

OVARIAN CARCINOID: A RARE EXPERIENCE

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ABSTRACT

Carcinoid tumors are neuroendocrine tumors. They occasionally secrete serotonin and associated factors that result in a unique constellation of features termed carcinoid syndrome. We report a case of primary ovarian carcinoid, which is a rare condition. A 54 year old lady presented at Obstetric Gynecology Department of NMCTH with history of gradually increasing mass in lower abdomen for 3 years. She also had intermittent flushing of face, sweating, shortness of breath, and multiple episodes of loose stools. Examination revealed a mobile mass felt separate from the uterus. Ultrasound, tumor markers and CT scan abdomen were performed. She then underwent staging laparotomy. Histopathology and immunohistochemistry were suggestive of neuroendocrine tumor. After surgery her symptoms disappeared and she was discharged on the 4th postoperative day.

KEYWORDS

Carcinoid, tumors, laparotomy, Nepal

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INTRODUCTION

Carcinoid tumors are neuroendocrine tumors that originate from the derivatives of the embryologic primitive gut.¹ These tumors can be “non-functioning” or “functioning”. Non-functioning tumors present as a mass, whereas functioning types may present with signs and symptoms mediated by the secretion of several biopeptides. This unique constellation of features is known as **carcinoid syndrome**.

These tumors usually arise from the gastrointestinal tract, mainly the appendix,² however they may originate from various other organs as well. Primary carcinoid tumors of the ovary constitute only 0.5% of all carcinoid tumors and 0.1% of all ovarian neoplasms.^{1,2} We report a rare case of a primary ovarian carcinoid tumor (OCT).

CASE REPORT

A 54 year old postmenopausal lady presented to Obstetric and Gynecology Department of NMCTH with complains of mass in lower abdomen for 3 years. The mass had gradually increased in size and was associated with continuous vague type of pain and heaviness. She did not give any history of difficulty in micturition, blood in urine, blood in stools, or postmenopausal vaginal bleeding. However, she had episodes of sudden flushing of face associated with sweating and shortness of breath that ended with passage of multiple loose stools. Shortness of breath was unrelated to physical activity or position. These episodes occurred almost five to six times a day. There was no history of significant weight loss. Her past medical history was not significant. She did not smoke or consume alcohol. She had not had any prior medical consultation nor had she had any over the counter treatment for the same symptoms.



Fig. 1: Uterus with left ovarian mass

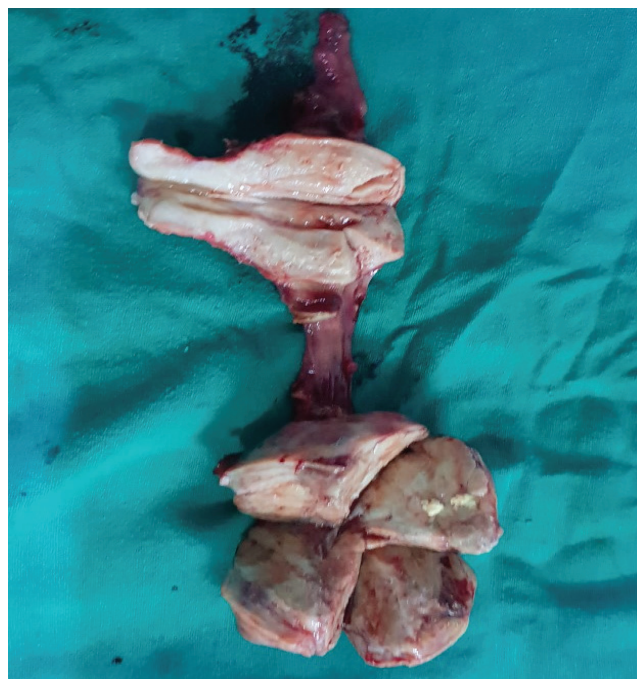


Fig. 2: Cut section of the uterus and ovarian mass

On examination, she had a flushed appearance. Her blood pressure was 120/70mmHg, pulse 80/min, respiratory rate 20/min and temperature 97.2°F. General examination revealed no other abnormality. Examination of the respiratory and cardiovascular systems was also normal. Abdominal examination revealed a soft mass of around 8 x 8 cm in her left iliac fossa that was irregular in outline, mobile, and non-tender. On bimanual examination, a solid mobile mass of around 8 x 8 cm could be felt separate from the uterus.

