Rare disease

Primary angiosarcoma of the thyroid gland in an young Iranian woman

Fariba Binesh,¹ Ali Akhavan,² Hossein Navabii,³ Mohammad Hossein Dadgarnia,⁴ Vahid Zand⁵

¹Department of Pathology, Yazd Shahid Sadoghi University, Yazd, Islamic Republic of Iran;

²Department of Radiotherapy, Yazd Shahid Sadoghi University, Yazd, Islamic Republic of Iran;

³Shahid Sadoghi University of Medical Science, Yazd, Islamic Republic of Iran;

⁴Department of ENT, Yazd Shahid Sadoghi University, Yazd, Islamic Republic of Iran;

⁵Yazd Shahid Sadoghi University, Yazd, Islamic Republic of Iran

Correspondence to Dr Fariba Binesh, binesh44@yahoo.com

Summary

Angiosarcoma of the thyroid gland is a rare and aggressive primary malignant tumour of the thyroid, originally reported in patient from Swiss Alpine region. Here, the authors describe a case of primary angiosarcoma of the thyroid in an 21-year-old Iranian woman who presented with a rapidly enlarging neck mass associated with compressive symptoms.

BACKGROUND

Angiosarcoma is one of the rarest form of soft tissue neoplasms, comprising less than 1% of all sarcomas.¹ Angiosarcoma has been reported in the literature as primary tumour in various anatomic sites including breast,^{2 3} thyroid,⁴ heart⁵ and lung.⁶ Angiosarcoma of the thyroid is a primary malignant tumour in the thyroid showing evidence of endothelial differentiation.⁷ It is a rare tumour and represents 2–10% of thyroid malignancies.⁸ Angiosarcoma of the thyroid was originally described by Swiss authors as early as 90 years ago in patients from Alpine region.^{9–11} However, there have been cases from non-Alpine regions in the literature. Here, we describe a case of primary angiosarcoma of the thyroid in an 21-year-old Iranian female who presented with a rapidly enlarging neck mass associated with compressive symptoms.

CASE PRESENTATION

An 21-year-old Iranian female developed rapid enlargement of the neck over a period of 8 months associated with dysphonia and dysphagia in the last few weeks prior to admission. Physical examination revealed a neck mass. The overlying skin was violaceous in colour. The mentioned mass was anterior and more prominent on the right side. It was pulsatile, fixed and there was leakage of blood from its surface (figure 1). There was no evidence of cervical lymphadenopathy. There was no other medical (such as thyroid dysfunction) or surgical history.

INVESTIGATIONS

The results of preoperative blood investigation including full blood count, renal function test, clotting profile and thyroid function were within normal limits. Direct laryngoscopy showed decreased motion of the right vocal cord. CT-scan of the neck revealed a large vascular mass arising from thyroid gland (figure 2). CT scan of the neck showed that there was some mass effect with displacement of the airway (figure 3). There was no obvious sign

of retrosternal extension. Chest x-ray films showed clear lung field. The patient underwent surgery. There was a haemorrhagic cystic lesion arising from the thyroid, extending beyond the confines of the thyroid capsule. The surgery was very difficult and incisional biopsy was done. Since the patient had no respiratory symptoms, insertion of a stent was not considered. On microscopic examination, sections revealed anastomosing angiomatous structures formed by large fusiform epithelioid cells with a vesicular nucleous and large eosinophilic cytoplasm showing partially solid islet structures, also extensive haemorrhage and necrosis were observed (figure 4). Based on these findings, the case was diagnosed as having thyroid angiosarcoma.

DIFFERENTIAL DIAGNOSIS

Clinically the differential diagnosis of angiosarcoma of thyroid includes a variety of entities including undifferentiated thyroid carcinoma, approximately 2–5% of all thyroid carcinoma are undifferentiated carcinoma. But the vast majority of patients are older people. Most patients present with a rapidly growing neck mass. In addition, the less differentiated angiosarcoma may resemble anaplastic thyroid carcinoma, however, anaplastic carcinoma shows abortive follicles and thyroglobin immunoreactivity. Riedle thyroiditis is a densely sclerotic inflammatory process which causes the thyroid gland to become adherent to adjacent structures. However, atypical mitosis, abnormal vascular spaces and the presence of marked cellular pleomorphism are not present. Microscopically, the well-differentiated angiosarcoma may be quite similar to pseudoangiosarcomatous changes in thyroid follicular epithelium after the needle aspiration injury and reactive endothelial hyperplasia occurring spontaneously in goitre, but there is however a tendency for the channel in angiosarcoma to anastomose with one another and to produce dilated sinusoids, and also has a more infiltrative dissecting pattern at its interface with the normal surrounding tissues.

