CLINICAL CASE

CERVICAL FIBROSARCOMA ASSOCIATED WITH PAPILLARY THYROID CARCINOMA – COMMENTS ON A CLINICAL CASE

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Abstract

Sarcomas are malignant tumours rarely found in adults, having in the head and neck a percentage of about 1% of all malignant tumours found at this level. They evolve both from bone tissue and from soft tissue, being differentiated depending on the mesenchymal cell from which they derive. Fibrosarcoma can evolve anywhere in the head and neck, the most common locations being represented by the cervical region and the paranasal sinuses.In this article, the authors present a rare case by the clinical association of two histopathological entities in the case of a male patient, aged 71. The surgical resection of two left laterocervical formations was performed, the histopathological result and the immunohistochemical tests setting the diagnosis of 2nd stage fibrosarcoma and papillary thyroid carcinoma, for which the patient was under a complex specialized cancer treatment.

Keywords*: cervical fibrosarcoma, papillary thyroid carcinoma, cervical surgery*

Introduction

Fibrosarcoma is a mesenchymal malignant tumor. This is composed of a malignant proliferation of fibroblasts with a variable amount of collagen and it classically features an architecture of "herringbone" type [1].

Fibrosarcoma comprises a percentage of 1 to 3% of soft tissue sarcomas, these in turn representing a group of neoplasms with mesodermal origin, which constitutes about 1% of the cancers of the adults and respectively 7% of the paediatric malignancies [2].

In the adult it is found between the third and sixth decade of age, and the locations of choice are represented by the extremities (especially thighs and forearms) and torso [3, 4].

They are unusual tumours for the head and neck, being found in these areas only in 0.05% of all fibrosarcomas. Of the risk factors with a possible involvement in the etiology of these neoplasias we mention the radiation exposure followed by trauma, diseases such as Paget's disease, fibrous dysplasia or chronic osteomyelitis. Fibrosarcomas can evolve both intraosseously and in the soft tissues [5].

In the field of ENT, after the paranasal sinuses, the second favourite location is the neck region. It appears as a painless tumor formation of hard consistency. It has a low rate of lymphatic metastasis, but a high percentage, of up to 50% of local recurrence, despite the radical surgical excision. The prognosis depends on the histopathological framing degree of the fibrosarcoma and on the patient's age, 5-year

survival being between 50% and 75%, possibly higher in the case of children [6-10].

Papillary thyroid carcinoma is the most common neoplasm of the thyroid gland, being incriminated in up to 80% of thyroid cancers. The highest points of occurrence of this neoplasia are met in the 3rd or 4th decade of life, most often in women, in a ratio ranging between 1.6 to 1 and 3 to 1 [11]. Papillary thyroid carcinoma received its name due to the histological characteristics of papilliform growth, but it can also show a follicular growth pattern (follicular variant of papillary carcinoma, a subtype that was once presented as follicular carcinoma) [12].

Thyroid cancers and particularly papillary carcinoma are found in patients with a history of irradiation. This neoplasia generally shows poor symptoms or it may be distinguished as a mass or an asymptomatic thyroid nodule. Of the symptoms found in this pathology we mention neck pain, dyspnea or dysphagia, stridor, paralysis of vocal cords, hemoptyses [13].

Lymphatic metastasis in the cervical ganglia is commonly found (in more than 50% of cases). Generally, papillary thyroid carcinoma shows a low growth pattern and it has a high prognosis depending on tumor size (excellent prognosis in sizes below 1.5 cm), with a 10-year survival rate ranging between 80% and 90% [14].

Case presentation

The authors present the case of a male patient aged 71, who was admitted to the clinic for the deformation of the left laterocervical region by the occurrence of a tumor formation about 6 months ago, accompanied by pharyngeal paresthesia and intermittent dysphagia. Of the pathologic personal antecedents of the patient we mention essential hypertension stage II, type II diabetes without complications and dyslipidemia.

