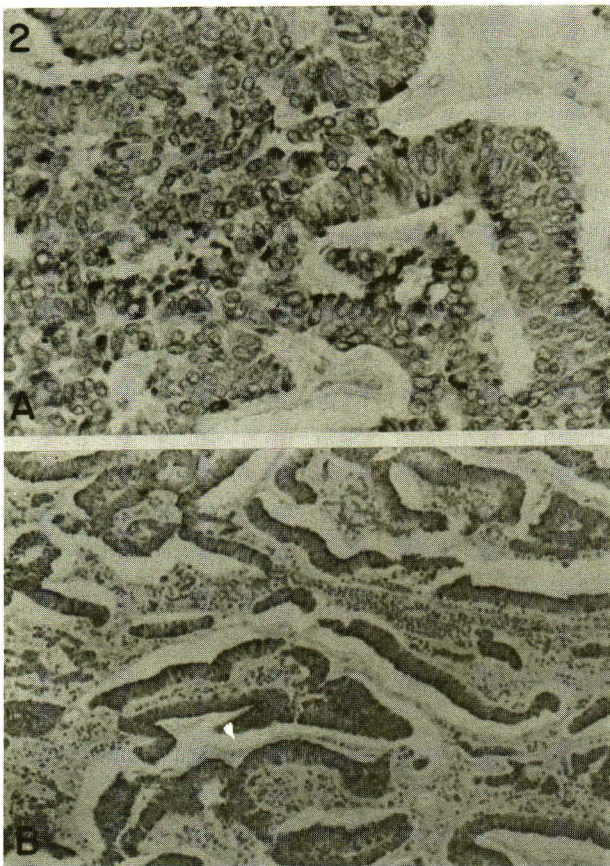


Figure 2. Immunohistochemical stains.

- A. Positive reactivity to chromogranin.
(PAP×400)
B. Positive immunostain for NSE. (PAP×200)

Discussion

The findings of ovarian cyst with sebaceous glands, hairs, and keratin content provided our diagnosis of ovarian dermoid cyst. However, ribbons of neoplastic cuboidal and columnar cells could have been mistaken for a Sertoli-Leydig cell tumor or adenocarcinoma. The positivity to chromogranin, NSE, lack of crystal of Reinke in the stromal cells and the presence of abundant dense-core granules are evidence against such possibilities. Furthermore, the histologic appearance and immunohistochemical findings as well as the ultrastructural features, were entirely compatible with descriptions of trabecular carcinoid.^(3,7) Thus we considered the ovarian tumor as being a trabecular carcinoid arising within an ovarian dermoid. The correct diagnosis of the former lesion can only be established with the help of electron microscope findings as seen in our tumor. Furthermore, a unilateral carcinoid tumor of the ovary without evidence of extraovarian spread and

the presence of teratomatous component indicate its ovarian origin while bilateral ovarian tumors with peritoneal involvement are evidence of metastases to the ovary.⁽³⁾

Clinically, trabecular carcinoid may occur in patients at any age group. In a series of 18 cases reported by Robboy et al, the patients' age ranged from 24 to 74 years with an average age of 45.3 years.⁽³⁾ Most patients are presented with abdominal pain and a pelvic mass. In contrast, insular carcinoids larger than 4 cm can give rise to the carcinoid syndrome in about one-third to one-half of cases and this syndrome may be present for many years before the tumor diagnosis.⁽²⁾ Such different clinical manifestations are perhaps related to the morphologic aspects of the secretory granules. Most insular carcinoids contain strong reducing substances resulting in positive reactions to Fontana-Masson, diazo, and ferric ferricyanide methods while trabecular tumors contain only a weak reducing substance for ferric ferricyanide, and were negative with Fontana-Masson and diazo stains.^(2,3) Moreover, under the electron microscope the insular carcinoids are seen to contain irregular, pleomorphic dense core granules whereas the trabecular tumors possess smaller, round and uniform granules as was observed in our case.⁽⁷⁾ Additionally, the insular pattern which is midgut derivation often produces a large amount of 5-hydroxytryptophan (5 HT).⁽⁹⁾ The trabecular carcinoid, particularly of the hindgut derivative, produces no excess of 5 HT.