Figure 1 Gross photograph shows large neck mass with violaceous overlying skin.

Figure 2 CT-angio (sagittal plane) showing large vascular mass arising from thyroid gland.

TREATMENT

In our case, because this hypervascular tumour was encased the great vessels and trachea, en bloc resection was not possible. On the other hand, because of numerous

small arteries that supply the tumour, arterial embolisation was also impossible. Inevitably the patient received chemotherapy by using adriamycin (20 mg/m^2) and ifosfamide (2500 mg/m^2), both of them for three consecutive

Figure 3 CT scan (axial plane) showing large mass which displaced airway.

days that repeated every 3 weeks for three courses. It resulted in partial improvement and decrease of oozing, but the response was incomplete. According to patient's symptoms and incomplete response, local radiotherapy started with cobalt 60 machine, by using anterior/posterior and posterior/anterior parallel opposed fields and 180 cGY daily dose. The treatment continued until 2600 cGY. But the disease course was complicated by extensive tumour necrosis and abscess formation, which was successfully

treated with antibiotics. Subsequently, she refused radiotherapy and discharged.

OUTCOME AND FOLLOW-UP

Several weeks after discharge, she developed progressive dysphagia, dysphonia and stridor. CT-scan showed progression of the disease and therefore failed radiotherapy. Her condition worsened and she died 4 months after diagnosis.

Figure 4 Photomicrograph shows anastomosing angiomatous structures & large fusiform epithelioid cells showing partially papillary structures (H&E ×10).

DISCUSSION

Angiosarcoma may occur in any location in the body, they rarely arise from major vessels. In contrast to the deep location of most soft tissue sarcomas, angiosarcoma have a predilection for skin and are usually found on the scalps of older people. Primary angiosarcoma is a rare tumour of the thyroid gland, but it is the most common form of the thyroid sarcoma and occurs primarily in the endemic goitre regions of Europe.¹² The mean age of affected patients is 60 years and there is a slight male preponderance.¹² In contrast, the presented case was a 21-year-old Iranian woman. Primary thyroid angiosarcoma usually arises from a long standing nodular goitre.⁴ Endothelial proliferation after recurrent intranodular haemorrhages occurring in a long standing nodular goitre have been considered to initiate neoplastic transformation.¹³ Our case had no history of nodular goitre and her thyroid function tests were within normal limits. Thyroid angiosarcoma is more common in the Alpine region or Europe and its incidence is about 2–10%.⁸ Cases are sporadic in other regions of the world and the exact incidence is unknown.⁹ This case is the first Iranian case which has been described. The histopathologic appearances of this neoplasm varies greatly. The well-differentiated lesion may be quite similar to pseudoangiosarcomatous changes in thyroid follicular epithelium after the needle aspiration injury and reactive endothelial hyperplasia occurring spontaneously in goitre. The less differentiated angiosarcoma may resemble anaplastic thyroid carcinoma, however, anaplastic carcinoma shows abortive follicles and thyroglobulin immunoreactivity. Surgery is the cornerstone of treatment in all subtypes of soft tissue sarcoma but head and neck sarcomas are not

amenable to wide local excision with generous margins of normal tissue due to anatomic constraints. The use of adjuvant radiation is more liberal in this site because local recurrence can be the cause of death.¹⁴ Surgery alone or radiation therapy with or without chemotherapy may yield dismal results. Mark *et al* reported that only 2 of 19 patients (10.5%) who received a single modality treatment well alive at the time of their review, and all 19 had a component of local failure.¹⁵ Therefore, the consensus is to use a combined modality approach including excision of disease with negative margins, if possible with adjuvant radiotherapy with or without chemotherapy but despite the different treatment modalities available, prognosis is generally poor. In our case, radical surgery was impossible because of tumour extension. The patient received radiotherapy and initially responded partially, but then tumour progressed and finally she died.

Learning points

- ▶ Thyroid angiosarcoma is a distinct entity and should be considered and included as a differential diagnosis of poorly differentiated thyroid neoplasms.
- ▶ This has been incidentally diagnosed in regions outside its typical geographical location.
- ▶ This soft tissue neck tumour may complicate postoperative management due to their bleeding tendency and aggressive infiltrative behaviour, and carry a dismal prognosis.

Competing interests None.

Patient consent Obtained.

