Metastatic Malignant Struma Ovarii Presenting as Paraparesis from a Spinal Metastasis

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A 42-yr-old woman had a solitary metastases to her spine (T3) from a malignant struma ovarii. The thyroid was excluded as the site of the primary cancer. The lesion caused paraparesis. The spinal metastasis was treated by surgery and two doses of 131I (200 mCi each time). The patient responded very well and is entirely free of symptoms and signs. Repeat whole-body 131I scan shows no abnormality.


Struma ovarii is a very rare ovarian tumor which can present in various ways. It can be discovered on pathologic examination of an asymptomatic ovarian mass, it can present as a cause of ascites and or hydrothorax, it can cause hyperthyroidism (1–3) and very infrequently malignant transformation of the tumor can occur and be a source of metastases. This tumor is extremely uncommon (4,5). It is defined as, “a teratoma in which thyroid tissue is present exclusively or forms a grossly recognizable component of a more complex teratoma” (6). The thyroid tissue is usually benign (7) but malignant changes have been described (8–13). Distant metastases are extraordinarily rare (14) and Pardo-Mindan and Vazquez (15) in 1983 could only find 18 cases in the world literature including a patient of theirs. Bony metastases from malignant struma ovarii have been described in only five patients (1,16–19).

The purpose of this case report is to describe a patient who has a single metastatic thyroid carcinoma (of ovarian origin) in the spine. The primary ovarian lesion contained both benign thyroid and papillary thyroid carcinoma. She has been treated successfully by surgery and radioiodine 131I.

For several years she had upper backache which had been attributed to stress; however, a radiograph from a chiropractor’s office on 2/22/84 showed complete loss of the body of the second thoracic vertebra. When she had an episode of acute abdominal pain in February, 1985 a left irregular, firm, tender ovarian mass (10 x 8 cm) was found on gynecologic examination. A mature cystic teratoma containing benign thyroid tissue was diagnosed pathologically. In the months after the surgery she noted altered sensation from the breasts downwards. The sensory changes progressed until her transfer and first admission to our institute. She had a burning dysesthesia over the anterior thighs and numbness and tingling in lower legs and feet. She had been incontinent of urine on several occasions in the preceding 2 mo.

On examination she was euthyroid. There was loss of touch and pain sensation from T3 caudad. Power in lower limbs was normal, reflexes were hyperactive, and plantar responses were equivocal. Computed tomographic (CT) and magnetic resonance imaging (MRI) scans showed a large tumor mass at the level of T3 involving the body of T3, with a bilateral and anterior paraspinal components, plus a mass projecting posteriorly which impinged on the spinal cord. A myelogram showed obstruction to flow of contrast. The patient underwent an emergency operation to relieve pressure from the cord, to debulk as much tumor as possible, and to obtain tissue for diagnosis. The tumor was extremely vascular and on frozen section the pathology was interpreted as metastatic follicular carcinoma probably from a thyroid primary. Since the surgical approach was anterior the thyroid was inspected and palpated and judged to be normal. Thyroidectomy was not performed because of some uncertainty about the tissue of origin on frozen section. The spine was supported by a fibular graft and an external halo put in position.

The final pathologic diagnosis was follicular cancer of the thyroid metastatic to bone. A thyroid scan (200 mCi131I) was
technically difficult because of the external halo but showed uptake in the thyroid and in the spinal lesion. A whole body $^{131}$I scan (2 mCi$^{131}$I) confirmed these findings and showed no other areas of uptake. By this time the ovarian pathology had been reviewed and the diagnosis of struma ovarii containing both typical thyroid and papillary carcinoma was made.

Thyroideotomy was advised for two reasons. Firstly, from the scans it was clear that normal thyroid was concentrating considerably more $^{131}$I than the spinal metastasis, and if the metastasis was to be successfully ablated with $^{131}$I, removal of the bulk of normal thyroid would be important. Secondly, careful sectioning of the thyroid would ensure that a primary thyroid carcinoma was excluded. A near total thyroideotomy was undertaken on 7/4/86 with no complications. The operation was conducted after removal of the halo frame with the patient's head and neck stabilized with sandbags. No cancer was found.

A second whole-body $^{131}$I scan one month after surgery, without thyroid replacement, showed substantially more uptake in the spinal lesion with only a small remnant of thyroid visible. Uptake of $^{131}$I in the spinal lesion was 5%. The volume of the cancer was estimated to be 50 G. The patient was treated with 200 mCi$^{131}$I on 7/31/86. The neurological signs resolved entirely, the halo was removed in September, 1986, and she was able to commence studies in art school and recommence physical activities including tennis. A repeat MR scan on 12/19/86 showed no impingement of the cancer on the spinal canal and no cord compression with a decrease in the soft tissue mass. The patient was rescanned on 3/27/87, 72 hr after 2 mCi$^{131}$I and 4 wk after stopping exogenous thyroxine. At the time of scan TSH was 70 $\mu$U/ml, and $^{131}$I uptake in the lesion 1%. No other lesions were noted on scan. A second therapeutic dose of $^{131}$I (200 mCi) was prescribed at that time and tolerated well. She continues to be normal in all respects. Repeat whole body scan showed no abnormal uptake of $^{131}$I, at the time of scanning TSH was $>60$ $\mu$U/ml and Tg $<5$ ng/ml.

