

Late Manifestation of Peritoneal strumosis and widespread functioning lesions in the setting of Struma Ovarii simulating “Highly Differentiated Follicular Carcinoma” (HDFCO)

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Abstract:

Malignant transformation of struma ovarii is a rare event observed in about 5% of cases. We herein illustrate an unusual case that more closely simulated “highly differentiated follicular carcinoma” (HDFCO), another entity with relatively similar appearance with malignant struma ovarii (MSO). In our case, the peritoneal and systemic dissemination occurred 18 years following excision of ovarian struma that was reported benign on histopathology. Three features in the described case were noteworthy: (i) the highly functioning nature of the lesions (evidenced by low TSH even without thyroxine suppression), (ii) low/minimal FDG avidity of the foci, that could further reiterate the well differentiated nature of these lesions as expected in HDFCO and (iii) prominent involvement of rare sites such as spleen and liver, in addition to the usual sites such as lungs, peritoneum and bilateral adnexae.

Keywords: Peritoneal strumosis; Struma Ovarii; Radioiodine scan; FDG-PET/CT

Running Title: Late Presenting Peritoneal Strumosis - HDFCO

Introduction:

Struma ovarii is a rare monodermal variant of ovarian teratoma accounting for 2% of all mature teratomas. To be classified as struma ovarii, teratoma must be composed predominantly of mature thyroid tissue (>50%) (1,2). It is a benign condition, but malignant transformation can occur in around 5% of cases (3). It usually presents as pelvic mass and at times may present as pseudo-Meigs' syndrome in about 5% of cases (4). Benign thyroid tissue spreading to the peritoneal cavity and pathological examination of peritoneal implants showing multiple nodules of varying sizes of mature thyroid tissue similar to struma ovarii (10). The differential diagnosis of peritoneal nodules of struma origin (termed peritoneal strumosis) includes highly differentiated follicular carcinoma (HDFCO) (5) or typical thyroid carcinoma metastatic to the ovary, which has also been reported(6,7). Because of its harmless and matured appearance histologically, HDFCO cannot be diagnosed until the neoplasm spreads beyond the ovary, showing evidence of aggressive behavior (5). Histological features of thyroid carcinoma are found in 5–37% of struma ovarii (referred to as malignant struma ovarii). The nuclear features of papillary carcinoma are usually used as the criteria for diagnosis of malignant struma ovarii, and most cases reported are papillary carcinoma (8-11).

Case Report:

A 55 year old female, with histopathologically confirmed struma ovarii for which surgery was undertaken 18 years previously, recently presented with diarrhoea and lower abdominal pain. She had a previous history of similar presentation at 6 years following surgery in a different centre, when she had an incidentally detected right iliac fossa mass (7 x 6 cm) on ultrasonography. On Computed Tomography(CT) of the abdomen, there was evidence of colonic and peritoneal nodules and she had undergone revision surgery with removal of the right iliac fossa mass and colonic and peritoneal nodules. Histopathology showed mature thyroid tissue with many areas of follicular neoplasm within confirming it as strumosis peritonei. The iliac fossa mass was reported as recurrence of the struma ovarii.

In the present visit, abdominal Ultrasonography(USG)showed multiple peritoneal deposits, solid echogenic space occupying lesions in both lobes of liver, mixed echogenic mass with calcific foci within in the pelvic region and recurrence of bilateral adenexal masses. CT thorax showed multiple nodular lesions in both lungs being highly suspicious of metastasis. Biopsy from peritoneal nodules showed deposits of mature thyroid tissue with many areas of follicular neoplasm within indicative of recurrence of in the form of strumosis peritonei. 18-F-fluorodeoxyglucose(FDG)positron emission tomography with computed tomography showed multiple low grade FDG uptake in lesions in bilateral lungs, liver and spleen and bilateral adenexal masses (Fig 1A-1E) Neck (USG)was normal. The patient underwent total thyroidectomy, histopathology indicating normal thyroid gland and no evidence of malignancy. I-131 scan with 3 mCi (111 MBq) 4 weeks after surgery demonstrated multiple iodine avid lesions in the liver, bilateral lungs, spleen, multiple peritoneal deposits and bilateral pelvic deposits (Fig 2A). Following treatment with 7.4 GBq, post therapy I-131 scan (Fig 2B) during discharge showed more numerous peritoneal lesions compared to that of the diagnostic scan. At 3 months post-treatment she is clinically euthyroid with her thyroid profile (even without

