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Case Report

A rare expediting killer-primary thyroid gland sarcoma

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ABSTRACT

Primary thyroid malignancies can be classified into differentiated type (follicular, papillary, medullary, and anaplastic) and un-differentiated type or non-epithelial malignancies, particularly lymphomas and sarcomas. The reported frequency of primary thyroid sarcoma (PTS) cases ranges from 0.01% to 1.5%. Based on this fact, we present a case report of rare primary thyroid sarcoma (PTS) in Hospital Sibu, Sarawak. Our patient is a 78-year-old lady with multiple co-morbidities and her clinical deterioration is so rapid that its progression mimics thyroid anaplastic carcinoma which is commonly seen in female elderly as well. To diagnose such uncommon thyroid malignancy, the histopathology of the tumour needs to be conducted with immunohistochemical staining. Radiographic imaging such as computed tomography or magnetic resonance imaging of whole body is required for staging purpose and to look for metastasis. We analyze the clinical presentations of PTS and discuss its treatment and prognosis. The treatment of PTS must be assessed individually on case-by-case basis as currently there is no general consensus on treating this cancer.

Keywords: Primary thyroid malignancy, Primary thyroid sarcoma

INTRODUCTION

There are four common differentiated types of primary thyroid malignancy which are follicular, papillary, medullary, and anaplastic, while non-epithelial malignancies arising from thyroid can be particularly lymphomas. The reported frequency of primary thyroid sarcoma (PTS) cases is very low, ranges from 0.01% to 1.5%.[1-3]

Based on this fact, we present a case report of rare primary thyroid sarcoma (PTS) in Hospital Sibu, Sarawak. The aim of this study was to analyze clinical presentation of PTS which mimics thyroid anaplastic carcinoma and to discuss its rapid deterioration and prognosis. We believe that treatment of PTS has to be assessed individually on case-by-case basis as currently there is no general consensus on treating PTS.

CASE REPORT

A 79-year-old lady, non-smoker, with underlying hypertension, initially presented with rapidly enlarging, painless right neck mass for 2 months. Otherwise, there was no compressive symptoms, hoarseness, hyper or hypothyroidism symptoms, infective symptoms, tuberculosis symptoms and ear or nasal symptoms. She had no family history of malignancy. On examination, a huge, firm, and diffuse irregular right thyroid nodule was detected, measuring about 10×7 cm (Figure 1). Thyroid function test showed subclinical hyperthyroidism. Thyroid stimulating hormone (TSH) was found to be 0.245 mIU/l (reference range 0.27 to 4.2 mIU/l) and free T4 20.82 pmol/l (reference range 12-22 pmol/l). Ultrasound of the neck revealed a thyroid nodule image reporting and data systems (TI-RADS) bilateral thyroid nodules with lobulated irregular hypoechoic solid mass

occupying almost entire right lobe. Contrast-enhanced computed tomography (CECT) neck showed a large irregular heterogeneously enhancing mass seen in the right thyroid lobe measuring 6.1×4.8×10.2 cm with minimal retrosternal extension. Small left hypodense thyroid nodule.[4]

Trachea is deviated to left. Internal jugular vein thrombosis at proximal and distal segments. CECT thorax noted irregular calcified nodule at apical of right lower lobe measuring 1.1×1.0 cm. Metastasis cannot be excluded. FNA showed many singly dispersed and loosely cohesive malignant cells in the background of evident necrotic debris. The malignant cells feature enlarged, rounded and pleomorphic nuclei with prominent nucleoli and irregular nuclear contour. The cytoplasm is scanty to moderate in amount. Occasional multinucleated tumour cells are seen. No normal follicular cells. FNA findings revealed malignancy that classifies it as Bethesda category.[6]

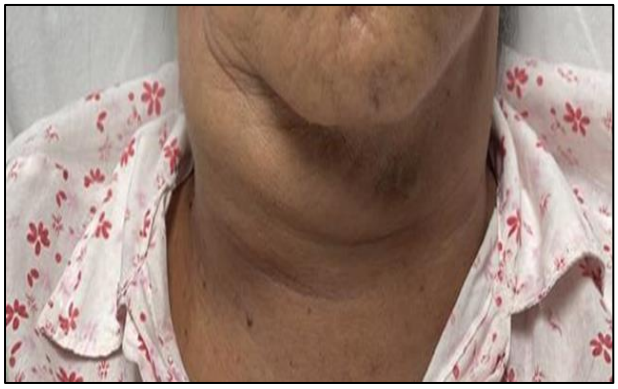

Figure 1: A 79-year-old woman with a rapid-growing neck mass.