The ENT clinical examination revealed the presence of a left laterocervical tumor formation measuring approximately 45/25 mm, oval shape, of hard consistency, slightly mobile on superficial plans and immobile on deep plans, superjacent teguments free.

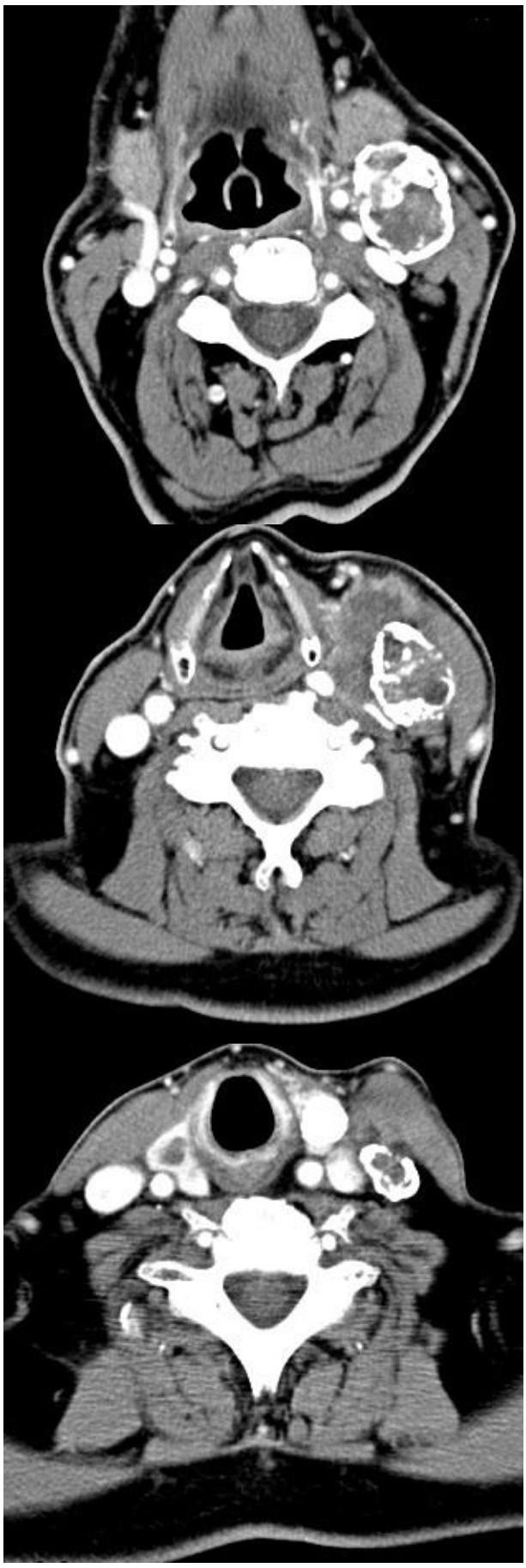

Figure 1- Preoperative cervical computed-tomography, axial section. Left laterocervical masses

The nasopharyngolaryngeal fiberscopic examination highlighted mild hypertrophy of the base of tongue and lingual tonsil bilaterally, infantile epiglottis, in omega, laryngeal asymmetry by rotation of the larynx to the right, vocal cords of normal appearance, hypomobility in the left hemilarynx, sufficient glottis space and free pyriform sinuses.

Laboratory tests did not show significant changes.

Cervical CT examination with contrast-enhancing agent highlighted the presence of an inhomogeneous infiltrate of 40/23 mm, left laterocervical, at the level of a plan passing through the upper edge of the thyroid cartilage, which showed a deleted boundary in relation to the medial face of the left sternocleidomastoid muscle and the left jugular vein, a spontaneously hypodense nodular image, hypocaptating, of 1 cm, in the right thyroid lobe, and laterocervical adenopathies non-calcified bilaterally, with maximum dimensions of 15 mm on the left side, and 19 mm on the right side. (Figure 1).

