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Case Report Open Access

Pleomorphic Sarcoma of Larynx: A Case Report

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Abstract

Malignant mesenchymal neoplasms affecting larynx are a rare tumors. Undifferentiated high-grade Pleomorphic Sarcoma (UPS), considered as malignant fibrous histiocytoma, is a high grade malignant neoplasm characterized by tumor cells with diffuse pleomorphism in the absence of a specific line of differentiation; we present a new case of undifferentiated high-grade pleomorphic sarcoma arising in larynx, and discuss pathological findings and surgical treatment of this rare tumor.

Keywords: Pleomorphic sarcoma; Malignant fibrous histiocytoma; Larynx

Introduction

Malignant mesenchymal neoplasms affecting larynx are a rare tumors. Undifferentiated high-grade Pleomorphic Sarcoma (UPS), considered as malignant fibrous histiocytoma, is a high grade malignant neoplasm characterized by tumor cells with diffuse pleomorphism in the absence of a specific line of differentiation [1-4]. The process mainly touches the males and affects all age groups. Wide surgical margin is often indicated because of their high local recurrence rates. However, due to its rarity there are scarce studies with no statistic power to define the better outcome [3,5,6]. We present a new case of undifferentiated high-grade pleomorphic sarcoma arising in larynx, and discuss pathological findings and surgical treatment of this rare tumor.

Clinical Case

60-years-old patient with no significant medical history, onset of symptoms up to 9 months for admission with permanent dysphonia, complicated by dyspnea requiring emergency tracheotomy, there was no history of dysphagia. On laryngoscopy, visualization of a well-defined, fleshy, firm mass of gray-white color, obstructing the laryngeal lumen with a planting base at the level of the right vocal cord; tomodensitometry shows the presence of a huge process of tissue density, well limit and pedicle to the right vocal cord (Figure 1).

On histological evaluation, after fixation by hematoxylin and eosin; a high grade pleomorphic malignant neoplasia, with spindle-cells arranged in a storiform pattern and a high mitotic index was identified. The tumor was present in all fragments of the sample and compromised the radial margins. The process revealed positive immune-expression for vimentin (diffuse), and negative staining for specific muscle actin, anti-PS-100, desmin, h-caldesmone and anti-CD34. The diagnosis of undifferentiated high-grade pleomorphic sarcoma of the larynx (storiform-pleomorphic malignant fibrous histiocytoma) was established.

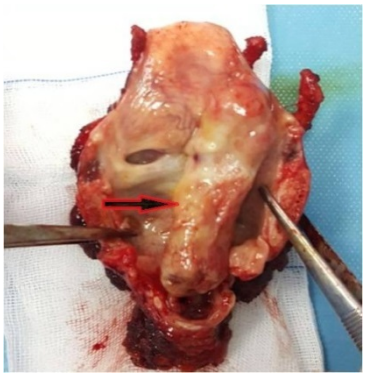

Figure 1: Huge process of tissue density, well limit and pedicle to the right vocal cord.

The patient was then submitted to a total laryngectomy without lymphadenectomy. Surgical specimen measured 7.0 cm × 2.0 cm × 3.0 cm, and showed are sidual, soft, gray-brown nodule affecting the lamina propria of the right vocal cord (Figure 2). There was no evidence of vascular or lymphatic invasion. Surgical margins were free of neoplasia. The patient does not present any post-operative complication. After 1 year of follow up, the patient has no clinic or radiologic signs of recurrence.

Comments

High-grade Undifferentiated Pleomorphic Sarcoma (UPS), or malignant fibrous histiocytoma, is a high-grade malignant tumor characterized by diffuse pleomorphic tumor cells in the absence of a specific line of differentiation. UPS is known from several histological aspects, but the mixed form of storiform and pleomorphic zones is the most common [1-4].

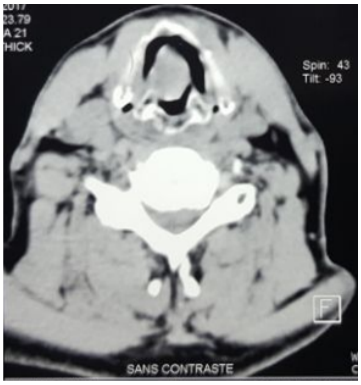

Figure 2: Surgical specimen showing elongated tumor pedicle to the right vocal cord and obstructing the respiratory tract.

