Abstract:
Carcinoid tumours are rare slow growing neoplasms that arise from neuroendocrine cells. Primary ovarian carcinoid tumours are rare, forming 0.3 percent of all carcinoid tumours. Carcinoid heart disease due to ovarian primary is very rare. A rare case of primary ovarian carcinoid tumour with isolated tricuspid regurgitation is presented. The patient had pelvic mass and was operated for clinical suspicion of ovarian tumour. A large solid yellowish tumour of size 12 cm was seen completely replacing the right ovary. Differential diagnosis of granulosa cell tumour and carcinoid tumour were considered histopathologically. Immunohistochemical study of the tumour showed positivity for Chromogranin and synaptophysin. No tumour was detected elsewhere by abdominal Ultrasonography and CT scan confirming the diagnosis of primary ovarian carcinoid tumour. Post operative review of the case details showed isolated tricuspid regurgitation on routine pre operative echocardiograph. The most common cardiac manifestation of carcinoid heart disease is tricuspid insufficiency. It is important to be aware of carcinoid tumour in the pathological diagnosis of ovarian tumours, even in the absence of any clinical indicator of carcinoid syndrome, as it carries a better prognosis and clinical outcome in comparison with most other malignant ovarian tumours.

Keyword: Primary ovarian carcinoid, Insular, granulosa, tricuspid regurgitation.

INTRODUCTION:
Carcinoid tumours are rare slow growing neoplasms that arise from neuroendocrine cells. Carcinoid tumours commonly occur in the gastrointestinal tract and lungs. However, primary ovarian carcinoid tumours are rare; forming 0.3% of all carcinoid tumours. Carcinoid syndrome due to primary ovarian carcinoid is even rarer. Cardiac lesions are present in one half of patients with the carcinoid syndrome. The most common cardiac manifestation is tricuspid insufficiency. We present here a rare case of primary insular type of carcinoid tumour of the ovary with isolated tricuspid regurgitation.
CASE REPORT:
A specimen of total abdominal hysterectomy with bilateral salpingo-oophorectomy of a 52 year old post menopausal woman was received. The patient was presented with pelvic mass and operated for clinical suspicion of ovarian tumour. On gross examination uterus appeared atrophic with normal cervix, left ovary and both side fallopian tubes. A large yellowish tumour of size 12×11×11 cm was seen completely replacing the right ovary, which was predominantly solid with few cystic areas. Microscopically, the tumour was composed of uniform population of polygonal cells with abundant granular eosinophilic cytoplasm and round to oval nuclei with fine chromatin. These are arranged as acini, nest with insular, micro and macro follicular pattern. No teratomatous component was seen either grossly or microscopically.

[Figure 1: Gross photograph of tumor showing cystic and solid cut surface with yellowish appearance]
[Figure 2: Picture showing acinar pattern of tumor cells. (H & E 100X)]

[Figure 3: Picture showing micro & macro follicular pattern (H & E 100X).]
[Figure 4: Picture showing uniform cells with eosinophilic cytoplasm & nuclei with fine stippled chromatin (H & E 400X).]

Initially histopathological differential diagnosis of granulosa cell tumour / carcinoid tumour was considered. With immunohistochemical study, tumour cells showed strong expression of chromogranin and synaptophysin. Final diagnosis of ovarian carcinoid tumour was made. After arriving at the final diagnosis we went through the case details retrospectively. Isolated tricuspid
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regurgitation was incidentally found in routine preoperative echocardiography.

Figure 5:Tumor cells showing positivity for chromogranin with immunohistochemistry (100X)

Figure 6: Echocardiograph picture showing tricuspid regurgitation.

The patient had no symptoms suggestive of carcinoid syndrome like episodic flushing of skin, cramps, nausea, vomiting and diarrhoea. Urinary 5-hydroxyindole acetic acid (5HIAA) level estimation was done post operatively and found to be within normal limits (6mg/day (normal 2-8mg/d)) probably due to the removal of ovarian carcinoid tumour. No tumour was detected elsewhere by abdominal Ultrasonography and CT scan.

To summarise this is a case of unilateral ovarian tumour which was yellowish in colour grossly. The histopathological and immunohistochemical examination confirmed the diagnosis of carcinoid tumour. No other tumour was detected elsewhere. There was isolated tricuspid regurgitation on echocardiography. In correlation with the above findings, we came to a conclusion of primary ovarian carcinoid tumour – insular type. The probable cause for isolated tricuspid regurgitation in this case may be ovarian carcinoid tumour. Preoperative Urinary 5-hydroxyindole acetic acid values are not available due to the lack of clinical suspicion.

DISCUSSION:
Primary ovarian carcinoid was first described in 1939 by Stewart et al. It is a rare tumour, only three of a series of 2837 cases of carcinoid reported by Godwin being of primary ovarian origin. Most arise within a dermoid cyst. Histologically they have been classified into three groups. The most common, as in the case described, is composed of cells in an insular pattern, typical of midgut derived carcinoids. Less common is the trabecular pattern, typical of foregut and hindgut derived carcinoids. In the strumal pattern there is a mixture of trabecular...
carcinoid and thyroid tissue\textsuperscript{6}. Primary ovarian carcinoids are difficult to distinguish from carcinoid secondaries in the ovary, but certain features including unilateral ovarian involvement and the absence of carcinoid elsewhere are suggestive of a primary ovarian origin\textsuperscript{5} as in our case. Despite their distinctive microscopic characteristics, ovarian carcinoids can be confused with several other primary ovarian tumours, especially granulosa cell tumour\textsuperscript{8}. The granulosa cells surrounding the lumen in Call-Exner bodies lack the eosinophilic cytoplasm that separates the nuclei from the lumen of a carcinoid acinus, and they typically have pale, grooved haphazardly oriented nuclei. The insular type is the only one associated with the carcinoid syndrome which is present in about a third of cases.\textsuperscript{4} Carcinoid syndrome is characterized by episodic flushing of the skin, cramps, nausea, vomiting, and diarrhea. Carcinoid syndrome is caused by vasoactive substances secreted by the tumour into the systemic circulation. The key bioactive mediators released into the portal circulation by gut carcinoid tumours are readily metabolized by the liver and do not reach the heart in high concentration. Thus, gastrointestinal carcinoids (with venous drainage via the portal system) do not usually induce carcinoid heart disease, unless there are extensive hepatic metastases that release the relevant mediators directly into the inferior vena cava\textsuperscript{3}. Primary ovarian carcinoids drain directly into inferior vena cava and they can induce carcinoid syndrome in the absence of hepatic metastasis. Carcinoid heart disease is the cardiac manifestation of systemic syndrome caused by carcinoid tumours. The most common cardiac manifestation is tricuspid insufficiency. There are very few cases described in literature of carcinoid heart disease caused by an ovarian tumour. The largest series (17 patients, only two with heart disease) has been reported by Davis et al. In our case isolated tricuspid regurgitation was seen, without any other symptoms of carcinoid syndrome. We conclude that it is important to be aware of this entity in the pathological diagnosis of ovarian tumors, even in the absence of any clinical indicator of carcinoid tumor/syndrome, as it carries a better prognosis and clinical outcome in comparison with most other malignant ovarian tumors.

REFERENCES


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