Cytological diagnosis of Spindle Epithelial Tumor with Thymus-like Differentiation (SETTLE)

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Citation

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Abstract

Spindle epithelial tumor with thymus like differentiation (SETTLE) is a very rare tumor arising either from ectopic thymus or remnants of branchial pouches that retain the potential to differentiate along the thymic line.[1]; Herein is reported a case of SETTLE in a 50-year-old man who presented with dessiminated intravascular coagulopathy. The subsequent work up revealed the presence of a mediastinal mass. A presumptive clinical diagnosis of lymphoma was made, forwhich the patient received a single dose of radiation. The patient's condition detriorated andnpatient died 5 days later. At autopsy, a thyroid mass of 10x8 cm was found with necrotic friable yellow tan color, extending into the mediastinum, obstructing the superior vena cava with extension into the right atrium and right ventricle forming a hemorrhagic mass of 12x7x6 cm. A touch preparation from the thyroid lesion demonstrated a biphasic lesion consisting of spindle and epithelial glandular components. The tissue specimen from the case demonstrated the spindle cells to stain positively for cytokeratin and vimentin but were negative for thyroglobulin and calcitonin as well as other neuroendocrine markers confirming the diagnosis of SETTLE.

CASE REPORT

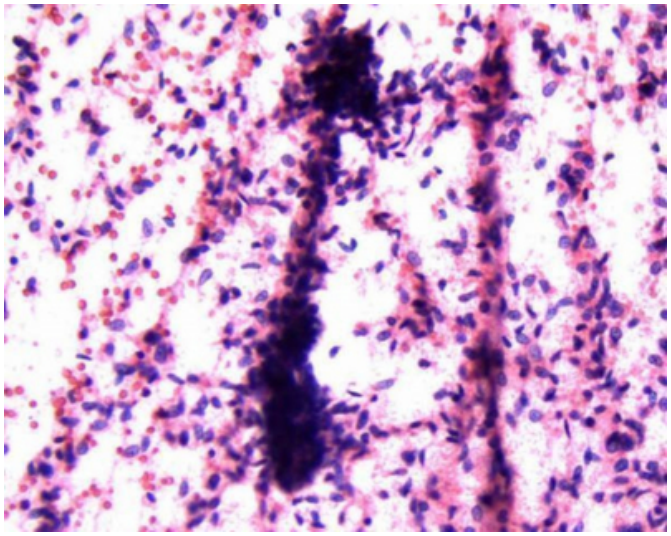
A 50-year-old man presented with shortness of breath and paroxysmal nocturnal orthopnea of 4 months duration. Fatigue and a 20 pound weight loss were noted over the same time period. Past medical history revealed multinodular goiter, smoker and alcohol abuse. On examination the patient was jaundiced with an elevated JVP. Initial investigations demonstrated disseminated intravascular coagulation. The patient was then transferred from a peripheral hospital to a tertiary centre where further investigations revealed a mediastinal mass, bilateral pleural effusion and ascites. Echocardiography showed a large intracardiac mass in the right ventricle and filling the right atrium originating from the superior vena cava but not from the heart itself. Because of the risk of the bleeding, a biopsy was not attempted, and a presumptive diagnosis of lymphoma was made and the patient was given a single dose of radiation. The patient died 5th day postradiation.

DISCUSSION

Spindle epithelial tumor of thymus like differentiation is a rare tumor primarily affecting young individuals. The youngest patient reported to have this condition was a 2-year-old girl who was presented with a right thyroid nodule.[1] Tong et al reported a case of monophasic SETTLE in a 16-year-old girl on whom no epithelial cells were identified in either fine needle aspiration biopsy or the subsequent hemithyroidectomy specimen.[2] Usually SETTLE has little to no mitotic figures with only focal necrosis. However a case report of 29 year old diagnosed by fine needle aspiration biopsy and histologically confirmed to have SETTLE demonstrated numerous mitotic figures among the spindle cells with focal necrosis.[3] The fine needle aspiration biopsy interpretation may be mistaken with synovial sarcoma, medullary and papillary carcinoma. Su L et al reported a diagnosis of synovial sarcoma for a case which was later proved to be SETTLE by histology.[4] The eldest patient reported to have SETTLE was a 59-year-old man who presented with thyroid enlargement who developed bilateral pulmonary and widespread metastasis.5 Despite development of metastasis in this patient, the patient had long term survival, attesting to the indolent behaviour of the tumor.[5] In the current case both spindle and epithelial cell components were present in the thyroid lesion (Figure 3). At the time of the autopsy, touch preparations were obtained from the mediastinal mass ,revealed the two mentioned components. The spindle cell component had an elongated nuclei, fine chromatin, inconspicuous nucleoli, and scanty cytoplasm as were seen in the touch preparations (Figure 1).

