Highly-differentiated follicular carcinoma arising in struma ovarii with metastatic omental involvement

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Introduction

Highly-differentiated follicular carcinoma of ovarian origin (HDFCO) is a recently recognised entity, first described by Roth & Karseladze in 2008.[1] The diagnosis can be made when struma ovarii without histological evidence of malignancy (i.e. papillary or follicular carcinoma) results in extra-ovarian disease. HDFCO can resemble either normal thyroid, colloid/adenomatous nodule, or an adenoma-like lesion.[2]

Considered a type of struma-derived thyroid-type carcinoma,[3] HDFCO is distinct in that the diagnosis of 'carcinoma' cannot be made unless extra-ovarian disease is found. As such, it is possible, indeed usual, for there to be an interval of decades between the original finding of struma and the subsequent finding of extra-ovarian disease and diagnosis of HDFCO (the longest reported being 48 years).[4]

The entity is very rare, with only 28 cases described in the literature that we are aware of.[2,5,6] The prognosis appears to be very good, with no definite reported deaths from the disease, although studies are limited in this respect.[2,6] Lesions can be locally destructive, however, for example when affecting the facial bones.[2] The natural history is otherwise effectively unknown as patients are typically treated aggressively with radio-iodine therapy, which seems to be effective.

We present a case of HDFCO which had spread at the time of its original detection.

Case report

A 55 year-old presenting with acute breathlessness was found to have Meigs's syndrome. A laparoscopic hysterectomy and bilateral salpingo-oophorectomy and omental biopsy was performed. The omentum was adherent to the abdominal wall. Macroscopically the left ovary was replaced by a cyst measuring 220 x 160 x 50 mm, with a smooth serosal surface and a solid/cystic interior with a yellow, greasy cut surface. The omentum appeared unremarkable.

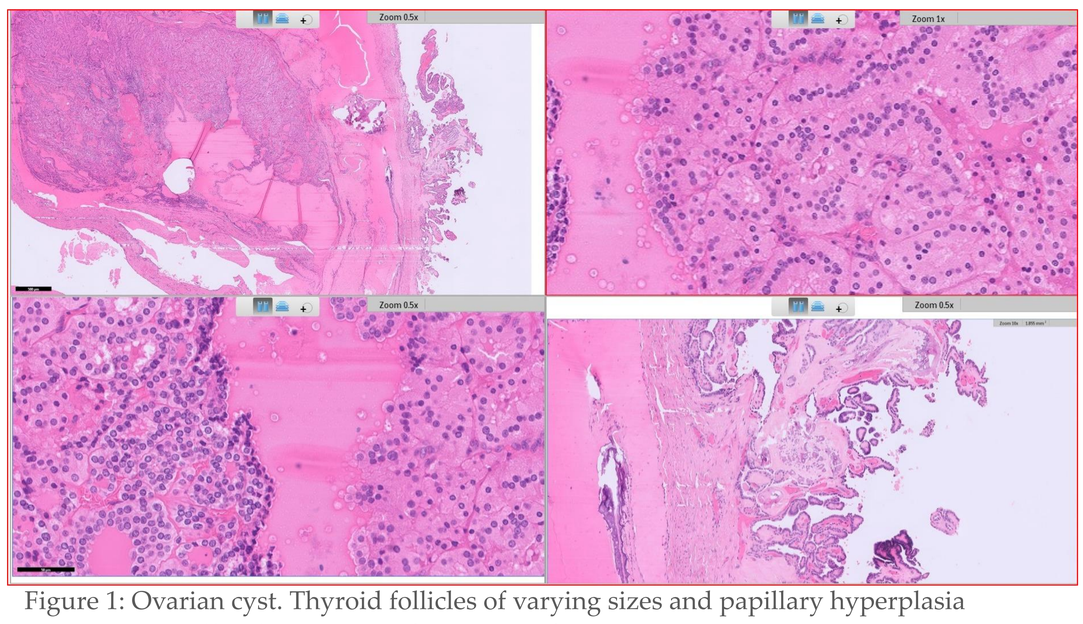

Figure 1: Ovarian cyst. Thyroid follicles of varying sizes and papillary hyperplasia

Microscopically there were features of a mature cystic teratoma, containing predominantly struma ovarii, with accompanying adipose tissue, squamous epithelium, and pilosebaceous units.

