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Malignant struma ovarii in an 11-year-old girl

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ABSTRACT

Struma ovarii is a highly specialized form of mature ovarian teratoma in which contains greater than 50% thyroid tissue and absence of uniform diagnostic criteria combined with the rarity of the tumor contributes to the large discrepancy in the reported frequency. When the characteristic features of thyroid carcinoma are present, the lesion is termed malignant struma ovarii. An 11-year-old girl presented with palpitation and right lower pain that was diagnosed with right ovarian mass and was ultimately treated by right abdominal salpingo-oophorectomy. Postoperatively, she was diagnosed with a malignant struma ovarii. The patient was subsequently treated with thyroidectomy and is currently disease free.

Malignant struma ovarii is a medical rarity and previous studies have reported cases with struma ovarii in adults and to our best knowledge this is the first describing struma ovarii in a girl before puberty.

Conclusion: according to results, Struma ovarii should be considered in the differential diagnosis of female presenting with hyperthyroidism and without clear etiology; and radioactive uptake and scan and thyroglobulin levels can be useful diagnostic tools.

1. Introduction

Struma ovarii is nearly always a teratoma because there are many cellular components in an ovarian tumor. Malignant struma ovarii is very rare. Struma ovarii is approximately 0.3% of all ovarian tumors and 3% of ovarian teratomas. The age presentation of Struma ovarii is between 40 and 60 years old [1]. The first case of ovarian teratomas containing thyroid tissue was reported in 1889 by Boettlin [2]. Struma ovarii with Thyroid ectopic tissue can be reported accidentally after adnexectomy, but it's occurrence as a primary diagnosis of hyperthyroidism is rare [3]. Recently an unusual rare Pancreatic Struma with Papillary Thyroid Carcinoma is reported [4].

Ludwig Pick recognized that struma ovarii composed of thyroid tissue in the early part of the 20th century [5].

Struma ovarii is a highly specialized form of mature ovarian teratoma. While approximately 15% of teratomas contain thyroid tissue, struma ovarii can only be diagnosed when thyroid tissue is the predominant element [6].

Absence of uniform diagnostic criteria combined with the rarity of the tumor contributes to the large discrepancy in the reported frequency [6].

Most cases of malignant struma ovarii are subclinical. The typical patient presents with pelvic pain. It is rare for patients to present with overt signs of hyperthyroidism [7] such as Weight loss, fatigue, heat intolerance, tremor, and palpitations [8]. Both papillary and follicular carcinomas have been reported in a setting of struma ovarii with follicular carcinomas being more common [7]. Rare forms of hyperthyroidism due to extrathyroidal production of thyroid hormones such as malignant struma ovarii can be identified by whole-body scanning techniques [9]. As in our country compared to others, different etiologies, presentations, type of drugs, age groups and prevalence have been noted for thyroid disorders [10–14] we aimed to report an unusual case of follicular and follicular variant of papillary thyroid carcinoma causing hyperthyroidism in unusual age in Struma Ovarii.

2. Case report

An 11-year-old female presented with palpitation. She was diagnosed with hyperthyroidism and underwent therapy with methimazole for a year. She also experienced mild abdominal Right Lower Quadrant pain and due to lack of probable response to medical therapy, thyroid scan was done 20 min after IV injection of 5 mci radiotracer in anterior

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Nomenclature

IV injection	intravenous injection
LT4	levothyroxine
TSH	thyroid stimulating hormone
Tg	triglyceride
FT4	free T4
WBS	whole body scan

projection. In the scan, no functional thyroid tissue in the thyroid gland bed was mentioned and further evaluation with ultrasound and radiologic modalities were recommended.

Regarding to the abdominal pain and normal thyroid scan, Abdominopelvic ultrasound was done. I showed a huge 113*112*100 mm ill-defined heterogeneous solid-cystic complex mass with volume measured about 670 ml within pelvic cavity with anterior replacement of bladder (Fig. 1). No sign of ascites was seen.

MRI of abdominopelvic with and without contrast showed huge ill-defined lobulated heterosignal mass measuring about 11*10*9 cm in pelvic cavity with heterogenous nodular enhancement that was limited by abdominal wall anteriorly, by sacrum and rectum posteriorly and by pelvic side wall laterally and compressed urinary bladder. No metastasis and adenopathies were noted.

After ruling out the pregnancy, patient underwent right salpingo-oophorectomy.

