Chula Med J Vol. 46 No. 4 April 2002

Angiosarcoma of the thyroid gland : a case report

Mana Taweevisit*

Thamathron Assanasen*

Taweevisit M, Assanasen T, Angiosarcoma of the thyroid gland : a case report. Chula Med J 2002 Apr; 46(4): 333 - 9

This is a case of angiosarcoma of unknown etiology of the thyroid gland, the incidence of which is rare outside the European alpine region. The patient experienced enlarged neck mass for three months without any history of trauma. She received right lobectomy and postoperative radiotherapy. Gross specimen reveals spongelike filled with blood accompanied by extensive necrosis. This feature suggests vascular neoplasm. Histology, moreover, demonstrates numerous vascular channels lined by pleomorphic cells and loss of polarity. The immunoperoxidase technique was used to support the existence of neoplasm of the thyroid gland with endothelial differentiation.

Key words : *Angiosarcoma, Thyroid gland.*

Reprint request : Taweevisit M, Department of Pathology, Faculty of Medicine, Chulalongkorn University, Bangkok 10330, Thailand.

Received for publication. January 10, 2002.

* Department of Pathology, Faculty of Medicine, Chulalongkorn University

มานะ ทวีวิศิษฏ์, ธรรมธร อาศนะเสน. มะเร็งหลอดเลือดของต่อมไทรอยด์: รายงานผู้ป่วย 1 ราย. จุฬาลงกรณ์เวชสาร 2545 เม.ย; 46(4): 333 - 9

สาเหตุของมะเร็งหลอดเลือดของต่อมไทรอยด์นั้นยังไม่ทราบแน่ชัด มะเร็งชนิดชนิดนี้พบได้บ่อยเฉพาะแถบเทือกเขาแอลป์ในทวีปยุโรปเท่านั้น ส่วนประเทศอื่น ๆ พบอุบัติการณ์น้อยมาก ผู้ป่วยมีประวัติคอโตเร็วมากภายในเวลา 3 เดือน และเข้ารับการผ่าตัดร่วมกับรังสีรักษา เมื่อมองก้อนมะเร็งด้วยตาเปล่า ลักษณะจะมีรูพรุนคล้ายฟองน้ำ เต็มไปด้วยเลือดและเนื้อตายจำนวนมาก ซึ่งเป็นลักษณะเด่นของมะเร็งหลอดเลือด การวินิจฉัยด้วยกระบวนการทางจุลพยาธิวิทยาสามารถแสดงได้ว่าหลอดเลือดที่เพิ่มขึ้นอย่างมากมายนั้นถูกบุด้วยเซลล์มะเร็งที่มีขนาดและรูปร่างต่าง ๆ กัน และจัดเรียงตัวอย่างไม่เป็นระเบียบ ไม่มีทิศทางแน่นอน อิมมิวโนฮีสโตเคมียืนยันพยาธิสภาพอย่างแน่ชัดว่าก้อนมะเร็งมีต้นกำเนิดมาจากผนังหลอดเลือด

คำสำคัญ : *มะเร็งหลอดเลือด, ต่อมไทรอยด์*

Angiosarcoma of the thyroid gland is extremely a rare disease.[1] It is only a common neoplasia in the European alpine regions, else where it is almost unknown.[2,3] Because of its rarity, the tumor is not included in most reports about thyroid cancer, but some possibility in a differential diagnosis of anaplastic thyroid carcinoma.[4,5]

We report a case of primary angiosarcoma of the thyroid gland, who was treated by lobectomy and postoperative radiotherapy, at King Chulalongkorn Memorial Hospital. The purpose of the report is to show the gross and histo-pathological features of the sarcoma, which was confirmed by immuno-histochemical study. The data also provides a further evidence that angiosarcoma of the thyroid gland is a distinct entity of endothelium in origin that differs from anaplastic thyroid carcinoma.

