Abstract. Papillary thyroid carcinoma with fibromatosis-like stroma (PTC-FLS) is a rare type of thyroid carcinoma that does not receive adequate follow-up. This study describes a case of PTC-FLS on the right side of the neck. The patient consented to a right hemithyroidectomy with a central compartment lymphadenectomy. Pathological examination revealed that the two initial lesions were consistent with benign histological performance. Using immunohistology, the proliferated stromal cells were revealed to be positive for vimentin and smooth muscle actin (SMA), but negative for thyroglobulin, S-100 and CK. Post-operative follow-up was scheduled at 3-month intervals for 2 years and then at 6-month intervals for the next 3 years. There were no signs of tumor recurrence at the 5-year follow-up.

Introduction

Papillary thyroid carcinoma is a common carcinoma. During the past several decades, an increasing incidence of thyroid cancer has been observed in several countries, including China. However, papillary thyroid carcinoma with fibromatosis-like stroma (PTC-FLS) is rare and was first reported in 1832 (1,2). A discerning characteristic of PTC-FLS is the formation of fibroma (nodular tumor composed of stromal cells) without large growth of papillary carcinoma. The histogenesis and clinicopathological features of PTC-FLS have not yet been fully clarified, due to the rarity of the disease (3,4). Further investigation into PTC-FLS is necessary. In the present study, one particular PTC-FLS case was reported, and the diagnosis and management of this tumor was reviewed through assessment of the literature.

Case report

A 42-year-old female patient was admitted to Shaoxing Hospital (Shaoxing, China), due to a mass on the right side of her neck. This mass had gradually enlarged over the previous 7 months. Upon physical examination, a 3-cm firm nodular mass was revealed in the right lobe of her thyroid gland. Serum free tetra- and tri-iodothyronine (FT4 and FT3) and thyroid stimulating hormone (TSH) tests, in addition to routine laboratory examinations (including thyroid function tests) all exhibited normal results. Additionally, thyroid peroxidase antibody (TPOAb), thyroglobulin antibody (TgAb) and thyroid stimulating hormone receptor antibody (TRAb) were at the normal level. A right hemithyroidectomy with a central compartment lymphadenectomy was performed. Post-operative histologic examination revealed a fibroma-like tumor (Fig. 1). Histological examination identified a complex neoplasm in which a minor component of typical papillary carcinoma was intermingled with a dominant and exuberant stromal proliferation. Immunohistology results revealed that the proliferated stromal cells were positive for vimentin and smooth muscle actin (SMA), but negative for thyroglobulin, S-100 and CK. Post-operative follow-up was scheduled at 3-month intervals for 2 years and then at 6-month intervals for the next 3 years. There were no signs of tumor recurrence at the 5-year follow-up.

The study was approved by the Ethics Committee of Shaoxing Hospital, China Medical University, Shaoxing, China. Written informed consent was obtained from the patient prior to the study.

Discussion

Here, we presented a case of PTC with fibromatosis-like stroma. Papillary thyroid carcinomas typically contain fibrous stroma of varying degrees and are occasionally almost completely replaced by hyalinized fibrous tissues with multiple or sole microcalcification. However, an extremely large stromal component in PTC is rarely found, which is a typical feature of PTC-FLS (5). Fibromatosis is a tumor-like lesion that develops from fascia or aponeurosis of fibrous tissue. It occurs mainly in the shoulder, chest and back, but rarely in the thyroid. The etiology of PTC-FLS is unknown;
Therefore, this mass may be and vinblastine or
be easily misdiagnosed as a malignant tumor (7). Although
contrast-enhanced CT scans reveal the rich blood supply
in thyroid fibromatosis, this is ineffective in differentiating
thyroid fibromatosis from thyroid cancer. The definitive
diagnosis of PTC-FLS may be determined by examining an
intra-operative frozen section and a post-operative paraffin
section. PTC-FLS must be treated surgically as a low-grade
malignant tumor. As the recurrence rate for local excision is
20-45%, complete excision needs to be performed (9). In
certain cases, when tumors invade nerves and significant
organs, it is difficult to perform this procedure. Invaded
organs have a greater impact on the patient’s quality of
life, rendering complete surgery a contradictory solution.
However, we suggest that complete surgery should be
performed if possible.

For patients who do not wish to undergo surgery or cannot
afford the operation, local excision in addition to radiation
therapy may be an effective treatment. Radiation therapy is
more suitable, as the recurrence rate is reduced with a radia-
tion dose of 50-60 Gy (10). However, there are also numerous
studies demonstrating the ineffectiveness of radiotherapy (10).
A number of systemic therapies have been studied for inoper-
able cases of fibromatosis, such as antiestrogens; nonsteroidal
anti-inflammatory drugs (NSAIDs), including indomethacin,
sulindac and cyclo-oxygenase-2 (COX-2) inhibitors; interfer-
ons and various other drugs, including methotrexate in
combination with doxorubicin, ifosfamide and vinblastine or
vinorelbine. In addition, more recently, target therapy with
imatinib has been applied in platelet-derived growth factor
receptor-beta (PDGFR-β)-positive tumors. Chemotherapy with
a low-dose of methotrexate and vinblastine (or its less
neurotoxic analog, vinorelbine) on a weekly basis has been
demonstrated to achieve a 70% relapse-free survival rate at
10 years (11). Likewise, indomethacin has been demonstrated
to be effective in certain cases of fibromatosis (12). Traditional
Chinese medicine may also be effective in this disease.

The prognosis of this tumor depends on its size and
location. Typically, the 10-year survival rate is 94% and the
20-year survival rate is 86% (11). However, for patients with
aggressive fibromatosis involving vital organs or large blood
vessels, the survival rate is lower.

References

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