CASE REPORT

METASTATIC RADIOIODINE AVID *STRUMA OVARII* ASSOCIATED WITH PSEUDO-MEIGS' SYNDROME

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We report a case of 21 years old lady who presented with ascites, left adnexal mass and elevated CA-125. With suspicion of ovarian malignancy, she underwent left salpingo-oophorectomy with omental biopsy. Histopathology revealed: 'follicular variant of papillary thyroid carcinoma arising in *struma ovarii*' with metastatic papillary thyroid carcinoma in omental and peritoneal nodules. Patient underwent total thyroidectomy followed by radioactive iodine therapy for metastatic omental and peritoneal disease. Post-therapy whole body scan, revealed extensive I-131 avid disease metastatic disease involving the chest, abdomen, pelvis and the musculoskeletal system. Patient was treated with multiple doses of high dose radioactive iodine. She became symptom free on supra-physiologic doses of oral thyroxin however her high thyroglobulin levels and residual radioiodine avid metastatic disease required further treatment. In literature a few cases of *struma ovarii* have been reported with elevated CA-125 and associated pseudo-Meigs' syndrome. The treatment for this rare disease is still not standardized and poses a therapeutic challenge. Our case emphasizes the need for a multidisciplinary approach for managing *struma ovarii*.

Keywords: *Struma ovarii*, Meigs syndrome, Pseudo-Meigs' syndrome, Papillary thyroid carcinoma, Radioiodine

J Ayub Med Coll Abbottabad 2015;27(3):731–4

INTRODUCTION

Struma ovarii, 'goitre of the ovary' is a rare, mostly benign ovarian tumour that was first described by Boettlin in 1889.[1,2] This monodermal teratoma is characterized by the presence of thyroid tissue comprising more than 50% of overall tissue.[3] Yoo *et al*. have reported that the abdominal pain and distension or a palpable mass are the most common symptoms at presentation.[4]

Meigs syndrome represents a triad of ovarian fibroma, hydrothorax, and ascites in which ascites is expected to spontaneously resolve after surgical removal of the ovarian tumour. When ascites and hydrothorax are associated with tumours (benign or malignant) other than ovarian fibroma this qualifies as Pseudo-Meigs' syndrome.[5] Pseudo-Meigs' syndrome with *struma ovarii* is a rare finding and reported in up to 5% of cases.[6] The thyroid tumours within the *struma ovarii* are predominantly papillary thyroid carcinoma (PTC), classical PTC or follicular variant of PTC. No correlation has been found in the histopathological features of thyroid tumour cells and the clinical outcome of *struma ovarii*.[7] Currently, no definite protocols and guidelines exist about the treatment of patients with malignant *struma ovarii*. We highlight the role of multidisciplinary approach and radioiodine ablation treatment in malignant *struma ovarii*.

CASE REPORT

A 21 years old, unmarried woman presented with one month history of abdominal pain and distention. On workup, ultrasound followed by CT abdomen revealed a complex left adnexal mass and moderate to severe ascites. She underwent left salpingo-oophorecctomy, omental biopsy with ascitic fluid drainage. Figure-1 shows histologically: a. terminal deoxynucleotidyl transferase (Tdt) stain, b. High magnification, and c. Low magnification that revealed 'follicular variant of papillary thyroid carcinoma, arising in *struma ovarii*' from a 15cm mass with ruptured ovarian capsule (T3a). Omentum was involved by the tumour; however, fallopian tubes were spared. Postoperative CT scans showed minimal pelvic free fluid and post-surgical changes in left adnexa. CA-125 was elevated [35.9 u/ml–Normal <2u/ml]. In view of histology of papillary carcinoma, thyroid was evaluated for involvement. Patient was clinically and biochemically euthyroid. Tc99m pertechnetate thyroid scan and ultrasound thyroid demonstrated normal thyroid function and morphology. Thyroglobulin levels were 0.66ng/ml with elevated anti thyroglobulin, i.e., 1317 IU/ml [Normal reference <20 IU/mL], which essentially rendered thyroglobulin levels as unreliable. After review by multidisciplinary team, patient underwent total thyroidectomy and omentectomy. Histopathology from thyroid revealed benign thyroid

tissue. However, histopathology from diaphragmatic, *cul de sac*, peritoneal nodules and omental fat was positive for metastatic papillary thyroid carcinoma (follicular variant).

After thyroidectomy patient underwent metastatic work up. Diagnostic radioactive iodine (I-131) whole body scan (Dx WBS) was acquired with low dose radioactive iodine [Infinia II GE]. The scan revealed thyroid remnant with no evidence of I 131 avid metastasis (Figure-2). CT of the abdomen and the pelvis also did not reveal any morphological nodal or visceral disease.

