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"RARE CASE OF INCIDENTALLY DIAGNOSED PRIMARY INSULAR OVARIAN CARCINOID"

KEYWORDS: Postmenopausal bleeding, ovarian carcinoids, insular ovarian carcinoid.

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ABSTRACT **INTRODUCTION:** *Carcinoids account to approximately 0.1% of the ovarian neoplasms,and among carcinoid clan they are found in 1% of cases. Primary ovarian carcinoids are most asymptomatic (40-60%), unilateral (stage I disease).* **CASE REPORT***: 59yr old, with 8yrs of menopause, came with complaints postmenopausal bleeding. On evaluation USG abdomen showed Uterus measuring 10.4X6cm, ET=8mm with multiple fibroids, right ovarian cyst measuring 2.8X3cm.D&C showed features of simple endometrial hyperplasia.CA125 was normal. She underwent Total Abdominal Hysterectomy and Bilateral Salphingoophorectomy. Histopathology report: Simple hyperplasia of endometrium, Focal adenomyosis and fibroid uterus, Right ovary revealed features suggestive of ?Juvenile Granulosa Cell tumor/ ?Neuroendocrine tumor of ovary. Immunohistochemistry showed positive for Pan CK, Synaptophysin, Chromogranin and Ki-69 was <1%, confirmed diagnosis of Well differentiated Neuroendocrine tumor of Ovary. Gastrointestinal.* **CONCLUSION:** *Surgical management is the mainstay for the treatment of primary ovarian carcinoid. At present there is no consensus on the role of chemotherapy.*

BACKGROUND

Carcinoid tumors were first described by Lubarsh, in 1888.They are rare, slow-growing neoplasms that arise from the neuro-endocrine cells1.Primary Ovarian Carcinoid tumors are rare ovarian neoplasm. They constitute 0.1% of all ovarian neoplasias and 0.3% of all carcinoid tumors2. They can be usually seen with a dermoid cyst, mucinous cystadenoma, sertoli-leydig cell tumor, or Brenner tumor. Pure forms are rare and have a low-malignant potential.

CASE REPORT:

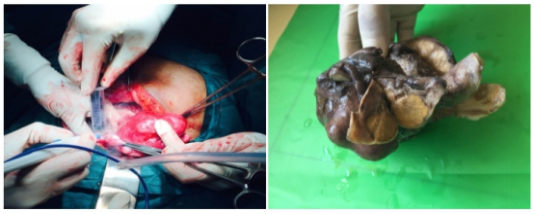
A 59 year old woman, para 3 living 3 , who was menopausal for the last 8years came to gynecology clinic with complaints of one episode of vaginal bleeding for 3days, 3 months prior to consultation. There were no other significant complaints, except for that she is a known asthmatic since childhood. Per-vaginal examination was unremarkable.

Investigations: Pelvic ultrasonography showed multiple fibroid uterus with endometrial thickness of 8mm and a right ovarian cyst of 2.8X 3cm contralateral adnexa was unremarkable. CA 125 was normal.

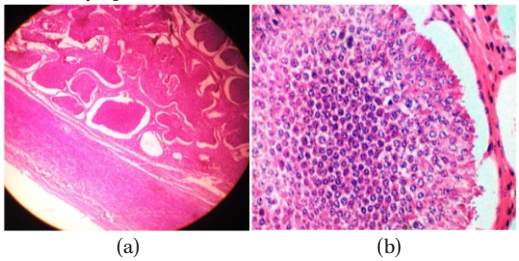
Management: A diagnostic endometrial and endocervical curetting was done and pathology report suggested simple hyperplasia without atypia. Following which she underwent a Total abdominal hysterectomy with bilateral salphingo-oophorectomy in view of endometrial hyperplasia and ovarian cyst.

Intraop findings: Uterus was approximately 10weeks size with multiple subserosal and intramural fibroids. Right ovary was enlarged and cut section was yellow, solid cystic in consistency.

Post-operative period was uneventful.