Case report

The patient was a 56 – year - old woman with a history of thyroid mass for eight months. She denied any history of symptoms associated with hyperthyroidism. About three months before her hospitalization, the mass grew rapidly. A physician gave her tablets of medication, but there was no improvement. Her physical examination revealed a well-defined mass in the right lobe of her thyroid gland, measured 10 x 15 cm. The mass, in addition, was fixed, firm, and smooth. Neither bruit nor tenderness was detected. The smear of a fine needle aspiration (FNA) exhibited colloid without evidence of malignancy. The patient was treated with a right lobectomy of the gland. The gross specimen consisted of a huge ovoid mass, measured 10.5 x 8.0 x 6.5 cm and weighed 220 grams. Its cut surfaces showed extensive areas of necrosis with hemorrhage, resulted from blood-filled cystic cavities resembling hematomas in the mass (Fig. 1). The thyroid parenchyma was observed at the periphery of the mass. The thyroid capsule was intact. Histologically, there was proliferation of neoplastic cells arranged in irregular vascular channels, infiltrating the thyroid parenchyma (Fig. 2). There was a tendency of these vascular channels to communicate with each other, forming anastomosing networks of sinusoids that contained numerous erythrocytes. Some forms of neoplastic-lined papillae were noted focally (Fig.3). Stromal fibrous connective tissue was increased with the presence of mature collagen. The neoplastic cells possessed enlarged pleomorphic vesicular nuclei, prominent nucleoli with marked mitoses and moderate to abundant pale acidophilic cytoplasm (Fig.4), containing some eosinophilic hyaline globules. Some of tumor cells adopted epithelioid appearance with cytoplasmic cavities containing red blood cells. The immuno-histochemistry was done, using antibodies of factor VIII-related antigen (Fig. 5), vimentin, cytokeratin (KB), epithelial membrane antigen (EMA), and thyroglobulin. The malignant cells reacted to factor VIII-related antigen and vimentin but was negative to cytokeratin (KB), epithelial membrane antigen (EMA), and thyroglobulin. The adjacent thyroid parenchyma also exhibited nodular goiter. A few lymph nodes taken had no metastasis. She was readmitted a month later because of a hematoma of her thyroid gland. The hematoma measured 8 x 10 cm, replaced the previous lesion that mentioned above. An amount of clotted blood was also removed. The patient underwent adjuvant postoperative local radiotherapy.

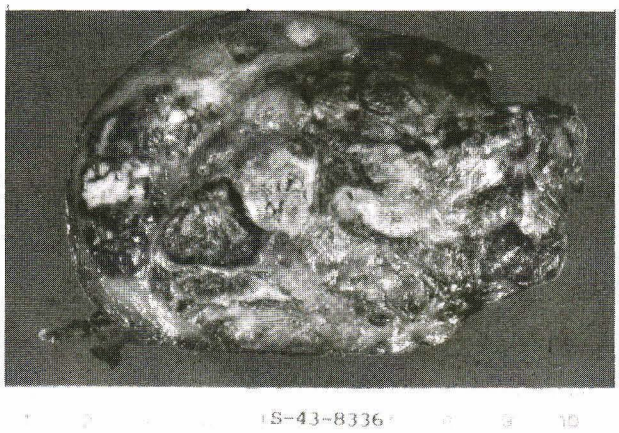

Figure 1. This tumor is characteristically necrotic and hemorrhagic.

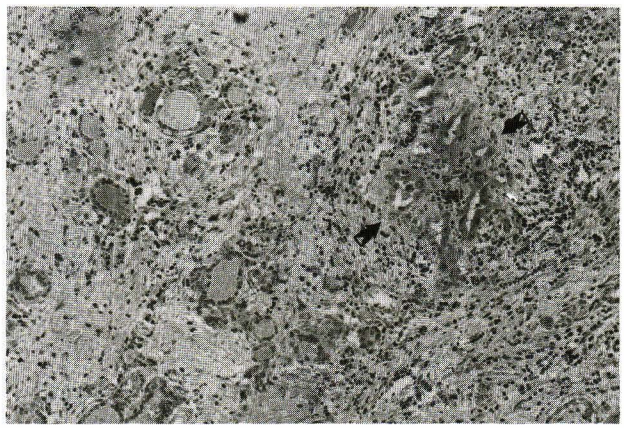

Figure 2. Angiosarcoma (arrows) infiltrating thyroid parenchyma. (H&E stain x 40)

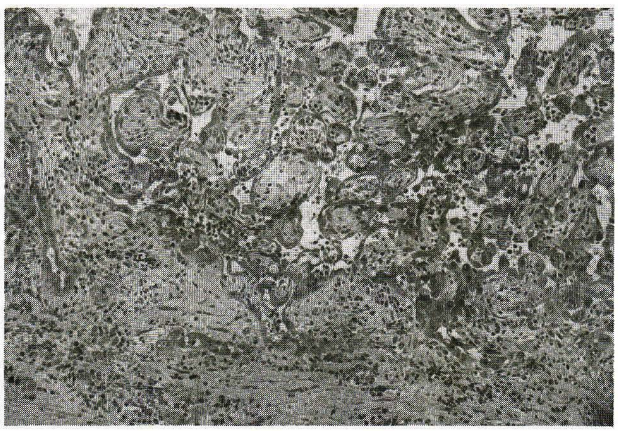

Figure 3. Foci of clear-cut anastomosing vascular channel formation with focal papillary growth of angiosarcoma. (H&E stain x 40)