UPS is a rare tumor that originates from the larynx with male predominance (3/1) and affects all age groups (6-68 years). Anatomically, it essentially touches the plane glotique laryngeal. Symptoms vary; can range from a simple feeling of strange pharyngeal body, to dysphagia or even dyspnea [3-5], usually, the USP compresses the somatic soft tissue and steadying its growth and the thigh is the most frequent location, followed by the upper end. Retroperitoneal UPS are giving a symptom like digestive disorders, malaise, slimming, and signs of increased abdominal pressure. In the literature a relationship between UPS and antecedent radiotherapy is evoked [1,3,6,7]. Our patient, though, did not have exposure to radiation therapy.

On gross examination, laryngeal UPS are sessile to polypoid, firm generalities; often ulcerated lesions, with a yellow-brown to gray-white appearance. Sometimes on can find nodular forms fleshy, multi-lobulated and fleshy, large diameter, sometimes more than 10 cm. On the skin surface, the myxoid, hemorrhagic and necrotic zones are common features [3,5,8-10]. Upon microscopic examination, the UPS may have pleomorphic, spindle cell, round cell and epithelioid appearance, and an unidentifiable line of differentiation. The storiform zones consist of spindle-shaped cells entangled in short bundles in a cartwheel, or storiforms, around slit-like vessels [1,3,5,8].

The diagnosis of the USP based on in-depth studies of sections stained with hematoxylin-eosin, and immunohistochemical finds its interest for the elimination of other tumors pleomorphic. USP has characteristics of fibroblasts/myofibroblasts, and may show positive expression for smooth muscle actin. Focal expression for cytokeratins can be found. Desmin and h-caldesmon are typically negative. The differential diagnosis includes other malignant tumors with comparable degree of cell pleomorphism such as sarcomatoid carcinoma, fibrosarcoma, myxofibrosarcoma, pleomorphic forms of liposarcoma, leiomyosarcoma, rhabdomyosarcoma, osteosarcoma, and chondrosarcoma [3,4,6,8-10].

Squamous cell carcinoma (SCC) is the most common laryngeal tumor, with predilection for the glottis and supraglotic region [3,8]. The SCC mainly affects men and is related to tobacco and alcohol. The tumor originates from the squamous mucosa that has undergone squamous metaplasia, or dysplasia appearing in the epithelium. Then tumor invasion occurs either by contiguous, or *via* lymphatic vessels and blood vessels to regional ganglia [3,8,9]. Remote metastases are rare and can occur at advanced stages of the disease TNM staging, Resection margins, proliferative index, and lymph vascular and per neural invasion are clinical predictors. Lymph node metastasis is a sign of poor prognosis for SCC [3,8]. This biological behavior of USP differs significantly from that of SCC; USP is a high-grade mesenchymal malignant neoplasm derived from the connective tissue of the larynx, which is the reverse of SCC; haematogenous dissemination to the lungs is common, with no tendency to develop cervical metastases. The favorable prognostic factors related to the specific survival of the disease are: AJCC stage I or II, negative surgical margin, superficial localization, myxoid subtype, and less than 50 years; wide surgical margins are indicated by the high rate of local recurrence, ranging from 44% to 73% [3,5,6,8-10].

An en bloc resection is treatment of choice, small lesions can be managed by excision under direct laryngoscopie, but for large lesions wide field laryngectomy is required. The indications for adjuvant radio-therapy are high grade tumor, positive surgical margins, large tumor size, and recurrent disease. Chemotherapy can be used in those patients with resectable lesions. The differentiation of tumor, size, vascular invasion and metastasis are the prognosis factors for survival and the 5 years survival is approximately 60% [10,11].

Conclusion

UPS arising in larynx is a very rare mesenchymal malignant tumor, which more commonly affects males. Immunohisto-chemistry is a fundamental tool to establish the diagnosis.

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