REFERENCES

1. **Weiss SW**, Goldblum JR. Malignant vascular tumors. In: Weiss SW, Goldblum JR, eds. *Enzinger and Weiss's Soft Tissue Tumors*. Fourth edition. St Louis, MO: Mosby 2008:703.
2. **Donnell RM**, Rosen PP, Lieberman PH, *et al*. Angiosarcoma and other vascular tumors of the breast. *Am J Surg Pathol* 1981;**5**:629–42.
3. **Macías-Martínez V**, Murrieta-Tiburcio L, Molina-Cárdenas H, *et al*. Epithelioid angiosarcoma of the breast. Clinicopathological, immunohistochemical, and ultrastructural study of a case. *Am J Surg Pathol* 1997;**21**:599–604.
4. **Astl J**, Dusková J, Límanová Z, *et al*. Hemangiosarcoma of the thyroid gland. A case report. *Neuro Endocrinol Lett* 2000;**21**:213–16.
5. **Wakely PE Jr**. Angiosarcoma of the heart in an adolescent. A light and electron microscopic and immunohistochemical study. *Arch Pathol Lab Med* 1987;**111**:472–5.
6. **Sheppard MN**, Hansell DM, Du Bois RM, *et al*. Primary epithelioid angiosarcoma of the lung presenting as pulmonary hemorrhage. *Hum Pathol* 1997;**28**:383–5.
7. **Eusebi V**. Angiosarcoma. In: DeLellis RA, Lloyd RV, Heitz PU, Eng C, eds. *World Health Organisation Classification of Tumours. Pathology and Genetics Tumours of Endocrine Organs*. Lyon: IARC Press 2004:113–14.
8. **Ryska A**, Ludvíková M, Szépe P, *et al*. Epithelioid haemangiosarcoma of the thyroid gland. Report of six cases from a non-Alpine region. *Histopathology* 2004;**44**:40–6.
9. **Maiorana A**, Collina G, Cesinaro AM, *et al*. Epithelioid angiosarcoma of the thyroid. Clinicopathological analysis of seven cases from non-Alpine areas. *Virchows Arch* 1996;**429**:131–7.
10. **Lin O**, Gerhard R, Coelho Siqueira SA, *et al*. Cytologic findings of epithelioid angiosarcoma of the thyroid. A case report. *Acta Cytol* 2002;**46**:767–71.
11. **Mills SE**, Gaffey MJ, Watts JC, *et al*. Angiomatoid carcinoma and 'angiosarcoma' of the thyroid gland. A spectrum of endothelial differentiation. *Am J Clin Pathol* 1994;**102**:322–30.
12. **Egloff B**. The hemangioendothelioma of the thyroid. *Virchows Arch A Pathol Anat Histopathol* 1983;**400**:119–42.
13. **Eusebi V**, Carcangiu ML, Dina R, *et al*. Keratin-positive epithelioid angiosarcoma of the thyroid: a report of four cases. *Am J Surg Pathol* 1990;**14**:737–47.
14. **Brennan MF**, Singer S, Maki RG, *et al*. Sarcomas of the soft tissue and bone. DeVita VT, Lawrence TS, Rosenberg SA, Weinberg RA, DePinho RA, eds. *DeVita, Hellman & Rosenberg's Cancer: Principles & Practice of Oncology*. Eighth edition. Philadelphia, PA: Lippincott Williams & Wilkins 2008:1778.
15. **Mark RJ**, Tran LM, Sercarz J, *et al*. Angiosarcoma of the head and neck. The UCLA experience 1955 through 1990. *Arch Otolaryngol Head Neck Surg* 1993;**119**:973–8.

This pdf has been created automatically from the final edited text and images.

Copyright 2011 BMJ Publishing Group. All rights reserved. For permission to reuse any of this content visit <http://group.bmj.com/group/rights-licensing/permissions>.

BMJ Case Report Fellows may re-use this article for personal use and teaching without any further permission.

Please cite this article as follows (you will need to access the article online to obtain the date of publication).

Binesh F, Akhavan A, Navabii H, Dadgarnia MH, Zand V. Primary angiosarcoma of the thyroid gland in a young Iranian woman. *BMJ Case Reports* 2011; 10.1136/bcr.03.2011.4042, date of publication

Become a Fellow of BMJ Case Reports today and you can:

- Submit as many cases as you like
- Enjoy fast sympathetic peer review and rapid publication of accepted articles
- Access all the published articles
- Re-use any of the published material for personal use and teaching without further permission

For information on Institutional Fellowships contact consortiasales@bmjgroup.com

Visit casereports.bmj.com for more articles like this and to become a Fellow