The surgical intervention – cervicotomy is decided, with biopsy and ablation purpose, succeeding in the ablation of two tumor formations, namely:

- The first left laterocervical tumor formation that infiltrates and thromboses the internal jugular vein, excised en bloc with this;
- The second left laterocervical formation located paratracheal.

The surgical intervention showed a high degree of difficulty. There were required various elements of surgical technique and tactics for identifying the anatomical structures of the region, the dissection being performed with difficulty due to the presence of multiple adherences. There have been highlighted the important vascular and nerve structures of the region – cervical neurovascular bundle, hypoglossal and spinal nerves. The main element of anatomic and surgical risk was represented by the common carotid artery and its branches, the first tumor showing multiple adherences in the artery, being necessary a thorough dissection, millimeter by millimeter, including under the operating microscope, succeeding in its complete detachment from this level. (Figures 2-5).

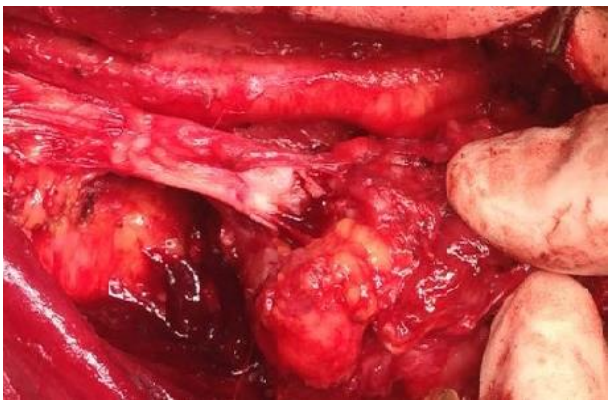

Figure 2 - Intraoperative aspect. Note the tumoral

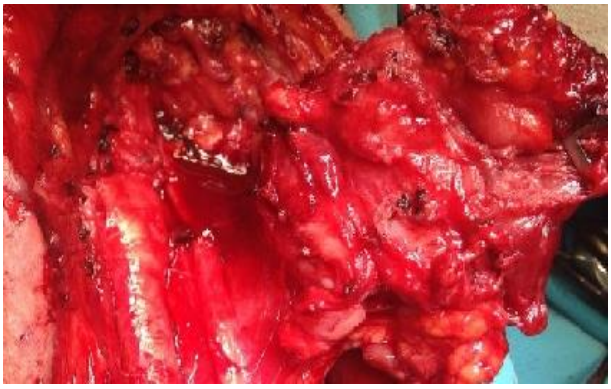

Figure 3 - Intraoperative aspect. Difficult disection mass that infiltrates and contain the left jugular vein from the common carotid artery

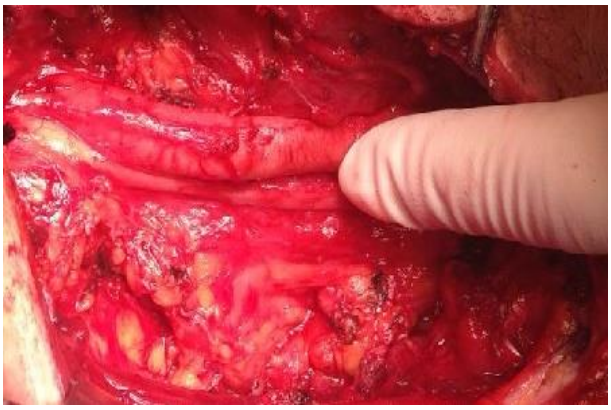

Figure 4 - Intraoperative aspect after resection

Excised pieces were sent to the pathologic anatomy laboratory which set the diagnosis of stage II moderately differentiated fibrosarcoma in the case of the first tumor and of papillary thyroid carcinoma in the case of the second tumoral formation.

The immunohistochemical tests performed confirmed the histopathological aspect and the immunophenotype specific to stage 2 fibrosarcoma, FNCLCC score 5 (differentiation 3, mitotic index 1, necrosis 1), respectively to papillary thyroid carcinoma.

