A case of thyroid angiosarcoma

Clinical History:

A 74-year-old man, living in Modena in a non-Alpine area, with a history of goitre for 20 years reported increasing dyspnoea, initially only under stress and later even at rest.

Imaging Findings:

The general practitioner prescribed a chest radiograph (Fig. 1), which showed a right mediastinal mass dislocating and compressing the trachea to the opposite side. The CT of the thorax demonstrated an enlarged right thyroid lobe with inhomogeneous enhancement, low density nodules and calcifications (Fig. 2).

After one month the right lobe of the thyroid was completely surgically extirpated. Histological specimen revealed a thyroid angiosarcoma reaching the stripe of resection, CD31+/CD34+.

After two weeks from surgery the patient reported thoracic pain. Chest radiograph showed again a right mediastinal mass dislocating the trachea (Fig. 3). CT showed a big hematoma (Fig. 4), with quite similar characteristics to the removed lesion. CT also revealed multiple suspicious pulmonary nodules (Fig. 5) and bone marrow lesions (Fig. 6). These findings were confirmed as hypermetabolic at PET-CT (Fig. 7).

A month later the patient presented with a pathological fracture of the right shoulder blade (Fig. 8, 9).

Discussion:

Thyroid sarcomas are malignant tumours of mesenchymal derivation. Angiosarcoma of the thyroid is a very rare entity almost unknown in most parts of the world except in European Alpine regions [1, 2, 3], where it represents about 16% of thyroid malignancies in association with iodine-deficient goitre [4]. Few cases without previous goitre have been reported [5].

Usually the lesion begins as a painless enlarging mass, poorly encapsulated and with infiltrative growth [1, 6]. The tumour is typically large with extensive areas of necrosis and hemorrhage, mimicking a hematoma. This type of tumour typically metastasizes to bone marrow and lungs [7].

Because of the nonspecific aspects at imaging studies (inhomogeneous mass with calcifications, areas of necrosis and hemorrhage) the differential diagnosis with goitre and other thyroid tumours is difficult. Moreover also after extensive surgery the occurrence of hemorrhage can resemble a very early recurrence. Either biopsy or complete resection of the tumour with a detailed histological investigation is usually the only way to make a diagnosis [8]. Histological specimen reveals freely joined channels lined by typical endothelial cells associated with a papillary configuration, that is due to a predominantly intraluminal growth pattern. The nuclei of the epithelioid endothelial cells are often large, vesicular and endowed with a large basophilic or amphophylic nucleolus connected by
chromatin strands to the nuclear membrane. Mitoses, typical and atypical, are found in large numbers. The pattern of growth is nearly always highly invasive and tumour necrosis is prominent. The tumour cells express vascular markers such as Factor VIII, CD 31 and CD 34 [2, 9, 10, 11]. The tumour is locally aggressive and has a destructive behaviour with high recurrence rate. The treatment of choice is radical surgery but if the tumour cannot be treated by surgery, then radiotherapy and eventually chemotherapy is indicated [5]. The prognosis is not favourable. Data on survival are dispersed but Goh et al. reported average 5-years survival of 33.3% [4].

**Differential Diagnosis List:** Thyroid angiosarcoma with pulmonary and bone marrow metastasis., Goitre, Thyroid adenoma, Thyroid cyst, Thyroiditis, Thyroid papillary carcinoma, Thyroid follicular carcinoma, Thyroid anaplastic carcinoma, Thyroid medullar carcinoma, Thyroid lymphoma, Thyroid sarcoma

**Final Diagnosis:** Thyroid angiosarcoma with pulmonary and bone marrow metastasis.

**References:**


Goh SG, Chuah KL, Goh HK, Chen YY (2003) Two cases of epithelioid angiosarcoma involving the thyroid and a brief review of non-Alpine epithelioid angiosarcoma of the thyroid. Arch Pathos Lab Med127:E70–E73


Description: The chest radiograph shows right mediastinal mass with maximum diameter of 8 cm, dislocating and compressing the trachea to the opposite side. Origin:
Description: The chest radiograph shows right mediastinal mass with maximum diameter of 8 cm.
Origin:
Description: The arterial phase of dynamic CT of the torax demonstrated an enlarged right thyroid lobe with inhomogeneous enhancement, low density nodules and calcifications, dislocating and compressing the trachea to the opposite side. Origin:

Description: The venous phase demonstrated the poor enhancement of the lesion. Origin:
Description: CT multiplanar coronal reformat of the thorax showing craniocaudal extension of the mass and its relationships to the main mediastinal structures. Origin:
Description: The chest radiograph 2 weeks after surgery showed a right mediastinal mass dislocating the trachea to the opposite side. Origin:
**Description:** The chest radiograph 2 weeks after surgery showed a right mediastinal mass. **Origin:**
Figure 4

a

Description: The CT 2 weeks after surgery showed a large hematoma in right thyroid lodge that has similar characteristics to the removed lesion. Origin:

b

Description: CT multiplanar coronal reformat showing a big hematoma following surgery with almost similar characteristics to the previous removed lesion. Origin:
Figure 5

Description: Lung windowing of the pre-surgery CT shows no pulmonary nodules - especially in the lower right lobe. Micronodular thickening of the interstitium in left lower lobe is appreciable. Origin:
Description: Lung windowing of post surgery CT (2 weeks interval) revealed multiple pulmonary nodules. Origin:
Figure 6

a

Description: The pre-surgery CT showed no bone marrow lesion at the level of the left shoulder.

b

Description: The CT performed 2 weeks after surgery revealed an osteolytic bone marrow lesion at the left scapula.
Description: The PET-CT examination showing hyper metabolic pulmonary lesions. Origin:
Description: The PET-CT examination confirmed bone marrow lesions. In the image a right iliac bone lesion. Origin:
Description: The PET-CT examination confirmed bone marrow (shoulder) and pulmonary lesions.
Origin:
Description: The PET-CT examination confirmed bone marrow lesions. In the image a right anterior iliac bone lesion. Origin:
Description: The radiograph showed a pathological fracture of the left shoulder-blade. Origin:
Figure 9

Description: CT confirmed the fracture of the left shoulder-blade. Origin:

Description: Multiplanar reconstruction confirms the pathological fracture of the left shoulder-blade. Origin: