CASE REPORT

Malignant Struma Ovarii—Management and Follow-up of a Rare Ovarian Tumor: A Case Report

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Abstract

Mature cystic ovarian teratomas (MCT) are the commonest germ cell tumors accounting for about 30–45% of all ovarian neoplasms and 60% of all benign neoplasms arising in the ovary. Only 2% of MCTs undergo malignant transformation. We report a rare case of a 48-year-old woman diagnosed with a primary malignant struma ovarii arising in a dermoid cyst of the ovary. The patient was admitted with the complaint of pelvic pain and a pelvic mass in the lower abdomen on gynecological examination and ultrasonography showed a dermoid cyst of the ovary. Preoperative tumor markers and routine biochemistry were unremarkable. A laparoscopic left-sided salpingo-oophorectomy was done. Histopathology of the tissue showed a malignant struma ovarii and immunohistochemistry (CK-19 levels) similar to the guidelines for primary thyroid gland disease. The patient was subsequently taken up for staging laparotomy with total abdominal hysterectomy with right salpingo-oophorectomy and retroperitoneal pelvic and para-aortic lymphadenectomy and omentectomy. The histopathological report of all staging tissues was normal and surgical staging IA was assigned. Thyroglobulin level was monitored in the postoperative period. She is free of the disease for the last 2 years.

Keywords: Case report, Dermoid cyst, Mature cystic teratoma, Patient care technician, Struma ovarii.

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Introduction

Germ cell tumors are derived from the primordial germ cells of the ovary and account for 20–25% of all benign and malignant ovarian neoplasms of germ cell origin.[1] Mature cystic teratomas (commonly known as "dermoid cysts") are the commonest germ cell tumors and predominantly found in young women of age group 30–40 years. It accounts for about 30–45% of all ovarian neoplasms and around 60% of all benign tumors arising in the ovary with a 0.17–1.4% reported incidence of malignant transformation. Teratomas are composed of well-differentiated derivations from at least two of the three germ cell layers (ectoderm, mesoderm, and endoderm) and about 10% of tumors are bilateral.[2] Most of the patients with dermoid cysts are asymptomatic and it may be discovered accidentally during gynaecologic investigations for other conditions or due to mass effect. When it is discovered during the reproductive age group, cystectomy is indicated. However, if discovered in postmenopausal age, the likelihood of malignant transformation of any of the components is very strong.[3] It is the most common ovarian tumor removed during gynecological surgery.[4,5]

Most mature cystic teratomas are unilocular, and filled with sebaceous material. The cyst wall is lined by squamous epithelium which is thin and compressed, often hyalinized ovarian stroma covers the external surface.[6,7] Ectodermal tissue (skin derivatives and neural tissue) is invariably present. Hair follicles, skin glands, and muscle lie within the wall. There is usually a raised protuberance projecting into the cyst cavity called the Rokitansky protuberance or nodule. Most of the hair arises from this protuberance. Whereas bone or teeth when present, tend to be located within this nodule.[8]

The mesodermal (fat, bone, cartilage, muscle) and endodermal components (gastrointestinal and bronchial epithelium, thyroid tissue) are also seen in the majority of cases.[2] Adipose tissue is present in 67–75% of cases, while teeth are seen in 31%.[2,6,8]

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Struma ovarii is a rare monodermal variant of ovarian teratoma accounting for only 2% of all mature teratomas. To be classified as a struma ovarii, teratoma must be composed of >50% of mature thyroid tissue.[9,10] It is usually a benign condition, but the malignant transformation is observed in about 5% of cases.[11,12] Hyperthyroidism may be present in 5–8% of cases, while thyroid cancer is present in less than 5% of all struma ovarii with metastasis in 5–23% of malignant struma ovarii patients.[13,14]

We report a case of malignant struma ovarii with a focus on papillary thyroid cancer in mature cystic teratoma which was diagnosed at an early stage (stage IA) and was managed appropriately.