Following the results obtained, the patient is directed to the oncology and endocrinology services for further investigations and appropriate treatment.

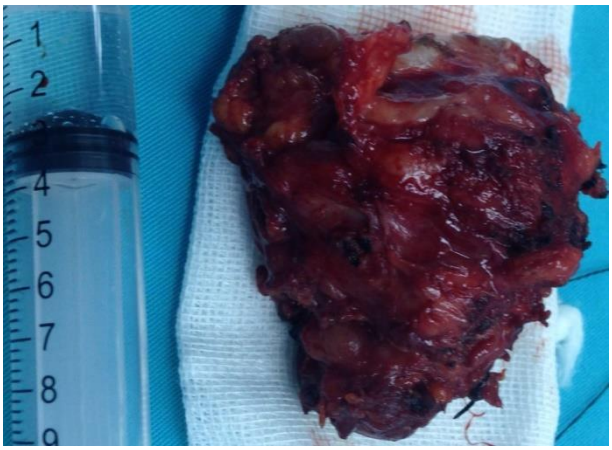

Figure 5 - The first laterocervical tumor that of the tumor was sent for histopathology examination

Thyroid hormone assessment showed eutiroidism, ATPO and calcitonin within normal limits. Ultrasound of the anterior cervical region showed in the middle third of the right thyroid lobe a mixed node of 13/8 mm, hypoechogenyc and transonic, discreetly vascularized peripherally, and in the left lobe, in the upper third, an imprecisely demarcated area of about 9/9 mm, that associates powdery microcalcifications, and an arciform coarse calcification in the lower part. The cervical-thoracic CT imaging examination with contrast-enhancing agent performed post surgery also certified the presence of a right, hypodense thyroid nodule.

After the clinical examination and the laboratory investigations, the patient underwent a new surgery about 2 and a half months after the first intervention, in which it was performed total thyroidectomy in a general surgery clinic. The histopathological result of the excised piece concluded the structure of papillary carcinoma, the follicular variant, in the lower half of the left thyroid lobe, focally encapsulated.

The complex oncological assessment led to the adoption of a treatment plan encompassing both neoplastic entities that this patient had presented. Thus, the patient received external cervico-supraclavicular radiotherapy – 50Gy and chemotherapy treatment with monocarboxylate carriers was initiated, initially dacarbazine, to which subsequently doxorubicin was added.

At the moment, the patient continues the chemotherapy treatment, this showing good tolerance both in the case of radiotherapy and of chemotherapy.

Discussions

Sarcomas are rare tumors comprising 1% of all neoplasias. These are classified histologically according to the tissue from which it derives, in the literature being described more than 30 subtypes. About 80% of sarcomas arise in the soft tissues and 20% from bone tissue [15]. Fibrosarcomas cover a range between 1% and 3% of all sarcomas and are rarely found in the head and neck, where the cervical region is the second location in terms of frequency, after the paranasal sinuses [2, 5]. This neoplasia affects the patients within an extended age range, that varies in literature, but most authors attest the occurrence between 30 and 60 years of age, also existing an infantile subtype that occurs in children younger than 5 years of age and which is associated with a better prognosis than in the case of adults. The exact causes of occurrence of the fibrosarcoma are not known, but the risk factors involved in its presence in the head and neck are represented by external causes such as radiation therapy or local scars and history of burns in its occurrence area [16], phenomena not expressed by the presented patient. In case of damage of soft tissues, there is no predilection regarding the sex to which it occurrs, but there are studies that attest a predominance of males [17-19].