The struma consisted of thyroid follicles of varying sizes and shapes, with focal calcification and small papillary elements. There were no nuclear features of papillary thyroid carcinoma and no vascular space invasion or significant mitotic activity.

The omentum contained foci of papillae covered by a single layer of cuboidal cells without nuclear features of papillary thyroid carcinoma. Strong immunoreactivity for TTF-1, BerEP4 and CK19 and negativity with WT-1 and calretinin confirmed thyroid differentiation.

The combination of struma ovarii without histological evidence of malignancy and extra-ovarian metastasis confirmed the diagnosis of HDFCO with omental metastases.

The multi-disciplinary team recommended radio-iodine therapy, however the patient failed to attend further appointments, and despite multiple attempts to arrange clinic appointments, was lost to follow-up.

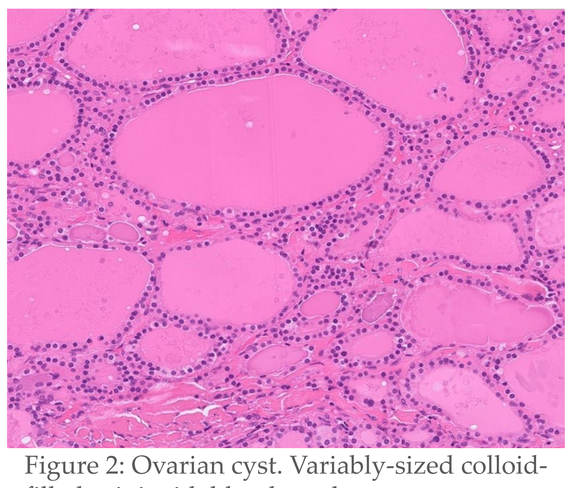

Figure 2: Ovarian cyst. Variably-sized colloid-filled acini with bland cytology

Discussion

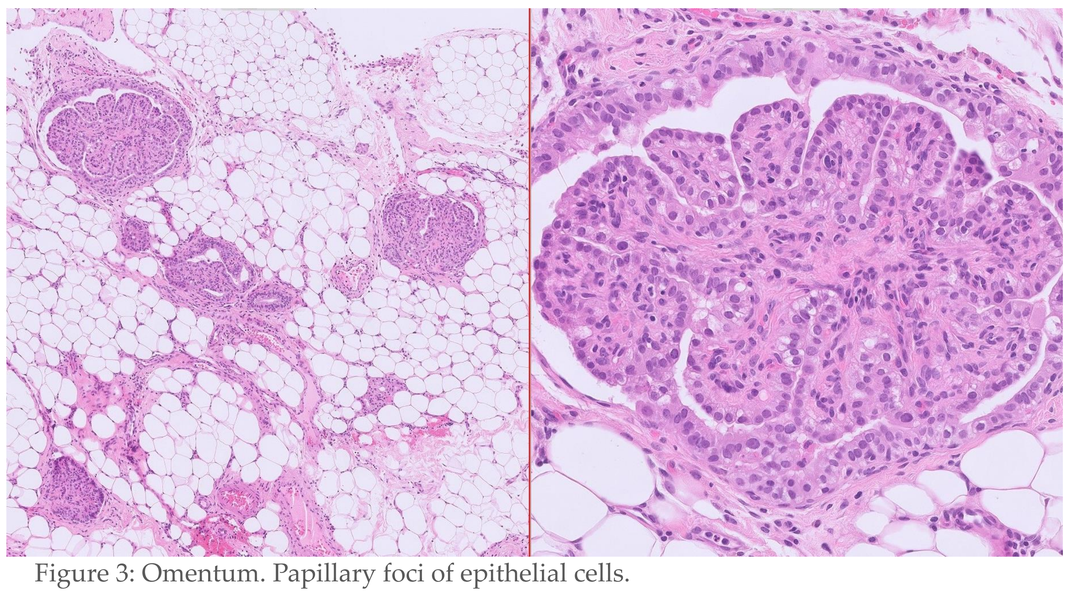

Figure 3: Omentum. Papillary foci of epithelial cells.