Figure 1

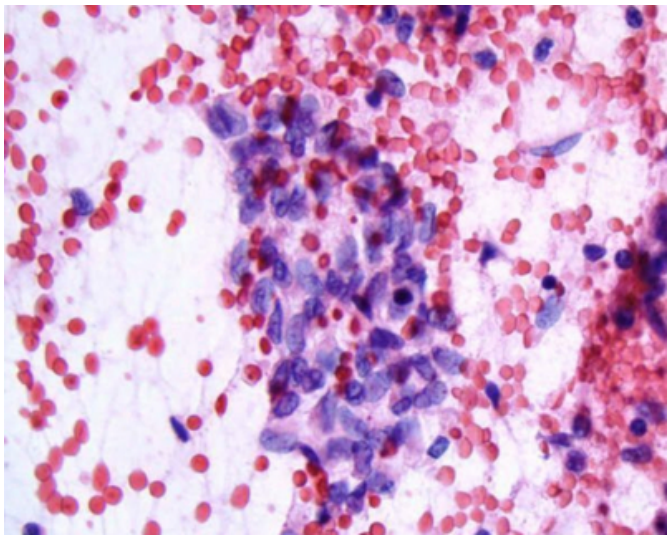
Figure 1: Biphasic cellular population of lesion as demonstrated. Spindle cell component of lesion with small cells with scant cytoplasm and some pleomorphism compatible with spindle cell component of lesion



Rare mitotic figures were noted. The glandular component showed a mixed pattern with tubules, papillae and epithelium lined cystic spaces. The glandular cells were cuboidal to columnar and the nuclei were round to oval (Figure 2).

Figure 2

Figure 2: Epithelial cell component of lesion with syncytial arrangement. The cells are larger than those seen in the spindle cell component. Occasion nuclear grooves are noted (arrows). (x400).



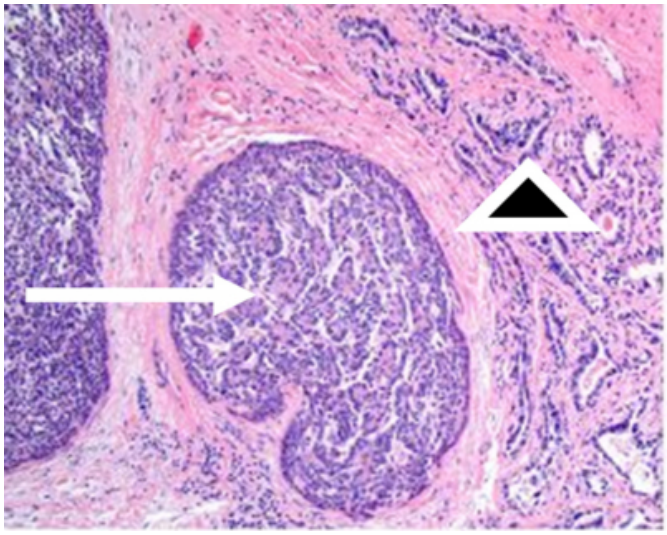
The cytologic features of the epithelial component demonstrated syncytial aggregates of cells with round to oval nuclei with occasional nuclear grooves. The chromatin was fine and nucleoli were inconspicuous (Figure2).

Histology of the tumor confirmed the cytological findings by the presence of

Lobulation imparted by the dense fibrous septae. The tumor was biphasic, composed of spindle cell and glandular epithelial component.(Figure 3).

Figure 3

Figure 3: A biphasic tumor show the glandular (triangle) and spindle cell (arrow) component.



The differential diagnosis includes sarcomatoid anaplastic carcinoma, this is characterized by rapid tumor growth and mortality within 1 year , tumor cells demonstrate overt nuclear atypia, frequent mitosis and necrosis. Medullary carcinoma can have spindle cells, gland like structures, and nuclear grooves (as encountered in this case Figure 1). Tumor cells of medullary carcinoma also have stippled chromatin and granular cytoplasm. Immunostaining for calcitonin and chromogranin are confirmatory for the diagnosis of medullary carcinoma.

SETTLE differs from thymoma as the extensive glandular pattern of SETTLE is not seen in thymoma. Histologically SETTLE lacks the jigsaw puzzle like lobulation, TdT – positive thymocytes, and ultrastructurally interdigitating cell processes. Distinction of SETTLE from synovial sarcoma is very difficult on the basis of cytology alone, but the spindle cells in synovial sarcoma show patchy cytokeratin immunoreactivity and tonofilament are not found ultrastructurally.[5]

CONCLUSION

SETTLE is a rare thyroid tumor. The cytologic and

histological appearance may mimic other thyroid tumors, especially those with a spindle cell morphology. Tumors most likely to be confused with SETTLE are sarcomatoid anaplastic cell carcinoma, medullary carcinoma, spindle cell variant of papillary carcinoma and thymoma. Immunoprofiling and attention to subtle morphologic details are key to correctly typing this rare tumor which appears to have an indolent behaviour.

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