Based on histopathologically proven peritoneal metastases the patient was treated with radioactive iodine ablation therapy (I131150 mCi). Serum thyroglobulin levels at point of therapy were 10.3 ng/ml, invalidated by elevated anti-thyroglobulins (384 IU/ml). Contrary to the Dx WBS, post therapy whole body scan (Rx WBS) at day 7 revealed multifocal I-131 avid metastatic disease involving skull, bilateral forearms, chest and abdomen (Figure-3). Following radioiodine therapy patient was started on oral thyroxin with the objective to suppress TSH (between 0.01–0.1 IU). Patient's disease responded to high dose radioiodine therapy and whole body SPECT-CT scan [Symbia T16 Siemens] acquired after second radioiodine therapy revealed residual I131 avid pulmonary nodules (Figure-4). In view of persistent I-131 avid metastatic disease patient has been treated with multiple dose of radioiodine (cumulative dose 450 mCi). Clinically she became symptom free; on 3 monthly follow up while taking thyroxin in supra-physiologic doses.

DISCUSSION

Struma ovarii rarely displays malignant potential. Association with Pseudo-Meigs syndrome is further rare and thus becomes a therapeutic challenge.[2,5,8]

Hysterectomy and bilateral salpingo-oophorectomy is the main treatment for *struma ovarii*. Since 95% of *struma ovarii* are benign and can occur in women of child bearing age it is critical to have a multidisciplinary approach. In the case of malignant and metastatic *struma ovarii* patient is to be treated on the lines of primary differentiated thyroid cancer.[9] The predominant benign outcome with up to 5% malignant potential poses a preoperative radiological challenge. Ultrasound, CT and MRI have been used to decipher benign from malignant with a few distinct features and considerable overlapping findings as well.[10] For the use of radioactive iodine total thyroidectomy is a prerequisite.

In literature a few case reports and reviews are available as a reference to plan the management of *struma ovarii* associated with thyroid malignancy. Devaney *et al*. studied 15 such cases with differentiated thyroid carcinoma; no recurrence was reported during the average follow-up period of 7.3 years, although no adjuvant radioiodine therapy was offered to these patients.[11]

DeSimone *et al*. have reported a literature review of 24 patients with *struma ovarii* and studied the use of radioactive Iodine.[12] In this review 4 patients were treated with thyroidectomy and radioactive iodine as adjuvant treatment and all remained disease free.

Seven patients received radioactive iodine for recurrent disease out of which 4 remained disease free while 3 developed further recurrences. DeSimone et al recommended that 'thyroidectomy and radioactive Iodine should be considered in the first line of management for malignant *struma ovarii*.[12]

Our case also benefitted from total thyroidectomy followed by multiple doses of radioactive Iodine. Malignant *struma ovarii* is a rare disease. Multi-organ involvement warrants a multimodality approach from the beginning for better clinical outcome.

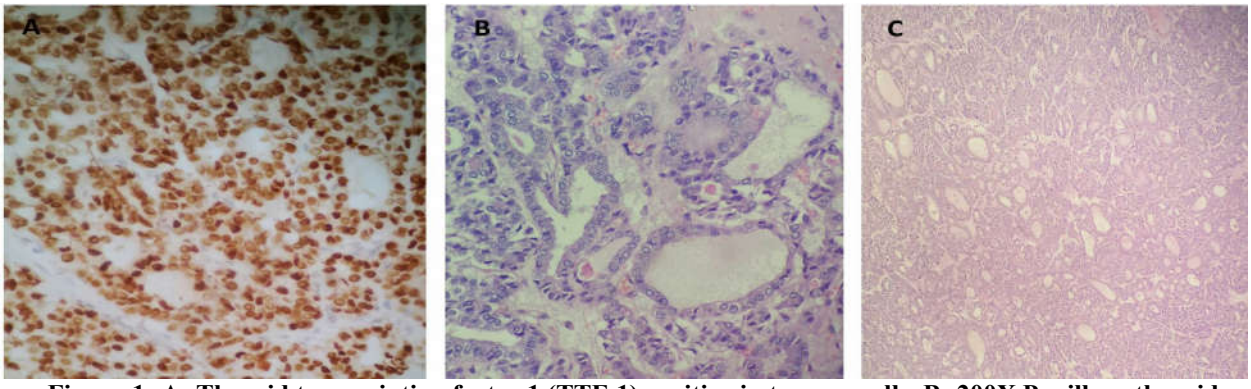

Figure-1: A: Thyroid transcription factor 1 (TTF-1) positive in tumour cells. B: 200X Papillary thyroid carcinoma C: 40X papillary thyroid carcinoma