Regardless of the clinical differences, primary ovarian trabecular carcinoids are usually benign and the prognosis is generally favorable after removal of the tumors. According to Robboy et al, 17 of their 18 patients with trabecular carcinoids survived up to 15 years postoperatively or died due to unrelated causes.⁽³⁾ Hence, unilateral oophorectomy is recommended in young women when the opposite ovary and the intestinal tract are free of tumors.⁽³⁾ Bilateral salphingo-oophorectomy with a hysterectomy is preferable in older patients. Chemotherapy may be useful as an adjunct to surgery in cases with evidence of tumor recurrence or metastases.

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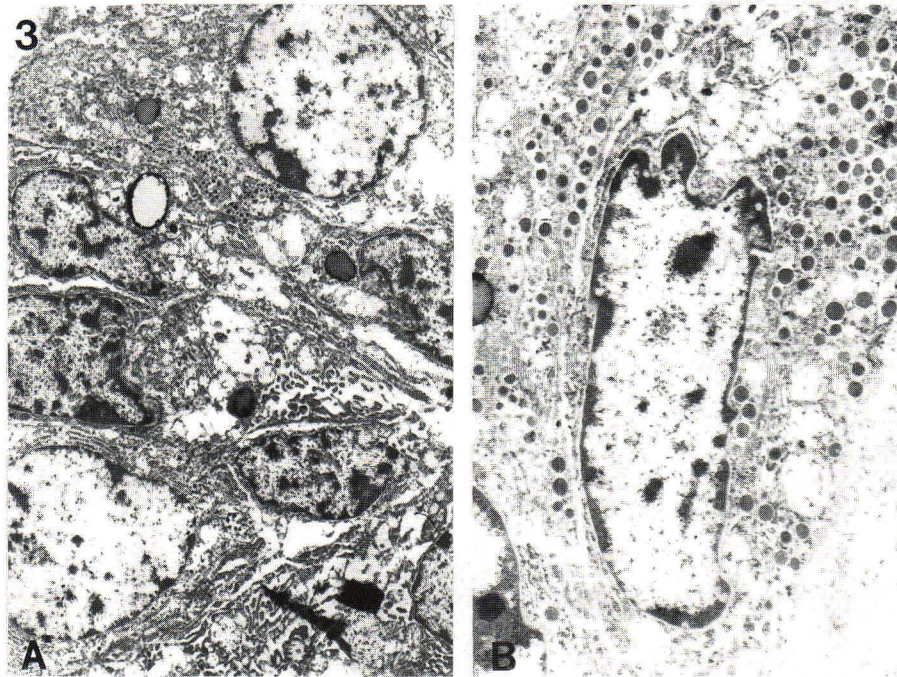


Figure 3. Electron micrographs of trabecular carcinoid.

- A. The “light” cells with oval or round nuclei and the “dark” cells with invaginated nuclei are demonstrated. ($\times 3750$)
- B. Small round dense-core granules in the cytoplasm of “light” cells. ($\times 9,000$)

References

1. Godwin JD 2d. Carcinoid tumors. An analysis of 2837 cases. *Cancer* 1975 Aug; 36(2): 560-9
2. Robboy SJ, Norris HJ, Scully RE. Insular carcinoid primary in the ovary. A clinicopathologic analysis of 48 cases. *Cancer* 1975 Aug; 36(2): 404-18
3. Robboy SJ, Scully RE, Norris HJ. Primary trabecular carcinoid of the ovary. *Obstet Gynecol* 1977 Feb; 49(2): 202-7
4. Robboy SJ. Insular carcinoid of ovary associated with malignant mucinous tumors. *Cancer* 1984 Nov; 54(10): 2273-6
5. Stangno PA, Petras RE, Hart WR. Strumal carcinoids of the ovary. An immunohistologic and ultrastructural study. *Arch Pathol Lab Med* 1987 May; 111(5): 440-6
6. Alenghat E, Okagaki T, Talerman A. Primary mucinous carcinoid tumor of ovary. *Cancer* 1986 Aug; 58(3): 777-83
7. Czernobilsky B, Segal M, Dgani R. Primary ovarian carcinoid with marked heterogeneity of microscopic features. *Cancer* 1984 Aug; 54(3): 585-9
8. Williams ED, Sandler M. The classification of carcinoid tumours. *Lancet* 1963 Feb 2; 1 (7275): 238-9
9. Hilton P, Tweddell A, Wright A. Primary insular argentaffin carcinoma of ovary. Case report and literature review. *Br J Obstet Gynecol* 1988 Dec; 95(12): 1324-31