3. Pathologic finding

The right side ovarian mass of an 11-years-old girl sent to pathology ward consisted of a large solid mass measured 12 × 12 × 10 cm with attached fallopian tube. The histopathologic sections (Fig. 2) revealed rare mature teratomal components like mature cartilage and salivary glands but most of the tumor composed of thyroid follicles with both atypical structural and cytopathological features with massive tumoral necrosis and marked capsular invasion (Fig. 3) which was consistent with the diagnosis of Follicular carcinoma along with some area of Follicular neoplasm with cytopathological specifications of papillary thyroid carcinoma including Orphan annie-eye nuclei that was compatible with the diagnosis of Follicular variant papillary thyroid carcinoma. Overall this mass was reported as malignant struma ovarii with two components of Follicular carcinoma and follicular variant of papillary thyroid carcinoma.

No malignant cell was seen in abdominal fluid cytology.

Postoperatively, thyroid function was evaluated, which showed a TSH level of 2.83uIU/ml (within normal limit), thyroid scan also was obtained which showed normal size with hemogenous tracer uptake, suggesting a normal scan. Thyroid ultrasound showed left lobe measuring about 35*10*9 mm and right lobe measuring about 35*8*11 mm with homogenous parenchymal echo and normal vascularity.

She underwent total thyroidectomy and remains free of symptoms and recurrence to this date.

4. Discussion

Struma ovarii is a rare highly specialized monodermal teratoma in which contain greater than 50%of thyroid tissue and may demonstrate all the pathologic features observed in the thyroid gland such as normal thyroid tissue, thyroid adenomas, and carcinomas, and represent less than 3% of all ovarian teratomas [15,16].

When the characteristic features of thyroid carcinoma are present, the lesion is termed "malignant struma ovarii" which is extraordinarily rare and accounts for only 5%–10% of Struma ovarii and 0.1% of all ovarian tumors [16].

Our case is an 11-year old- girl, however, it is most commonly seen in the fifth and sixth decades of life. The most common presenting symptom is a pelvic mass, and in 5–8% of cases clinical manifestations of hyperthyroidism are seen [17,18]. Although malignant struma ovarii causing hyperthyroidism is rare, this age presentation is very rare. Boyd JC et all reported malignant Struma ovarii in a 30-year old nulliparous patient [19] and suggested aggressive surgical management followed by radioiodine therapy for reduction of recurrence risk.

Our case presented with palpitation and slight abdominal pain, which according to Samina Makani et al. review study of cases with malignant Struma Ovarii a 45% of cases presented with a pelvic mass, 40% with abdominal pain, 9% with menstrual irregularities and 5% with hyperthyroidism [18].

Our case is an extremely rare form of ovarian tumor with the component of 60% well-differentiated thyroid follicular carcinoma, and 40% well-differentiated follicular variant of papillary thyroid carcinoma and the neoplasm shows capsular invasion, which according to Maria D. Navarro et al. study has been reported in only 5 cases of papillary and follicular carcinoma and 1 case of pure struma ovarii; capsular invasion has been described in the follicular variant of papillary carcinoma in Maria D. Navarro et al. case for the first time [20].

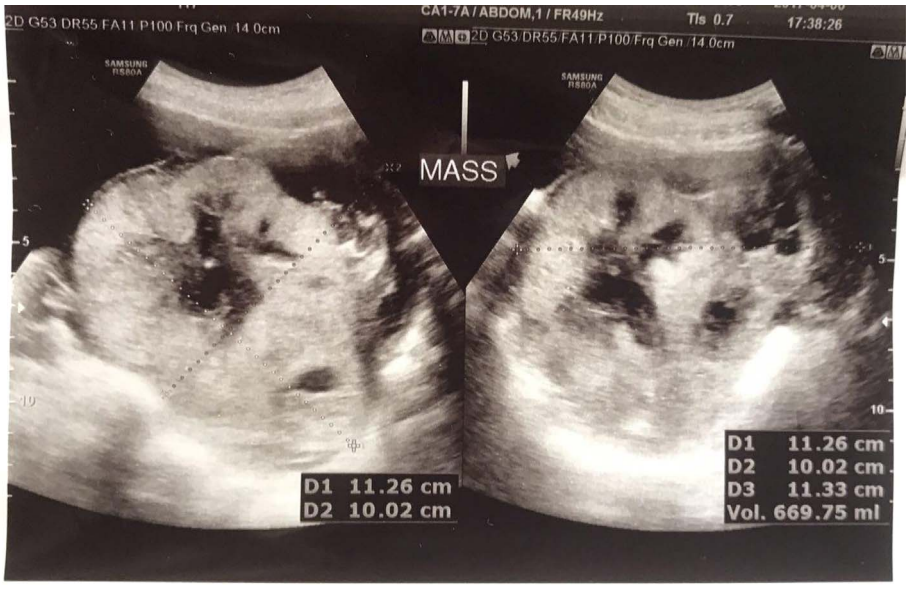

Fig. 1. Ill-defined heterogenous solid-cystic complex mass within pelvic cavity.