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CASE REPORT

Hemangiosarcoma of the Thyroid Gland. A case report

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Abstract

OBJECTIVES: The authors described a case of rare histological type of primary thyroid tumor—angiosarcoma. Angiosarcoma generally represents a rare connective tissue tumor in the head and neck region. Its biological behavior is locally aggressive and destructive with a high recurrence rate.

METHODS: A case of a rare histological type of primary thyroid tumor —angiosarcoma—has been described including fine needle aspiration biopsy, histology, and autopsy.

RESULTS: Irrespective of the complex approach the angiosarcoma of the patient referred to was diagnosed in a radically inoperative stage of the disease. Radiation was therefore applied after the tumor mass was diminished with the partial thyroidectomy and a correct histopathological diagnosis was rendered. Nevertheless, the locally destructive course was followed with a rather rapid spread of the tumor (lymph nodes, bones, lungs, and brain). Autopsy proved with certainty that the thyroid was the primary tumor location. A suspected kidney origin due to an uncertain sonography shadow of this region was excluded with a complete absence of any neoplastic spread of the angiosarcoma in the abdominal cavity at all.

Introduction

About twelve percent of soft-tissue sarcomas occur in the head and neck region [15]. Angiosarcoma is a rare type of sarcoma. Among other possible locations it occurs rather often in the head and neck region. In Pack and Ariel's study [13] seven from twenty cases and in Das Gupta's study [2] three from seventeen described angiosarcomas had this origin. The location in the thyroid gland is nevertheless quite exceptional [8, 11, 12]. On the other hand, the only well-documented sarcoma that does arise in the thyroid is either the angiosarcoma or the hemangioendothelioma [1, 4, 5, 9, 10, 14].

Biopsy or even complete removal of the tumor with a detailed histological study of the tissue is usually the only way to establish the sometimes difficult diagnosis.

Falk remarked that this lesion has been most commonly described from the Alpine regions of Europe [6].

Angiogenic sarcomas comprise malignant angioendothelioma, hemangioendothelioma and lymphangiosarcoma. These tumors are made up of malignant endothelial structures with a common fibrosarcomatous stroma and may contain both capillary and lymphatic elements [1, 10, 11].

Most angiogenic sarcomas begin as a painless, slowly enlarging firm mass, poorly encapsulated and infiltrative. They recur frequently at the site of excision and may be spread by blood to multiple distant organs. Regional node metastases also sometimes occur. Radiation therapy is used as a palliative measure [15].

Case report

A 68 year-old woman observed for seven months a tumor mass growing on the neck in the jugular fossa, i.e. in the thyroid region. During the last four months she lost about 15 kilograms of her body weight. For the last three weeks she suffered from a dyspnea associated with an upper-airways inflammation. The neck circumference was 46 centimeters when she visited the Otolaryngological Department.

On the neck inspection the tumor mass was predominantly visible in the jugular region without the skin involvement. Palpation proved a firm tumor associated with the thyroid gland tissue. Auscultation did not reveal a major overlaying vessel and whirring on auscultation was discovered.

The examination of the oral cavity, pharynx, larynx, salivary gland, skin of the scalp, face and neck excluded primary cancer of any of these locations.

The lung X-ray investigation and breast superior aperture proved right site trachea deviation and on the C7 level a trachea stenosis up to 50%. The enlargement of the upper mediastinum was also discovered.

On the CT-scans the tumor mass was centered on the left thyroid lobe with the trachea stenosis and spread into the upper mediastinum.

The fine needel aspiartion biopsy (FNAB) was performed (C128/97). Several smears stained MGG, alcian blue, oil red, and PAS were investigated. In the blood background were dyscohesive groups of large atypical cells with a variable nucleocytoplasmic ratio. The cytoplasm was mostly circumscribed, vacuolized in some, and there was irregular chromatin distribution and striking large nucleoli. Cytoplasm of the cells was PAS and oil red positive, thyreoglobulin immunocytochemistry negative. The cytology conclusion: the tumor was less differentiated, more probably of the metastatic origin than primary. In relation to the clinical information about a sonography shadow diam. 3 cm in the renal pelvis, this origin for additional staining results (PAS, oil red positivity) was considered possible.

As the patient had a trachea stenosis and progress of swelling with dyspnea problems surgery was indicated for liberation of the air ways. A tumor mass on the left kidney was discovered and investigated at the same time.