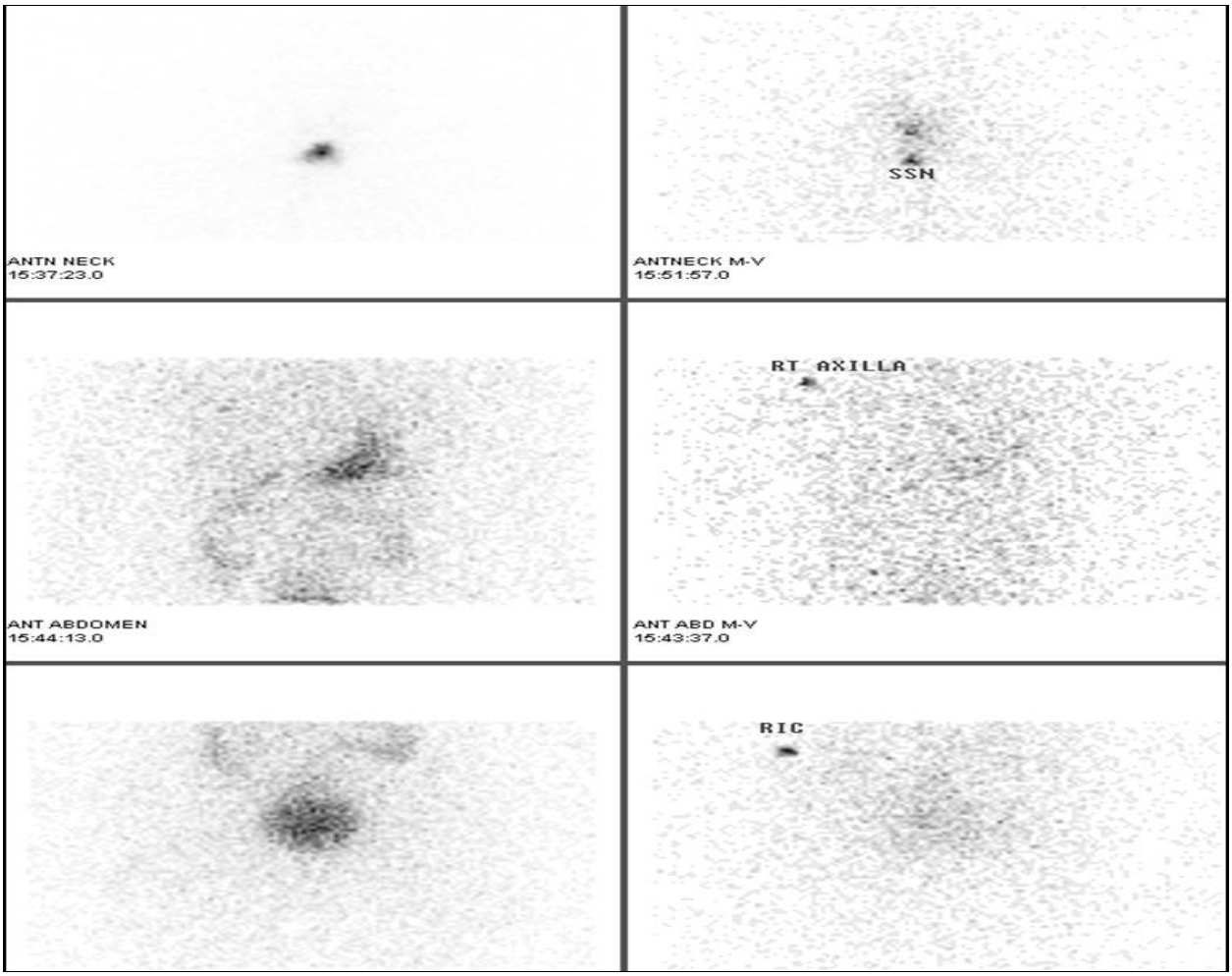

Figure-2: Diagnostic I-131 scan with whole body spot views in anterior acquisition shows foal avid uptake by the thyroid remnant. Physiological tracer distribution to the gastrointestinal tract and urinary bladder is seen