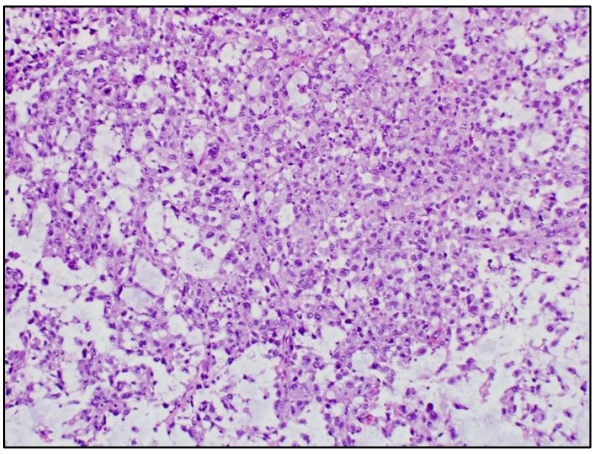

Figure 2: Infiltrative malignant cells with myxoid background (H and E 40X magnification).

Patient was given an appointment in one month time in view of patient not keen for any surgical intervention at that moment. However, she presented again about one month later with aspiration symptoms and respiratory distress. She underwent emergency tracheostomy with tumour debulking for airway protection. Thyroid gland tissues were sent for histopathological examination (HPE). Microscopically, HPE showed infiltrative malignant cells with myxoid background and extensive necrosis (Figure 2). They were round to spindly, displaying moderately pleomorphic and hyperchromatic nuclei with minimal to modest cytoplasm, and disposed in clusters, strands and discohesive single cells (Figure 3). No osteoid or chondroid formation identified. No follicular formation, colloid materials or nuclear features of papillary thyroid carcinoma identified.

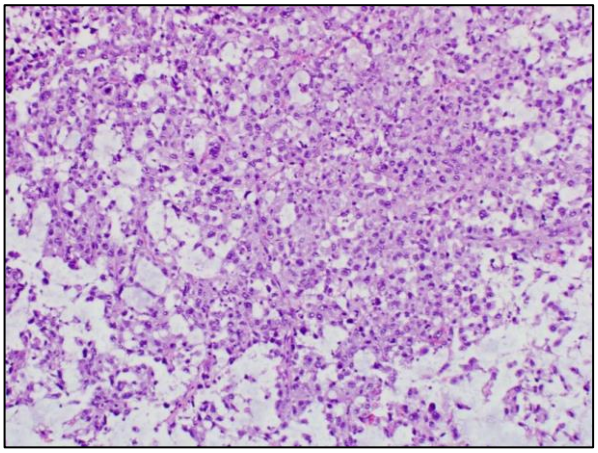

Figure 3: Cells exhibit pleomorphic and hyperchromatic nuclei with modest cytoplasm, and readily seen mitosis. No follicular formation, colloid materials, or nuclear features of papillary thyroid carcinoma identified (H and E 400X magnification).

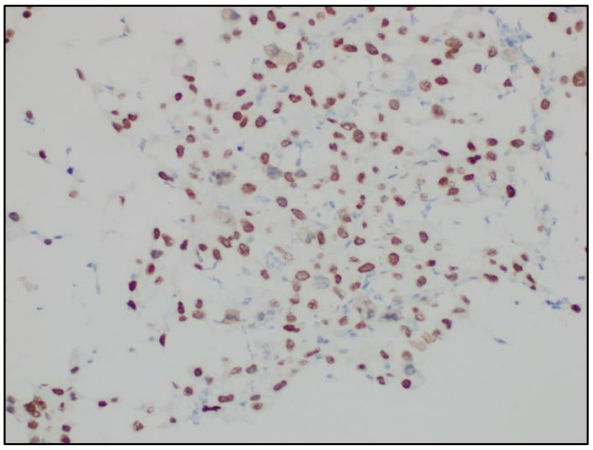

Figure 4: Diffuse positivity for SATB2 (200X magnification).

Immunohistochemical staining revealed the tumour cells were focally positive to smooth muscle actin (SMA) (Figure 4), indicating a smooth muscle origin and diffusely positive for Special AT-rich binding protein 2 (SATB-2) (Figure 5). H3K27me3 showed retain focal nuclear positively. Other tumour markers, CKAE1AE3, CAM5, EMA, PAX-8, TTF-1, Thyroglobulin, Desmin,

Caldesmon, Myogenin, S100, SOX-10, HMB45, LCA and CD34 were negative. Ki-67 proliferation index was 90%.

The overall findings were consistent with a high-grade sarcoma with myxoid background, differentials including high grade myxofibrosarcoma and myxoid pleomorphic liposarcoma. Patient's condition further deteriorating as she developed type II myocardial infarction and hospital acquired infection. Initial plan of surgical removal of thyroid gland and adjunct therapy (chemotherapy and radiotherapy) was unable to be done as patient's condition not stable to be proceeded. Patient succumbed to her disease around 3 months from her first presentation.

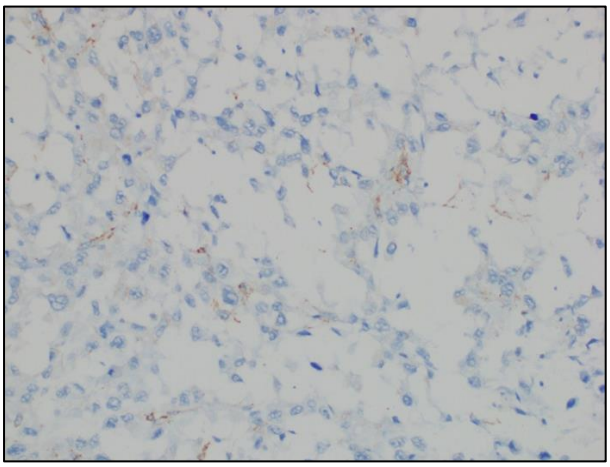

Figure 5: Focal positivity for SMA (200X magnification).