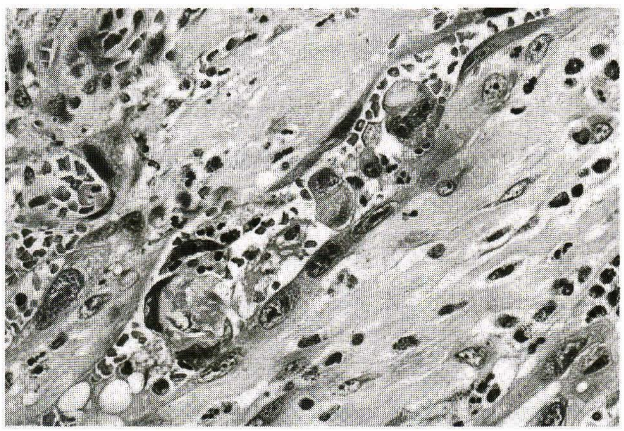

Figure 4. This high magnification shows the large vesicular nucleus and prominent irregularly shaped nucleolus that are typical of this tumor type. Abortive vascular lumina can be seen. (H&E stain x 400)

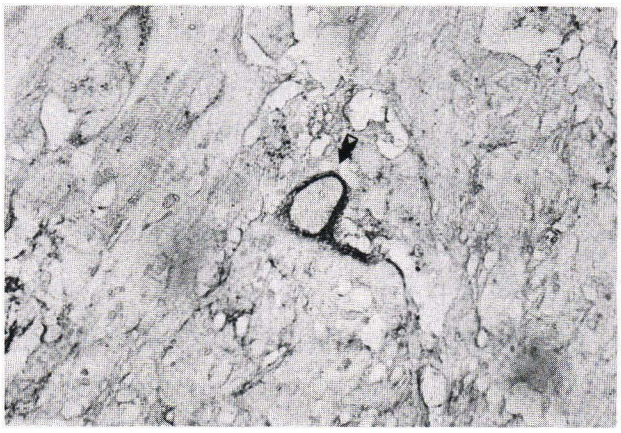

Figure 5. The plump epitheioid cells are strongly positive for factor VIII-related antigen (arrow). (H&E stain x400)

Discussion

Angiosarcoma or malignant hemagio-endothelioma, an extreme spectrum of endothelial differentiation, is the rarest form of soft tissue neoplasm, especially in the thyroid gland.[1,6] Its biological behavior is locally aggressive and destructive, well-known for its widespread metastases and high rate of recurrent.[7] Angiosarcoma of the thyroid gland occurs in the elderly; the majority of patients are older than 60 years. Its female: male sex ratio ranges between 1 and 1.9.[5] Most patients are from the Alpine zone with endemic goiters (approximately 16 % of thyroid malignancies).[2,3] Its incidence in Western Austria is 0.15 to 0.25 per 100,000 population per year.[1] In the United States, reports of the lesion are extremely scarce.[2] In Asia, there is only one reported document, about the disease in a Chinese patient.[5]

Its cause remains unknown.[5] In contrast the other site, there is no evidence to support that prior neck irradiation or contact with carcinogen, such as thorotrast or vinyl chloride are the risk factors of angiosarcoma of the thyroid gland.[5,8] The gross appearance of the tumor is often a huge mass exhibiting sponge-like filled with blood admixed with large areas of so-called ruberry hyaline.[1,5]

The microscopic evidence of angiosarcoma is the presence of anatomosing vascular channels with cleft-like spaces lined by atypical calls in the tumor.[1,5,8,9] The clue may be limited only to focal areas, while most of the lesion is arranged in poorly differentiated epithelial-like or fascicular pattern.[5] In addition, the complicated lesion can also discloses small areas of angiosarcoma-like within anaplastic carcinoma. Distinctions between angiomatoid thyroid carcinoma and true thyroid angiosarcomas are necessary.[4,10-12] Some authors believe that the tumors may be variant forms of undifferentiated carcinoma.[12] Therefore, the existence of this form is definitively confirmed to support the endothelial origin by recent immuno-histochemical analyses. [13]

The immuno-histochemical methods confirm our case; they revealed only positive results for factor vimentin and VIII-related antigen, found throughout the tumor. The presence of these markers suggested that the tumor was demonstrating endothelial differentiation.[13,14] The epithelial membrane antigen (EMA), and cytokeratin were markers of a wide variety of epithelial cells while the presence of thyroglobulin suggested follicular or papillary differentiation of thyroid carcinoma.[2,14]

However, the smear of FNA biopsy only contained colloid, the storage site of iodinated

hormones, without parenchymal cell. This evidence strongly shows that the submitted specimen is not the representative of the whole lesion. Indeed, angiosarcoma shows clusters of pleomorphic cells, concentrically arranged, or whorl patterns.[15]