The tumor markers were CEA 0.1 mg /l, SCC 1,O mg /l GMT O, l mg /l, AST 0.8 mkat/l and ALT O, 7 mkat/l, ALP 3.7 mkat/l, CRP 185 mmg/l. Transferin 1.38 g/l, the thyroid hormones level was next T3 1.59 nmol/l, T4 O,87 nmol/l, TSH 2.10mIU/l.

The liver ultrasonography was normal—free of focal lesions.

Total left hemithyroidectomy was performed. The surgical intervention was complicated with a left vocal cord paralysis.

The tumor had not spread over the thyroid lobe capsula.

The histological investigation (B 3195/97): a thyroid lobe 100x90x70mm, nearly all substituted with a grey to yellowish focally necrotic neoplastic tissue of a rather firm consistence. Microscopically a poorly differentiated highly desmoplastic polymorphocellular tumor with foci of possible epithelial cohesion. Other parts contained spindle cells. The nuclei were large, pale with striking nucleoli. Focally necroses, old hemorrhages and hemosiderophages were present. PAS and oil red staining were both positive as fine granules in the neoplastic cells cytoplasm, while alcian blue only slightly in the intersticium. The immunohistochemistry for Thyreoglobulin and Calcitonin was performed with DAB visualization at first,

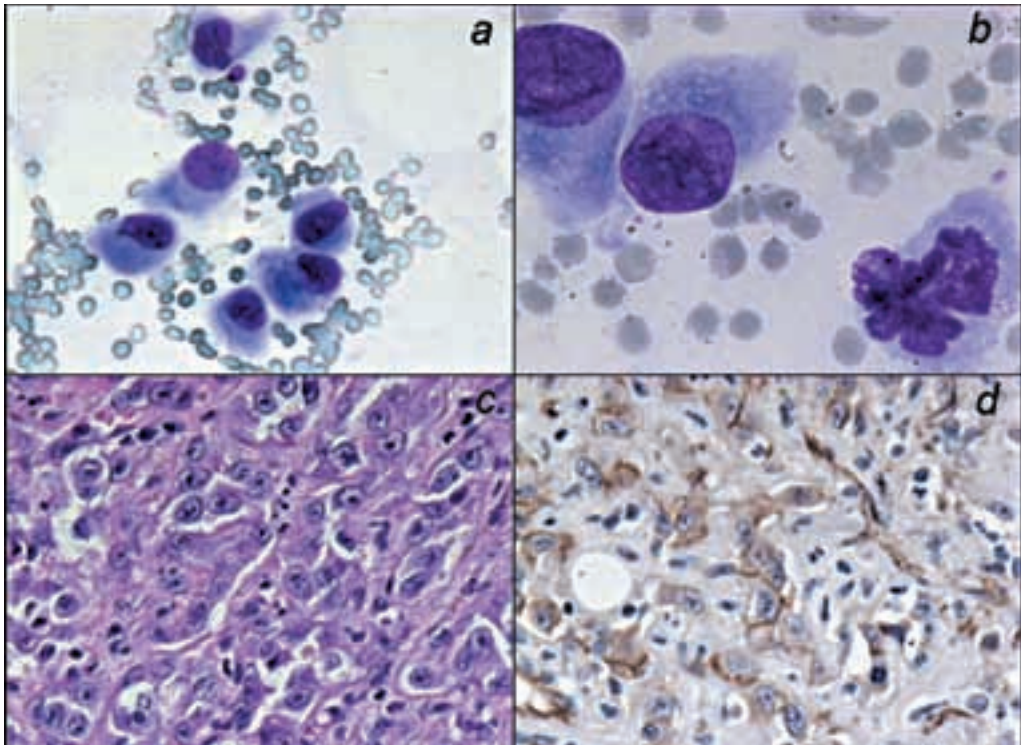

Fig. 1.
a - Fine Needle Aspiration Biopsy. Dyscohesive malignant cells among erytrocytes. May - Grünwald - Giemsa, 40x.
b - Fine Needle Aspiration Biopsy. Cellular details. May - Grünwald - Giemsa, 100x.
c - Histology. Solid tumor architecture. Hematoxylin - Eosin, 20x.
d - Immunohistochemistry. CD31 positivity. 20x.
(All images were acquired with the microscope Nikon Microphot FXA, color RGB video camera JVC TK 1070E, and LUCIA image analysis software.)