thyroxine suppression) being T4- 12.8 mg/dl; FT4 1.88 ng/dl and Thyroid Stimulating Hormone (TSH) of 0.0003 microIU/ml (demonstrating highly functional nature of the metastatic lesions) and is under TSH profile monitoring every 6 weeks). The serum thyroglobulin level was >300ng/ml. During writing this case the patient is doing well on follow up and is ambulatory. She is due for next cycle of radioiodine therapy in 3 months.

Discussion:

Literature regarding the course of malignant struma ovarii is based upon case reports and series, the predominant sites of metastases being adjacent pelvic structures and peritoneum, but rarely also been observed in lungs, liver, brain and skeletal sites. Roth et al (5) evaluated the relationship of HDFCO and cases reported as peritoneal strumosis and finally doubted the existence of the latter as a distinct clinicopathologic entity. They recommended an approach similar to the present strategy: local resection of the extraovarian tumor and subsequent thyroidectomy followed by radioactive iodine ablation for these patients.

Radioiodine scan is considered pivotal imaging modality in differentiated thyroid cancer useful in detecting the presence of disease and deciding on therapy. As seen in this case struma peritoneii mainly consists of differentiated thyroid tissue, radioiodine scan should be integrated in the follow-up of these cases. I-123 scan is the preferred choice owing to its better imaging characteristics compared to I-131; however, the former is not yet available in all countries. The low FDG avidity of the lesions on PET-CT is explained by presence of differentiated thyroid tissue in the lesions and goes in accord with its potential role in predicting disease biology.

Conclusion

The present case more closely simulated the entity of highly differentiated follicular carcinoma of ovarian origin (HDFCO) (5), where recurrence occurred twice (at 6th and 18th years after excision of ovarian struma, which was reported benign on histopathology), with only peritoneal metastasis at first but peritoneal and systemic dissemination during the second presentation. The functioning nature (evidenced by low TSH even without thyroxine suppression) and (ii) low/minimal FDG avidity of the foci is also noteworthy, that could further reiterate the well differentiated nature of these lesions as would be theoretically expected in HDFCO.