The treatment was similar to extrathyroid carcinoma enclosing of radical surgical excision and postoperative radiotherapy, because a complete removal of the growth is impossible.[1,5] The prognosis was very poor due to its rapid growth and early metastases.[7] The tumor was sarcoma in nature. It, hence, usually metastasizes through venous and capillary channels; but, rarely affects regional lymph nodes.[16] The small series of reports might indicate that the outcomes of the disease might not be uniformly deleterious.[17] The resistance to radiotherapy was also reported. The mean survival time was approximately 39 months.[17]

In this case, the patient suffered hematoma of her neck about one month after surgery but this condition usually occurs within the first few hours after operation because of bleeding from branches of the superior or inferior thyroid artery.[18] Furthermore, delayed bleeding is not illustrated in most literatures. So this finding is rather unsual and may cause by other factors; however, the removed hematoma was not submitted for pathological examination.

References

1. Enzinger FM, Weiss SW. Soft Tissue Tumor. 3rd ed. St. Louis: Mosby,1995: 641 - 58
2. Mills SE, Stallings RG, Austin MB. Angiomatoid carcinoma of the thyroid gland. Anaplastic carcinoma with follicular and medullary features mimicking angiosarcoma. Am J Clin Pathol 1986 Nov; 86(5): 674 - 8
3. Tanda F, Massarelli G, Bosincu L, Cossu A. Angiosarcoma of the thyroid : a light, electron microscopic and histoimmunological study. Hum Pathol 1988 Jun; 19(6): 742 - 5.
4. Lamovec J, Zidar A, Zidanik B. Epithelioid angiosarcoma of the thyroid gland. Report of two cases. Arch Pathol Lab Med 1994 Jun;118 (6): 642 - 6
5. Chan YE, Ma L, Boey JH, Yeung HY. Angiosarcoma of the thyroid. An immunohistochemical and ultrastructural study of a case in a Chinese patient. Cancer 1986 Jun 15; 57(12): 2381 - 8
6. Astl J, Duskova J, Limanova Z, Povysil C, Kuchynkova Z. Hemangiosarcoma of the Thyroid Gland. A case report. Neuroendocrinol Lett 2000; 21(3): 213 - 6
7. Mills SE, Gaffey MJ, Watts JC, Swanson PE, Wick MR, LiVolsi VA, Nappi O,. Weiss LM. Angiomatoid carcinoma and 'angiosarcoma' of the thyroid gland. A spectrum of endothelial differentiation. Am J Clin Pathol 1994 Sep;102 (3): 322 - 30
8. Schoen FJ. Blood vessels. In : Robbins Pahologic Basis of Disease. Robbins CK,ed. Vol. 5th ed. Philadelphia: W.B.Saunders, 1994: 510 - 11
9. Meis-Kindblom IM, Enzinger FM. Color Atlas of Soft Tissue Tumor. Philadelphia, 1996: 164 - 7
10. Ritter JH, Mills SE, Nappi O, Wick MR. Angiosarcoma-like neoplasms of epithelial organs: true endothelial tumors or variants of carcinoma?. Semin Diagn Pathol 1995 Aug; 12(3): 270 - 82
11. Halalau F, Laky D. Hemangiosarcoma of the thyroid gland. Histological diagnosis problems.

Morphol Embryol (Bucur) 1983 Jul-Sep; 29(3); 195 - 6

12. Cutlan RT, Greer JE, Wong FS, Eltorky M. Immunohistochemical characterization of thyroid gland angiomatoid tumors. Exp Mol Pathol 2000 Oct; 69(2): 159 - 64
13. Neri A, Aldovini D, Leonardi E, Giampiccolo M, Pedrolli C. Primary angiosarcoma of the thyroid gland. Presentation of a clinical case. Recenti Prog Med 1990 May; 81(5): 318 - 21
14. Colvin RB, Bhan AK, McCluskey RT. Diagnostic immunopathology. 2nd ed. New York: Raven Press,1995: 455 - 70
15. DeMay RM. Soft tissue tumors. In: DeMay RM. The Art and Science of Cytopathology: Vol 2. 6th ed. Hong Kong; 1996: 583
16. Lieberman MW, Lebovitz RM. Neoplasia. In: Danyanov I, Linder T, ed. Anderson's Pathology. 10th ed. St.Louis: Mosby, 1996: 523
17. Rhomberg W, Gruber-Mosenbacher U, Eiter H, Fritzsche H, Breitfellner G. Prongnosis and epidemiology of malignant hemangioendotheliomas of the thyroid gland. Schweiz Med Wochenschr 1993 Sep 4; 123(35):1640 - 4
18. Schwaztz SI, Shizes GT, Spencer FC. Thyroid and Parathyroid. In: Principles of Surgery: Vol. 2. 6th ed. McGraw-Hill Inc; 1994: 1643