Case Description

We present a case of a 48-year-old perimenopausal female, para 4 live 4, presented in the outpatient Department of Obstetrics and Gynaecology, Jawaharlal Nehru Medical College and Hospital with

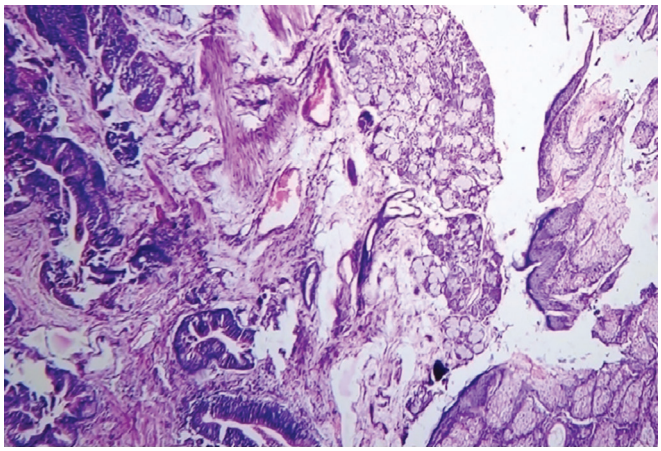

Fig. 1: Dermoid cyst showing skin with its adnexa and many endodermal glands. [H&E × 100]

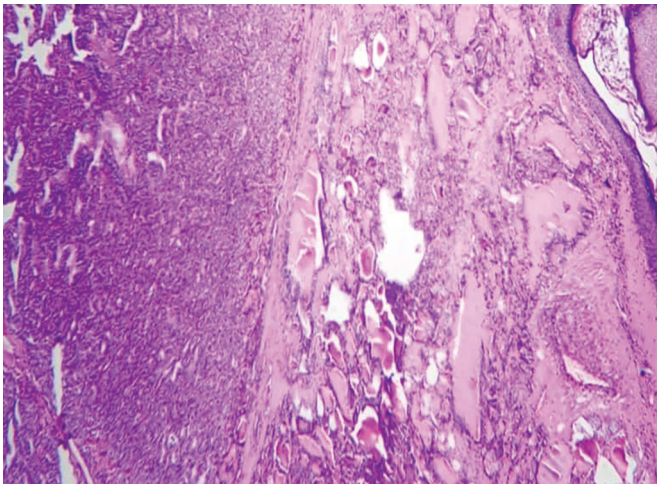

Fig. 2: Showing normal-appearing skin and adnexa of dermoid cyst along with numerous colloid-filled follicles along with solid areas comprising papillary thyroid carcinoma (extreme left). [H&E × 40]

the complaint of pain in the lower abdomen for 3 months. Her past medical history was uneventful and her menstrual history was normal. Her general condition was fair. There was no visible or palpable evidence of any neck swelling, breast abnormality, or lymphadenopathy. Her per abdominal examination showed a midline, large 14 weeks size, cystic, non-tender, well-defined, freely mobile (side to side) mass arising out of the pelvis. Her pelvic examination revealed a mass on the left side with a normal-sized uterus, with loss of intervening space and moving with cervical movement. Per rectal examination ruled out any nodularity in POD. Routine and specific investigations were carried out. USG's whole abdomen demonstrated a large cystic (79 × 72 × 75 mm) mass arising from the left side adnexal region, the lesion was thin-walled and showed mixed echogenicity with the sedimentation of echogenic material and few hyperechoic calcified foci likely ovarian dermoid cyst. CA 125 was 26 U/mL. Given clinical, radiological, and biochemical findings, the patient was taken for laparoscopy.

Her intraoperative finding revealed a left-sided cyst ovarian mass of 72 × 80 mm size. Left-sided salpingo-oophorectomy was performed, cyst was drained in a bag without any peritoneal spillage, whole ovary with cyst along the fallopian tube was taken out from the port, and tissue was sent for HPE (Figs 1 to 3).