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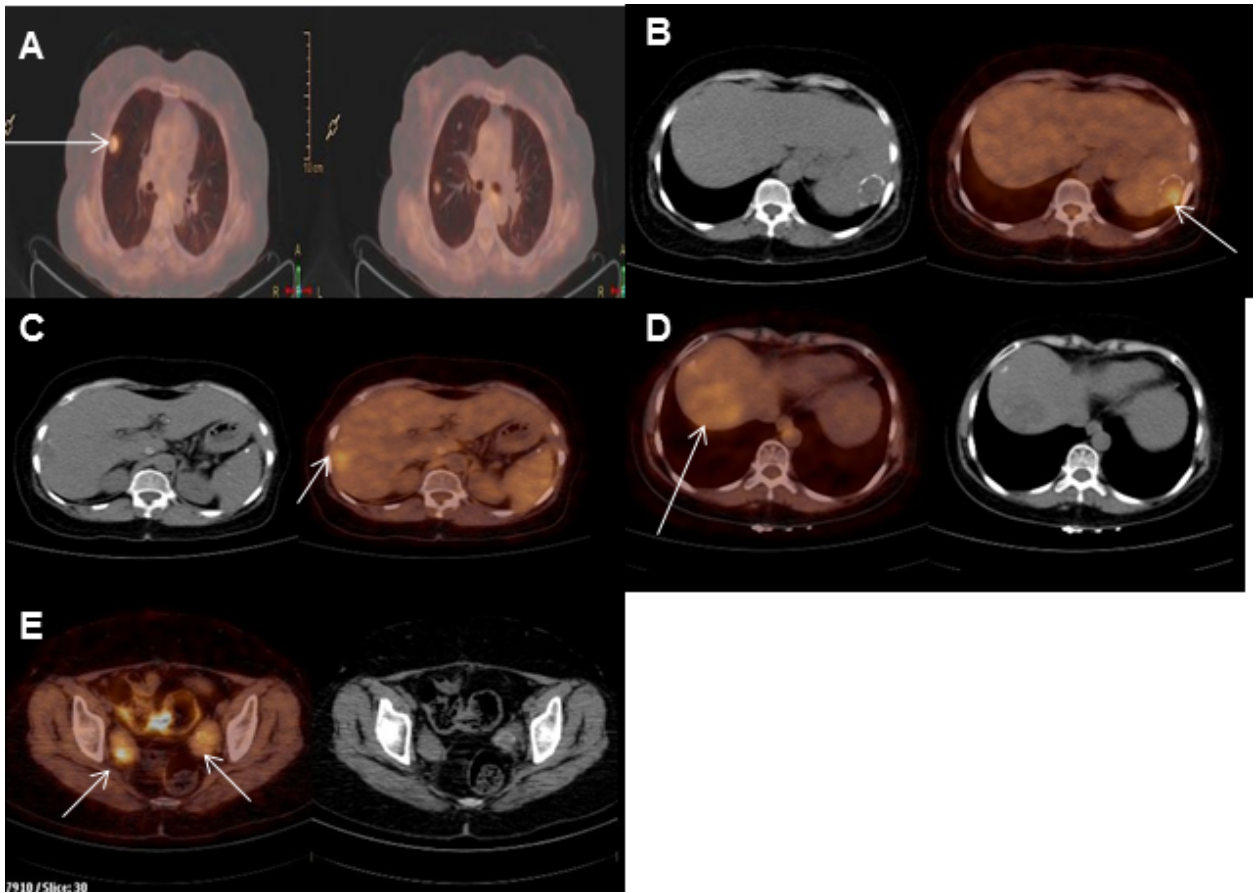