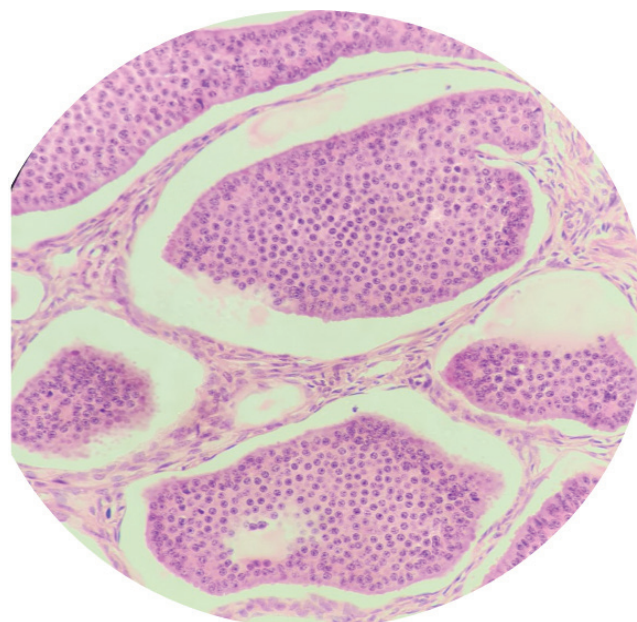


Fig. 3: Histopathology of the ovarian mass

An ultrasound of her abdomen was performed, which showed a complex left adnexal cyst of around 10 x 10cm. She then underwent a computed tomography scan of her abdomen, which showed a well-defined soft tissue density with heterogenous enhancement likely to be an immature teratoma. Her tumor markers (CEA, CA-125) were normal. Due to her symptoms of episodic breathlessness, medical consultation was done and she was advised for pulmonary function test, electrocardiogram and an echocardiogram, which all turned out to be normal.

With the provisional diagnosis of an ovarian teratoma, she underwent staging laparotomy followed by total abdominal hysterectomy and bilateral salpingo-oophorectomy with infracolic omentectomy. Per-operative findings revealed a normal uterus and right adnexa. A smooth glistening left ovary of around 10 x 10 cm with lobulations was seen (Fig. 1). Cut section showed solid yellow to grey white tissue with multiple cystic spaces (Fig. 2).

Her postoperative period was uneventful and her episodic symptoms of breathlessness and flushing disappeared after surgery. She was discharged on 4th postoperative day and had follow-up after histopathological reports.

The histopathology examination (HPE) showed uniform tumors cells in sheets and solid nests with acini within them (Fig. 3). The nuclei were centrally located, and nucleoli were stippled to hyperchromatic. Moderate amount of granular cytoplasm was present. The omental biopsy was normal. Immunohistochemistry report revealed the tumor to be immunoreactive (CK, synaptophysin, CGA and Ki-67 markers) suggesting a neuroendocrine tumor.

DISCUSSION

Carcinoid tumors are slow-growing neoplasms of neuroendocrine origin that are usually benign, but may rarely metastasize. The incidence has been reported to be approximately 0.28/100,000 populations/year and are most commonly seen in the gastrointestinal tract (67.5%), followed by the respiratory system (25.3%).³ Only 0.5% of carcinoids arise in the ovary and it constitutes only 0.1% of all ovarian neoplasms.¹ Thus patients presenting with ovarian tumors are rarely suspected of having a carcinoid. To the best of our knowledge this is the first case to be reported from Nepal.

These tumors mainly occur in postmenopausal women and are invariably unilateral.¹ Forty three percent of OCTs are associated with carcinoid syndrome.⁴ The syndrome consists of

episodic cutaneous flushing, cyanosis, abdominal cramps, diarrhea, carcinoid heart disease, and bronchoconstriction.⁵ These symptoms are due to direct production of serotonin and other similar humoral factors into the systemic circulation through the ovarian venous system bypassing hepatic deactivation.⁴ Our patient also presented with a few constellation of the above symptoms. However these symptoms are non-specific and a high degree of suspicion is required to clinically diagnose carcinoid syndrome.

Commonly performed imaging studies such as ultrasonography, CT scan, and MRI are also not diagnostic. Measurement of urinary 5-HIAA, a serotonin metabolite, or serum chromogranin A, a glycoprotein secreted by the tumor, may be more specific for carcinoid.⁶ The gold standard for locating functional tumor is radiolabelled somatostatin analog scintigraphy.⁷ Unfortunately these specific investigations are not available in Nepal.

A preoperative diagnosis is important as the patient's condition can be optimized to minimize tumor activity. Any stimuli such as anesthesia, surgery or stress can cause unpredictable and uncontrolled release of hormones leading to a devastating condition called carcinoid crisis. It is characterized by hypotension or hypertensive crisis and hemodynamic collapse, which may be unresponsive to conventional drug therapy.⁸ Fortunately, such an event did not occur in our case. Octreotide started at least 12 hours prior to surgery is commonly used to inhibit the action and release of hormones from the tumor.⁹

Due to the difficulty of setting an accurate preoperative diagnosis, most cases are diagnosed based on postoperative pathology findings.¹⁰ Even in our case too, the diagnosis was made by histopathology and further confirmed after immunohistochemistry. OCTs are histologically categorized into four groups: insular, trabecular, stromal and mucinous.¹¹ Insular carcinoid are considered to have low malignant potential and are more commonly associated with carcinoid syndrome; whereas mucinous variety tend to be more aggressive.¹ In our case, HPE report did not provide the histopathological type of OCT, hence its malignant potential could not be determined. However no evidence of metastasis was found.

The definitive therapy of OCTs is surgical resection. Total hysterectomy with bilateral salpingo - oophorectomy and surgical debulking of extra-ovarian spread is usually performed. Premenopausal women may be approached with a fertility-sparing surgery but with careful staging to exclude occult metastases.^{12,13} The role of systemic chemotherapy in patients

with metastatic carcinoid is unclear and is not recommended¹⁴ as no chemotherapy regimen has demonstrated a survival benefit or progression-free survival.

In conclusion, primary OCTs are rare but usually benign tumors with good outcome. It is important

to diagnose functional tumors as carcinoid crisis may be precipitated perioperatively. Though non-specific, the presence of symptoms of carcinoid syndrome should allow a strong clinical suspicion and indicate further testing. Surgical excision of the tumor is usually sufficient.

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