From a clinical point of view, fibrosarcomas show nonspecific symptoms, even poor, often manifesting as a painless tumor formation which gradually grows in volume. The symptoms associated with this growth vary depending on the tumor location and they comprise pain, paraesthesias, swelling and even ulcerations. On the computed tomography they may look homogeneous and without contrast medium uptake, and on the MRI they show a low or moderate signal both in T1 and in T2. The treatment of choice is the surgical one and consists of the complete excision, by observing the principles of oncological surgery, and

postoperative radiotherapy is indicated in the case of positive resection edges. With an unfavorable prognosis are fibrosarcomas with high histological degree (G1 – low degree, respectively G3 – high degree), tumor size over 5 cm, excision edges invaded by the tumor, invasion of the bone tissue, of the skin or neurovascular tissue. Metastasizing is taking place hematogenously. Local recurrence is found in up to 50% of cases, despite the radical surgical excision. Remote metastasizing occurs in 20-40% of cases and mainly affects the lungs. 5-year survival rate is between 50-75% of cases, some authors showing even lower values, located between 20-35% [5], and the recurrences appear frequently in the first two years after the initial treatment. In the case presented by us, the tumor formation excised has shown an intermediate degree of malignancy, namely G2, and the invasion of the internal jugular vein, multiple adherences to the adjacent structures may indicate an unfavorable prognosis of this patient, who, besides this aggressive neoplasia, also has another tumor formation in the thyroid, with the diagnosis of papillary thyroid carcinoma.

Papillary thyroid carcinoma is the most common neoplasia of the thyroid gland, with a frequency which reaches up to 80% of these. It predominantly affects females, in a ratio of up to 3:1, with the highest point of occurrence depending on age comprised between the 3rd and 4th decade of life [11]. As in the case of fibrosarcoma, we find irradiation as a risk factor (85% up to 90% have a history of exposure to low doses of radiation) [20]. Clinically, although the symptoms are nonspecific, they are noisier than in the case of fibrosarcoma, especially in advanced cases of disease and includes cervical pain, dyspnea, dysphagia, stridor, paralysis of vocal cords, hemoptyses [13]. Very often, papillary thyroid carcinoma metastasizes in the cervical ganglia in a ratio ranging between 50% and 80%, and in 80% of cases it also affects, microscopically, the contralateral thyroid lobe at the time of surgery, although recurrence occurs in it and in the cervical ganglia in less than 10% of cases [21]. This pathology has a slow growth pattern and is associated with a very good prognosis, with a 10-year survival rate between 80% and 90% [14]. Due to its slow growth over time, patients with papillary thyroid carcinoma respond well to treatment and prolonged survival, regardless of recurrence, has led to controversies regarding the extension of thyroidectomy in patients with well differentiated carcinoma [22]. Surgical treatment remains though the treatment of choice, the surgical options being total thyroidectomy or lobectomy. Quasi-total or total thyroidectomy is recommended following clinical and laboratory or biopsy certification of the disease in both lobes. Patients who have performed partial thyroidectomy showed an increased risk of local recurrence and cervical or lung metastasising [23]. Therapeutic management depends on tumor staging and may comprise, besides surgery, which remains the treatment of choice, the administration of 131I after surgery (it lowers recurrence rates), external radiation therapy if the tumor does not take 131I, thyroid hormone suppression and chemotherapy in the diffuse forms that do not show indications for other therapies (cytostatic of choice in monotherapy being doxorubicin). Mortality increases with the occurrence of complications such as local recurrence or remote metastasizing, but it also depends on the patient's age, which, if it exceeds 40 years of age, is associated with a more reserved prognosis [20].

Conclusions

This complex case represents a challenge in terms of diagnosis and therapy due to the association of a rare cancer located in the neck and with reserved prognosis, with a more common thyroid neoplasia and which has a better prognosis. Both pathologies showed advanced stages of progression and raise problems regarding response to treatment and long term prognosis. It could not be demonstrated a causal relationship in the occurrence of these cancers and their clinically concomitant evolution and literature data have not succeeded to certify a similar case either so far. The main therapeutic means in both pathologies was surgical treatment, following that the adjuvant cancer treatment contributes to the locoregional and overall control of the disease.

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