Histopathology report showed a mature cystic teratoma with struma ovarii with developing papillary thyroid carcinoma. The immunohistochemistry showed >75% of tumor cells with diffuse CK-19 positivity compatible with patient care technician (PCT). The fallopian tube was histopathologically unremarkable (Fig. 4).

Further management was discussed with the Gynec Oncologist, and she was advised to stage laparotomy with thyroidectomy for confirmation of disease, but she refused thyroidectomy. On staging laparotomy, intraoperative findings were as follows, the right-side tube, ovary, and uterus were normal. There was no fluid in the peritoneal cavity. Peritoneal surfaces, omentum, bowel, liver, and diaphragm all were normal. Peritoneal washings and liver and diaphragm surface biopsies were sent for cytology. A total abdominal hysterectomy with right-sided salpingo-oophorectomy with bilateral pelvic and para-aortic lymphadenectomy with omentectomy was performed. No malignant cells were seen in cytology. The histopathology of the right ovary, tube, uterus, lymph nodes, and omentum was normal, and surgical staging IA was assigned. The postoperative course was uneventful.

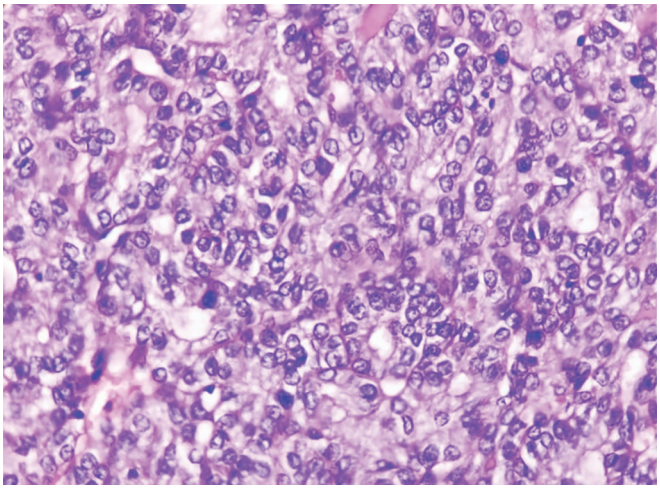

Fig. 3: Papillary thyroid carcinoma: showing solid areas and small follicles lined by cells exhibiting nuclear clearing, grooving, and overcrowding. [H&E × 400]

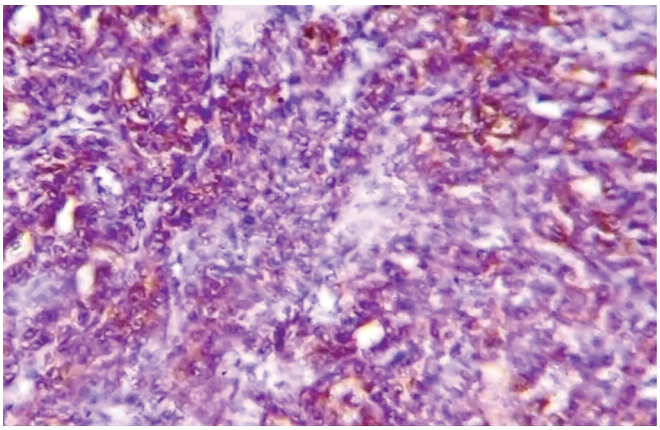

Fig. 4: Cytoplasmic positivity of galectin-3 in the tumor cells of PTC. [IHC × 400]

Thyroid function tests and the thyroglobulin level were also evaluated. Thyroid-stimulating hormone was suppressed and free levels of thyroid hormones and thyroglobulin were in normal ranges. Thyroid ultrasound scanning was also normal. She has been free of the disease for the last 2 years.