DISCUSSION

Thyroid malignancy is universally classified into differentiated type (follicular, papillary, medullary and anaplastic) and un-differentiated type or non-epithelial malignancies, particularly lymphomas and sarcomas. The reported frequency of primary thyroid sarcoma (PTS) ranges from 0.01% to 1.5%.[1-3] In a systematic review of primary thyroid sarcoma conducted by Alexey et al in 2015, the most frequent five types of thyroid sarcoma are angiosarcoma, malignant hemangioendothelioma, malignant fibrous histiocytoma, leiomyosarcoma, and fibrosarcoma.[4]

In this report, we present a rare case of primary thyroid sarcoma (PTS) in Sibu, Sarawak, Malaysia. The differentials include high grade myxofibrosarcoma (MFS) and myxoid pleomorphic liposarcoma. Our patient is a 78-year-old lady which her age and gender are compatible to the analyzed PTS cases by Alexey et al that there is a slight female and elderly predominance PTS.[4] In addition, it also matches the clinical findings of a case series report published by Gupta et al, which involved three cases of thyroid sarcoma. These three cases were all elderly ladies and the patients presented with a rapidly increasing thyroid mass with tenderness and were found to have lung metastasis with poor clinical outcome. Before diagnosing as primary thyroid sarcomas, it is vital to perform a thorough physical examination to rule out secondary sarcomas involving thyroid gland. Despite the neck swelling in our case, there was no other swelling or deformity noted over the head, larynx, and bilateral limbs. As Alexey et al concluded that 70.4% of patients in their review had no local or distant metastases at the time of diagnosis, it was unusual for our case that lung metastasis with inferior vena cava thrombosis was already detected in the CT thorax, which was only 1 month after patient's initial complaint of neck swelling.[4] An emergency operation (tumor debulking and tracheostomy) was performed due to impending airway collapse and some thyroid gland tissue was sent for histopathological examination (HPE).

Our patient's thyroid HPE showed high grade sarcoma with myxoid background which its differentials include high grade myxofibrosarcoma (MFS) and myxoid pleomorphic liposarcoma. Macroscopically, multiple fragments of tan brownish tissue measuring 45 mm in aggregate diameter were received. The tumor was characterized by infiltrative malignant cells with myxoid background and extensive necrosis, round to spindly displaying moderately pleomorphic and hyperchromatic nuclei, with minimal to modest cytoplasm, and disposed in clusters, strands and discohesive single cells. No osteoid, chondroid formation, follicular formation, colloid materials or nuclear features of papillary thyroid carcinoma was identified. By immunohistochemistry, the tumor cells are focally positive for smooth muscle actin (SMA) which can be expressed in sarcoma cells and diffusely positive for SATB-2. H3K27me3 showed retain focal nuclear positivity with high Ki-67 expression (90%).

High Ki-67 expression was defined as 20% or greater.[5] A higher level of Ki-67 means that the cancer cells are multiplying at a faster rate. Our case has expressed 90% of Ki-67 which correlates with the poor prognosis of this thyroid sarcoma and rapid deterioration of our patient's condition. It took about 3 months from the time of diagnosis to the demise of our patient.

Although the specimen obtained intraoperatively was highly suspicious of highgrade myxofibrosarcoma (MFS) or myxoid pleomorphic liposarcoma, the specimen amount is still inadequate to reach a conclusion among the pathologists. To our knowledge, only 5 cases of primary thyroid MFS have been reported in the English literature so far.[6] The most definite answer is to proceed with total thyroidectomy and send the whole thyroid tumor for HPE but this intervention does not suit our patient whose clinical condition was deteriorating gradually.

Alexey et al, has proposed in his study that there is no general recommendation regarding its therapy, in terms

of surgery to chemo-radiotherapy and follow-up of PTS and hence, it is impossible to compare different treatment approaches in PTS.[4] On the other hands, some literatures recommended surgery as the mainstay treatment while there is still inadequate evidence and clinical trials to study the effect of chemo-radiotherapy.[7] In our case, there was no further surgical intervention. Thyroidectomy or chemotherapy and radiotherapy done as our patient developed type II myocardial infarction and hospital acquired infection which her clinical condition was not optimized to undergo this surgery. Palliative care was initiated and the patient eventually succumbed around 3 months from her first presentation.

CONCLUSION

Primary thyroid sarcoma is a rare neoplasm that need to be differentiated from other non-differentiated thyroid carcinoma such as anaplastic, lymphomas, myxofibrosarcoma (MFS) and myxoid pleomorphic liposarcoma. As the final diagnosis rests on HPE, the treatment of PTS should be assessed on case-by-case basis although surgery is still the priority if patient is fit for it. In a case of clinical deterioration, palliative care should be initiated.

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