Histopathology: Revealed Simple hyperplasia of the endometrium, with multiple leiomyomas with areas of hyalinization and calcification. Right ovary showed sheets of tumor cells arranged in insular pattern, solid and focal microfollicles.Individual cells showed mild to moderate cytoplasm with neuroendocrine chromatin pattern and increased atypical mitosis with pleomorphism. No elements of teratoma. No lymphovascular emboli noted.

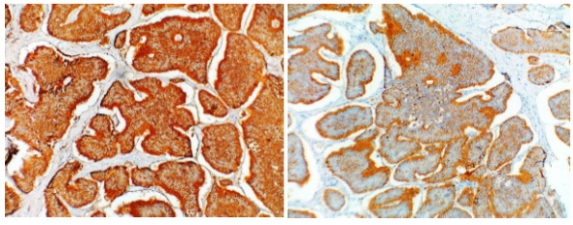


(a) (b)

Figure 1 (a) The microphotographs show a pure ovarian insular carcinoid tumor. They show monotonous tumor cells forming islets and layers (H&Ex10) ; (b)Under greater magnification, monotony, narrow cytoplasm, and salt-pepper type chromatin of the tumor cells are noted (H&Ex40);

Immunohistochemistry: Tumor cells were positive for Chromogranin (+++), Synaptophysin (++), Pan Cytokeratin (+). Ki-67 proliferation index was < 1%.

Under the light of these data the case was considered to be a well differentiated pure insular carcinoid tumor of ovary.



(a) (b)

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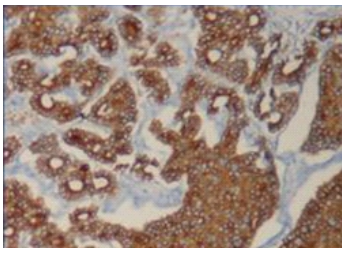

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Figure 2: Immunohistochemical examination tumor cells were diffusely positive for a)strong granular cytoplasmic chromogranin (Chromogranin x20);(b)Cytoplasmic synaptophysin (Synaptophysin x20); c)Pan-cytokeratin(Pan-cytokeratin x10).

Gastrointestinal tract was evaluated by CT/MRI and carcinoids elsewhere were ruled out.

Following the surgery patient is on regular follow-up initially after a month and later is on 3 monthly follow-up.

DISCUSSION:

Upto 90% of carcinoids arise from the appendix and the small bowel, remainder usually originate from the other gastrointestinal sites or the bronchi. Ovary, thymus and breast are very rare sites of origin[4]. Carcinoid tumors have a higher incidence in the 5th and 6th decade of life. The tumor has predilection to female gender[5]. They develop from well-differentiated neuro endocrine cells. Histologically classified as Insular, Trabecular, Strumal, Mucinous and mixed forms. Primary Insular form is the most common form, usually associated with ovarian teratomas6.Carcinoid syndrome is observed in approximately 1/3rd of the cases even in the absence of metastasis. It occurs at a rate of 43% in insular type and 25% of these cases were seen to originate from mature cystic teratomas[5].Although our patient had an insular type of ovarian carcinoid, she had no symptoms of carcinoid syndrome. Since the diagnosis was not suspected clinically the patient was not evaluated for the same initially.

It is crucial to distinguish between primary versus secondary ovarian carcinoid tumors. Carcinoid tumors arising from the gastrointestinal system particularly metastasize to ovaries and they are generally bilateral. In contrast, Primary ovarian carcinoids are unilateral3.The malignancy potential of ovarian primary insular carcinoid tumors should not be overlooked, even though they grow very slowly. They are usually diagnosed at FIGO stage 1. Literature reports have given a survival rate of 100% in the primary form limited to one ovary whereas it drops to 35% at advanced stage.

CONCLUSION:

Ovarian pure carcinoid tumors are rare. Differential diagnosis may pose difficulty for the pathologist in such cases, usually diagnosed in FIGO stage I. They have a low malignant potential, Surgery is usually sufficient mode of treatment and at present there is no consensus on the role of chemotherapy3.

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