PATHOLOGY
Ovarian Lesion

The left ovarian mass revealed abundant mature appearing follicular thyroid tissue as well as areas of typical papillary carcinoma. The former was characterized by variably sized colloid-filled follicles lined by bland, low cuboidal thyroid type epithelium (Fig. 1A and B). This tissue had irregular, bluntly infiltrative borders into adjacent ovary, but no vascular invasion. Foci of papillary carcinoma contained only occasional papillary structures, which were characterized by greater cellularity with nuclear crowding and large pleomorphic nuclei. “Orphan-Annie” chromatin clearing and nuclear grooves were also evident (Fig. 2A and B).

**FIGURE 1**
A: Low power of ovary showing irregular border between struma ovarii and adjacent ovarian stroma are noted (40×). B: Higher power shows fairly normal follicular architecture with considerable variation in follicle size which suggests a predominantly benign lesion (50×).
Spinal Lesion
Sections of bone from first and second thoracic vertebrae showed metastatic follicular thyroid carcinoma which was histologically quite bland, and which closely resembled the follicular thyroid tissue in the ovary (Fig. 3A and B). Immuno-peroxidase studies showed positive reaction to thyroglobulin. No papillary carcinoma was found.

Thyroid
The entire thyroid specimen was sectioned at 2.0-mm intervals. Macroscopically and microscopically there was no evidence of a primary thyroid cancer or any evidence of metastases.

DISCUSSION

Struma ovarii is a rare ovarian tumor. To fulfill this diagnosis the ovarian lesion should be composed entirely of thyroid tissue or contain grossly recognizable thyroid elements (6). In two series of 1000 and 2000 ovarian tumors, struma ovarii accounted for 0.3% (4) and 0.65% (5) of cases. Malignant struma ovarii are very rare and most malignant struma have not metastasized (20–22).

Identification of carcinoma in struma ovarii should be straightforward in instances where papillary carcinoma is present because the diagnosis is primarily a cytologic one. However, because ovarian strumas are typically not encapsulated, consequently capsular-invasive criterion cannot be used. Diagnosis of follicular carcinoma can be extremely difficult in the absence of vascular invasion. In our view, follicular carcinoma should not be diagnosed in a struma ovarii unless metastases are documented or unequivocal vascular invasion is present. We interpreted the vertebral metastases in this case as probable metastatic follicular variant of papillary carcinoma.

Struma ovarii can be asymptomatic, can cause ascites and hydrothorax (Meig's Syndrome) (1), or hyperthyroidism (1–3) or rarely symptoms due to metastases (8–18). Pardo-Mindan and Vazquez (15) could only find 17 cases with metastases in the literature and added a further case. Only five have been diagnosed with skeletal involvement which makes this case worthy of reporting. Wynne et al. (16) described a 26 yr old who developed pain in the right ischium 1 yr after removal of a “benign” ovarian teratoma which contained only thyroid. Biopsy of the ischium showed well differentiated thyroid tissue. The patient reported by Eerland (18) was a 60-yr-old who had a metastasis to the cranial

FIGURE 2
A: Foci of papillary architecture in the ovarian lesion are suggestive of papillary carcinoma (125x). B: Higher power showing nuclear crowding and pleomorphism and open chromatin distribution and characteristic nuclear grooves (arrows) (500x).
vault. Although the 67-yr-old patient of DeBakay and DeLeloczy (19) had a brain metastasis, at the end of their brief article they state the patient had "struma metastases arising in the skull and brain", but give no other details. Kempers et al. (1) describe a patient who developed liver and bone metastases 7½ yr after presentation with malignant struma ovarii. Gonzalez-Angulo et al. (8) make reference to a patient who had metastases to bone (17) but provide no clinical details.

In this patient the skeletal lesion was proven pathologically to be thyroid and this was substantiated by its ability to trap and concentrate iodine. The thyroid was excluded as the primary source by careful pathological evaluation of 2-mm sections, which were all normal. Thyroidectomy made possible therapy of the spinal metastasis with $^{131}$I. There are few reports of treatment of functioning metastasis by this approach (1,11). One with a pulmonary lesion did well (14). Although the number of cases of localized and metastatic malignant struma ovarii is small, it is thought that the prognosis is similar to that of similar stage and type of thyroid cancer arising in the thyroid.

REFERENCES

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