Figure 1

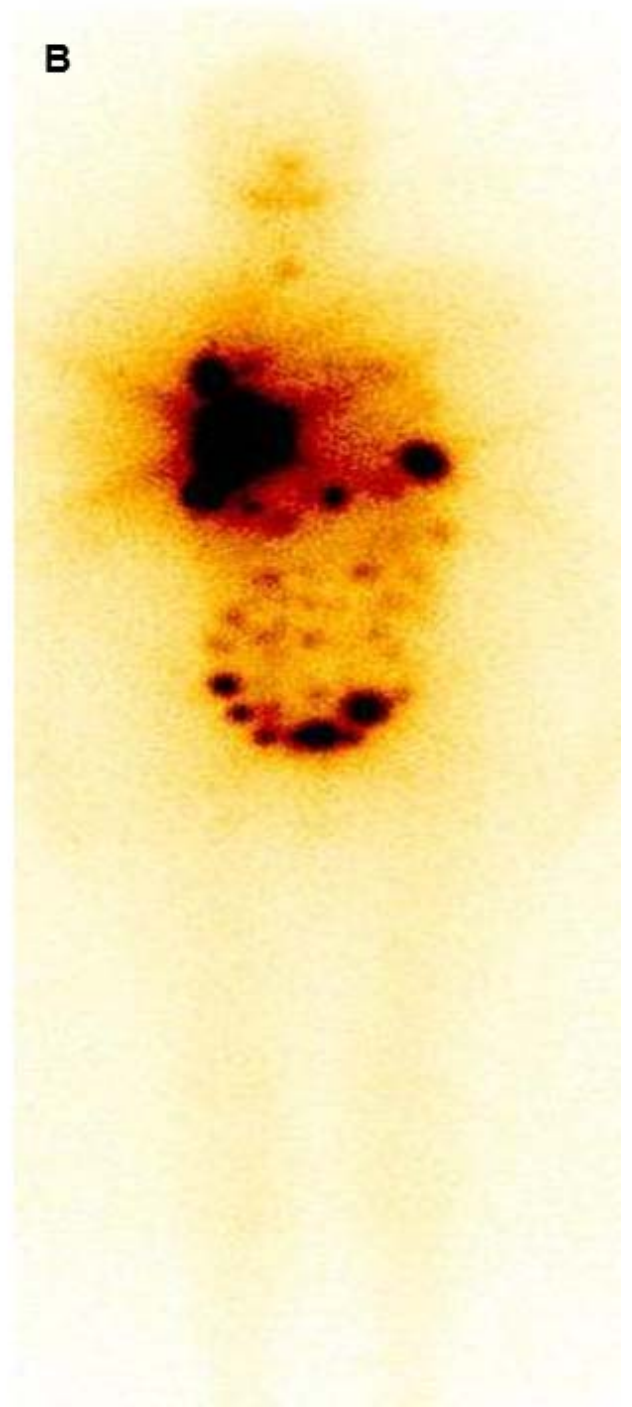
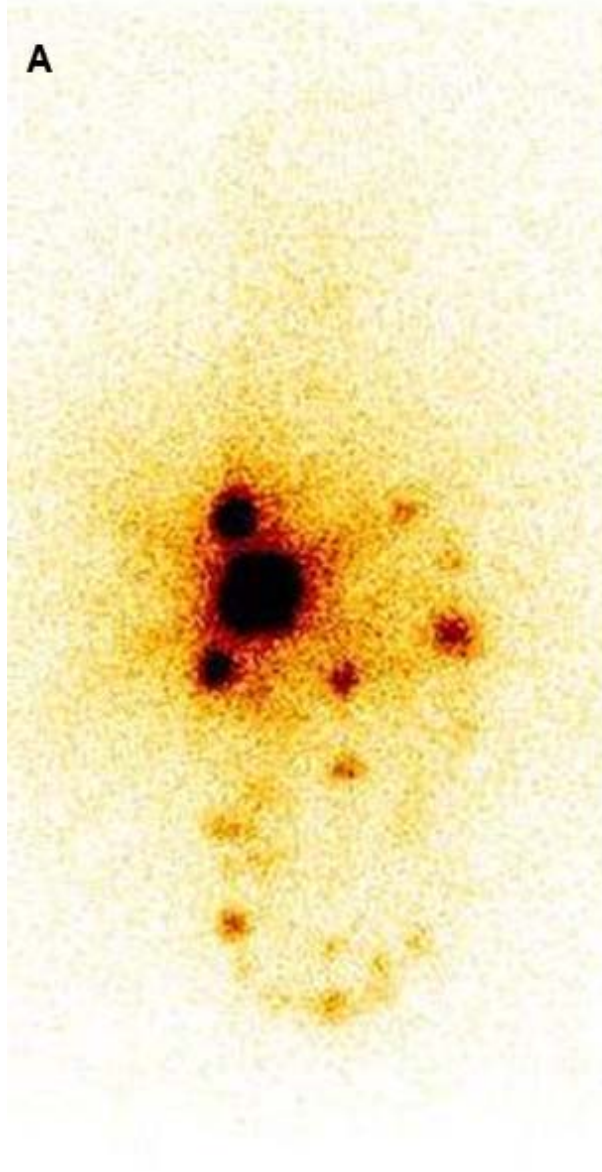


Figure 2