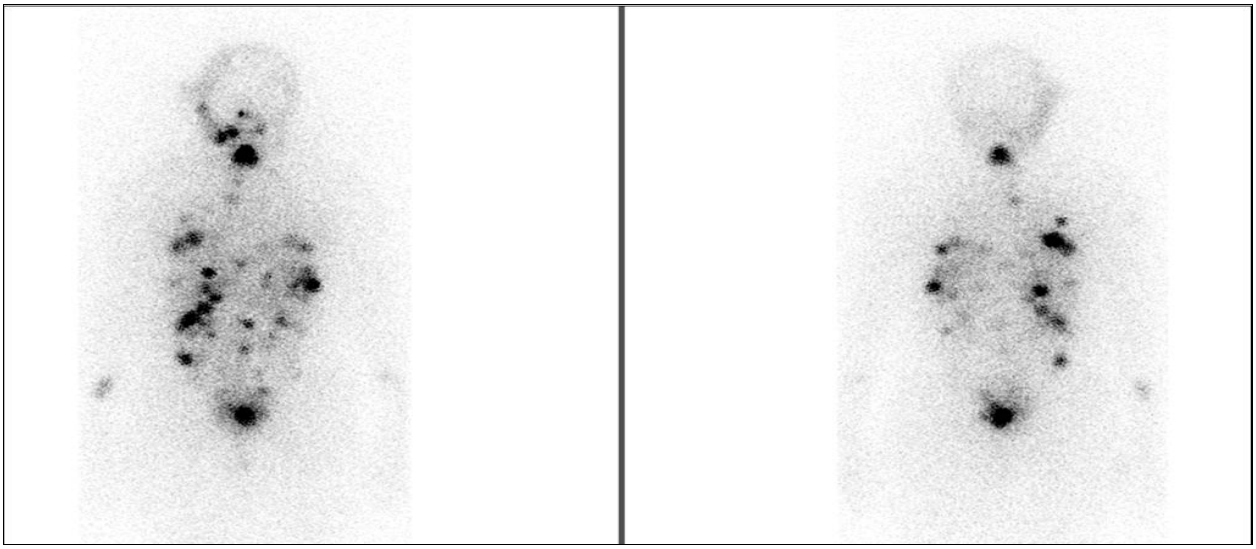

Figure-3: Post radioiodine ablation therapy (150mCi) whole body scan with anterior and posterior views acquired at day 7 showing multifocal I-131 avid disease involving skull, bilateral forearms, chest and abdomen.

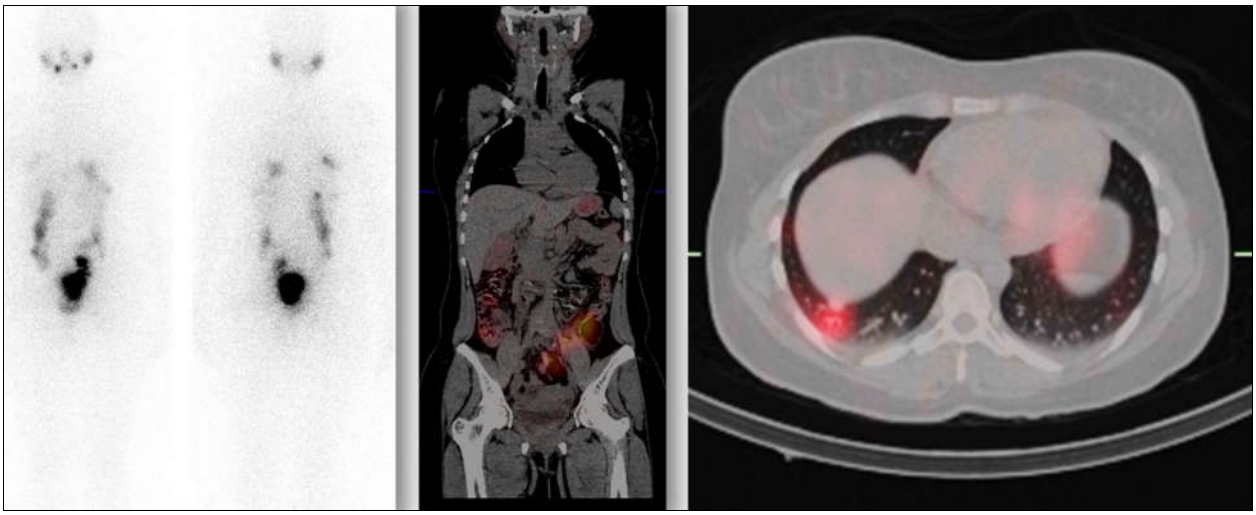

Figure-4: Post second radioiodine ablation therapy (200 mCi) whole body planar, fusion SPECT-CT coronal whole body and axial chest views show I-131 avid 8mm solitary pulmonary nodule in the basal segment of right lung. Physiological tracer distribution to the salivary glands, gastrointestinal tract and urinary bladder is seen.

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