The differential diagnosis in cases of HDFCO includes metastasis from a thyroid carcinoma, particularly a well-differentiated follicular carcinoma. This is partly the rationale for treatment with radio-iodine therapy. It has been postulated that such therapy may not be necessary in cases of HDFCO confined to the peritoneum and peritoneal nodes due to the slow rate of progression, however there is no evidence for this and the approach is not recommended.[2]

Follicular or papillary carcinomas (particularly follicular-variant papillary carcinoma) arising in struma ovarii which metastasise are another consideration, especially where the struma contains benign and malignant elements.[7] Thorough sampling of struma ovarii is recommended as a finding malignant elements is likely to indicate more aggressive behaviour.

From a pathologist's perspective, effective communication with the multidisciplinary team is an important aspect in such rare cases with distinguishing features which are important but may be subtle.

This case is unusual in that the diagnosis of HDFCO was made at the time of of its original detection; this is only the second such case confirmed as such that we are aware of, the other having been reported in Roth & Karseladze in their original paper.[1] The significance of this and whether it represents more aggressive disease is unknown.

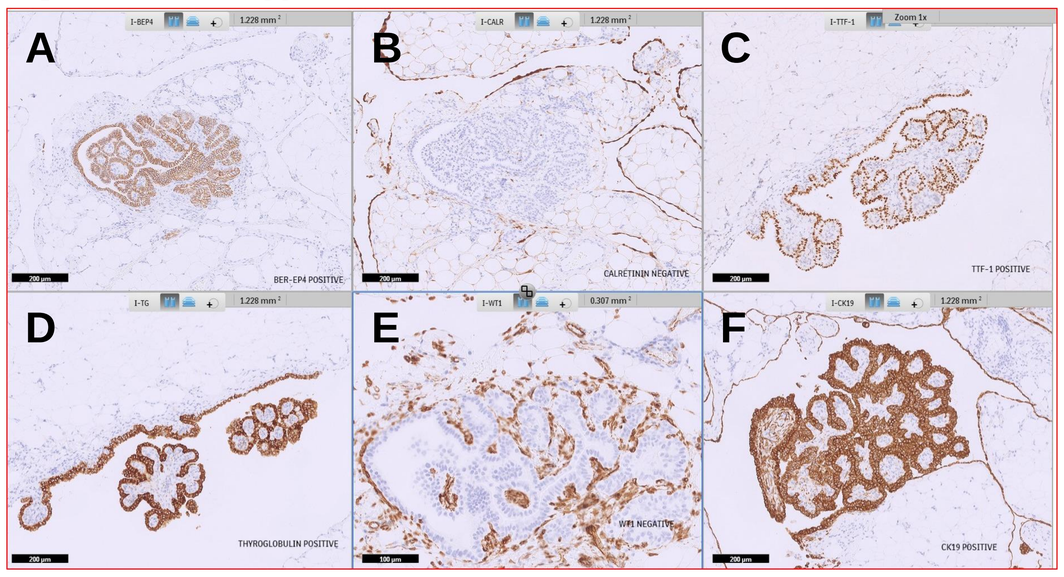

Figure 4: Omentum. A. BerEP4; B. Calretinin; C. TTF-1; D. Thyroglobulin; E. WT1; F. CK19.

Key points

- **HDFCO can be diagnosed when struma ovarii without histological evidence of malignancy is associated with extra-ovarian disease**
- **Diagnosis is often retrospective when metastasis is found, commonly decades after the original excision of struma.**
- **Prognosis appears to be good, however lesions can be locally destructive**
- **Treatment involves radio-iodine therapy**
- **Distinction from similar entities is essential as is clarity among the clinical team.**

Acknowledgement & References

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