Discussion

Mature cystic teratoma (MCT) is most prevalent germ cell tumor in women of reproductive age and responsible for 20% ovarian neoplasm.[15] They are often discovered incidentally on physical or sonographic examination. They may contain hair, teeth, or bone and fatty material. Malignant potential of an MCT is less than 2% of all mature dermoid cysts. Struma ovarii a very rare histological diagnosis is found in just 3% of ovarian teratomas, 2% of all germ cell tumors, and 0.5% of all ovarian tumors.[16,17] Malignant transformation is uncommon, in only about 5% struma ovarii.[16,18]

Malignant transformation starts in the postmenopausal period mostly in the 5th and 6th decade with a median age of 55 years; the common malignant transformation occurs in squamous cell carcinoma, adenocarcinoma, sarcoma, carcinoid, thyroid carcinoma, and melanoma.[19,20] Since the malignant transformation is a rarity and the malignant and benign forms represent a similar appearance, it is challenging to make a preoperative diagnosis. Rim et al.[21] reported that the malignant transformation of dermoid cyst occurred at an average age of 56.8 years, and 63% of the cases were older than 40 years at the time of diagnosis.

The symptoms at presentation are variable in both benign and malignant tumors with some patients presenting with acute abdominal pain and mass effects whereas others with constitutional symptoms such as fatigue, anorexia/weight loss and urinary symptoms.[22] Even though it is a neoplasm consisting of thyroid tissue, only 8% of patients with struma ovarii present with clinical hyperthyroidism.[23] Therefore, there are difficulties in the clinical diagnosis because of the absence of uniform diagnostic criteria due to the rarity of the disease. Various routes of metastasis described are as direct spread to the omentum and peritoneal cavity, the contralateral ovary, regional lymphatics to pelvic and para-aortic lymph nodes, and hematologic dissemination to the bone, lung, liver, and brain.[11,24]

Devaney et al.[25] have advocated that the same guidelines should be followed for pathological diagnosis of malignant struma ovarii as those for thyroid carcinomas. Metastasis is rare in patients with malignant struma ovarii (5%).[25]

There is controversy regarding the optimum treatment of malignant struma ovarii. The treatment alternatives include are radical surgery with or without thyroidectomy followed by adjuvant therapy that can be in form of external radiotherapy, chemotherapy, or thyroid suppression.[26] Various studies suggest that malignant struma ovarii should be treated with combination surgery including total abdominal hysterectomy, bilateral salpingo-oophorectomy, pelvic and para-aortic lymph node sampling and thyroidectomy followed by Iodine-131 (I-131) therapy. Thyroidectomy is a part of radical surgery for two reasons: first to exclude the possibility of primary thyroid carcinoma and second, we can recognize the metastasis, recurrence, and residual tissue after thyroidectomy with total body scanning with I-131 and serum thyroglobulin levels. Serum thyroglobulin is used as a tumor marker for follow-up in these malignant cases. As there have been only a few reported malignant cases, there is no consistent data on the protocol of management of such cases. For the patients desiring fertility, treatment might be challenging. As proposed by Dardik et al.[23] fertility-sparing surgery should be performed first followed by definitive surgery after the completion of childbearing.

The reported case was also advised for surgical staging and thyroidectomy followed by I-131 therapy, but the patient agreed to surgical staging but refused for thyroidectomy. The survival rates of 12–180 months have been reported in the literature in patients of malignant struma ovarii treated without thyroidectomy.[27] As with our case we are unable to determine the metastasis, the residual tissue, or recurrence without thyroidectomy, we think that thyroidectomy should be a part of treatment for better follow-up and prognosis.

Yücesoy et al.[28] have published a similar case report in which following staging laparotomy, the patient refused thyroidectomy, and follow-up was done with serum thyroglobulin levels. Till her follow-up for 18 months, her thyroid profile was normal and the patient was healthy.

Conclusion

Struma ovarii is a monodermal variant of ovarian teratoma, defined by the presence of >50% thyroid tissue that generally occurs in women between the ages of 40 and 60 and usually presents as a unilateral adnexal mass. In view of controversy regarding the management and follow-up of this rare entity, more data and reporting of such type of cases is required for establishing the optimum management protocol. It is also necessary for the gynecologist to be aware of these rare conditions to avoid unnecessary delay in management.

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