then repeated with immunophophatase reaction and blue visualization due to the uncertain product interference with the dispersely present hemosiderin. Both Thyreoglobulin and Calcitonin were negative. The epithelial markers (CAM 5,2, AE1-3, CEA were positive on some of the neoplastic cells. S100 protein was slightly positive as well. Vascular markers CD34 and CD31 were positive. For the immunohistochemistry results— concurrent epithelial and vascular marker positivity of the tumor—an electron microscopy investigation was performed using the formaline fixed and paraffin embedded material. No Weibel-Pallade bodies or microvillous cell surface was found. The cell junctions were of the tight junction type; real desmosomes were not present. Summarizing the results of all investigations, the diagnosis of hemangiosarcoma was favored. The thyroid was declared as a possible primary tumor site, but metastatic involvement could not be excluded either.

During the two months following the thyroidectomy, the scare swelling and respiratory problems appeared. The tumor spread widely in the neck muscles and involved the glottis. It was not radically operable any more. X-ray scans were indicative of pulmonary metastases and a suspicious neoplastic shadow in the left kidney. Actinotherapy with Co 60 was tried but the tumor progression was not slowed down. The patient died two months after surgery (nine month after the first symptoms were discovered).

Autopsy (N392/97) was performed two days after death. A tumor generalization was found involving the thyroid rests (right lobe, isthmus, left side residual tissue), neck muscles, larynx up to the glottis, and cervical lymph nodes. The metastatic spread involved the pericardium, parietal pleura, lungs, left cerebellar hemisphere, ribs, and small nodules were in the subcutis in the mammary region. No neoplastic infiltration was found in the abdominal cavity. Both kidneys were symmetrical, each of 150 gm weight, with only small cortical scars from arteriosclerotic nephrosclerosis. Other pathological findings included advanced aortic and coronary atherosclerosis, slight cardiac hypertrophy with the terminal signs of acute failure only. The cause of death was angiosarcoma generalization.

Discussion

Angiosarcomas are in general rare tumors [3, 5, 7, and 15]. Involvement of the head and neck region is

therefore usually reported in the literature. A locally aggressive behavior with an extension for long distance before the first metastases appear is characteristic together with a rather rapid clinical course. Most patients who are not cured with an initial treatment of unadvanced tumor die within three years. The recommended treatment starts with radical excision if possible. Radiation therapy may be effective in some patients and can be completed using Adriamycin [5].

Hemangiosarcoma has a local recurrence rate of about 36%. Metastases appear in 81% of the cases. The tumor causes death in 79% of patients in the first 5 years after the diagnosis [5, 15].

The diagnosis is mostly difficult; from FNAB nearly never completely conclusive. A thorough histological investigation including immunohistochemistry and electron microscopy.

The first symptoms are usually caused by the local spread of the tumor. The symptoms such as pain, dyspnea or dysphagia depend on the angiosarcoma location. Progression into the mediastinum with compressive complications has also been described.

The therapy is a challenge with frequent difficulties. Surgery represents the first step of choice in cases of radically removable tumor. When the tumor is not resectable, radiation must be considered, whereas the chemotherapeutical approach is not yet well established. Raaf 1992 [15] recommended tumor dose ranges from 40.8 to 61.2 Grays in five fractions per week. These data are in accordance with our experience.

In our patient the radical surgery following subtotal thyroidectomy was impossible. An alternative radiotherapy and chemotherapy was applied, unfortunately without a desired tumor response. Our patient died within two months after the diagnosis and therapy effort.

Conclusions

A case of a rare histological type of primary thyroid tumor—angiosarcoma—has been described including fine needle aspiration biopsy, histology, and autopsy.

Angiosarcoma represents a rare connective tissue tumor in the head and neck region. Its biological behavior is locally aggressive and destructive with a high recurrence rate.

Irrespective of the complex approach the angiosarcoma of the patient referred to was diagnosed in a radically inoperable stage of the disease. Radiation was therefore applied after the tumor mass was diminished with the partial thyroidectomy and a correct histopathological diagnosis was rendered. Nevertheless, the locally destructive course was followed with a rather rapid spread of the tumor (lymph nodes, bones, lungs, and brain). The autopsy proved with certainty that the thyroid was the primary tumor location. A suspected kidney origin due to an uncertain sonography shadow of this region was excluded with a complete absence of any neoplastic spread of the angiosarcoma in the abdominal cavity at all.

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