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# A Rare Case: Laryngeal Sarcomatoid Carcinoma

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**Abstract**

**Background:** Spindle cell carcinoma (sarcomatoid carcinoma) is a rare carcinoma that accounts for 2-3% of all laryngeal malignancies. Here, we present a case with laryngeal tumor diagnosed as Spindle cell carcinoma.

**Case Presentation:** Fifty-nine-year-old male patient was admitted to our clinic with the complaint of hoarseness and shortness of breath that has been increasing for 10 months. Indirect laryngoscopy performed on the patient, and a polypoid lesion was detected in the anterior of the left vocal cord. The patient was admitted to our clinic for direct laryngoscopy and the lesion was completely excised from the base. The case was diagnosed as "sarcomatoid carcinoma in histopathological examination.

**Conclusion:** Sarcomatoid carcinoma is usually diagnosed and treated at an early stage. So, it has a good prognosis. However, it has a poor prognosis in the advanced stage.

*Keywords:* Larynx, Polyp, Sarcomatoid carcinoma, Spindle cell carcinoma, Vocal cord

## 1. Background

Laryngeal cancer is the most prevalent cancer in the head and neck region. It account for approximately 3% of all malign tumors seen in adults. Around 85-90% of laryngeal cancers are squamous cell carcinoma (1-2). Spindle cell carcinoma (sarcomatoid carcinoma) is a rare carcinoma that accounts for 2-3% of all laryngeal malignancies (3-4). Sarcomatoid carcinoma, most commonly seen in the head and neck region, is a rare variant of squamous cell carcinoma. This tumor, which shows a very aggressive course, is mostly observed in patients receiving head and neck radiotherapy. These tumors are generally found in the anterior of the vocal cord as glottic localized and polypoid masses that have the ability to infiltrate and metastasize. The histogenetic origin and morphological classification of sarcomatoid carcinoma is not clear. It is important to distinguish sarcomatoid carcinoma from other laryngeal malignant tumors in the diagnosis.

## 2. Case Presentation

A fifty-nine-year-old male patient was admitted to our clinic with the complaint of hoarseness and shortness of breath that has been increasing for 10 months. Indirect laryngoscopy was performed on the patient, and a polypoid lesion, about 1-1.5 cm in size, was detected in the anterior of the left vocal cord (Figure 1). The surface of the growth was leukoplastic. In the magnetic resonance image (MRI) of the patient, the lesion was observed to be limited in the vocal cord. Bilateral vocal cord mobility were normal and no palpable lymphoadenopathy was found on the neck examination. The patient was admitted to our clinic for direct laryngoscopy. The lesion was completely excised from the base with direct laryngoscopy, considering the vocal cord polyp (Figure 2). The case was diagnosed as "sarcomatoid

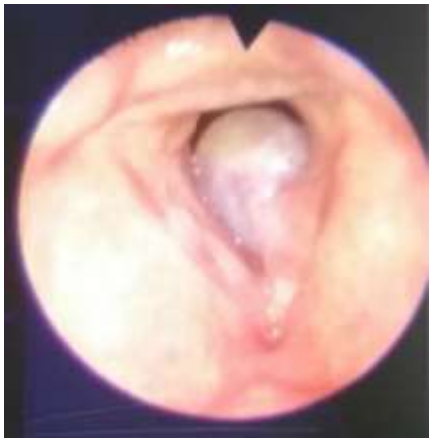

**Figure 1.** Polypoid lesion on the left vocal cord

**Figure 2.** Vocal cord polyp removed with direct laryngoscopy

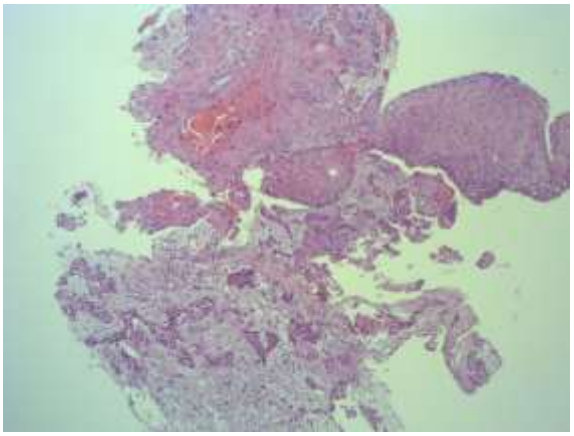

**Figure 3.** Classical invasive squamous cell carcinoma area in the superficial part of the lesion in the larynx biopsy material (Hematoxylin-Eosin stain X 50)

carcinoma" in the histopathological examination (Figures 3-5). It was reported that, there was no malign cell at the surgical margin. Furthermore, no lymph node and distant metastasis were detected clinically and radiologically. The patient was accepted as stage 1 larengeal cancer and was referred for an adjuvant radiotherapy.

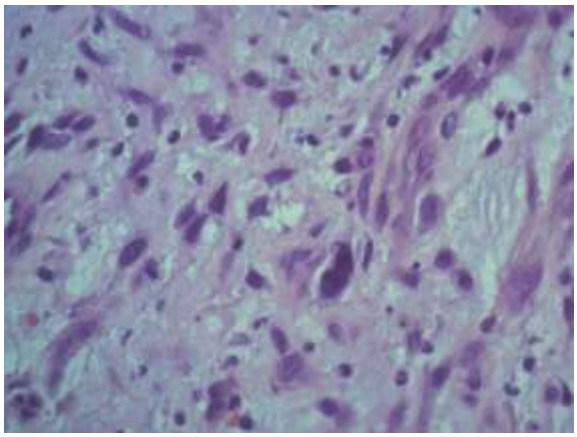

**Figure 4.** Histopathological findings of the deep stromal tissue sample of the lesion. In the subepithelial area, pleomorphic spindle-like tumor cells that do not form solid epithelial structures within the hypocellular myxoid stroma (Hematoxylin-Eosin dye X 400)

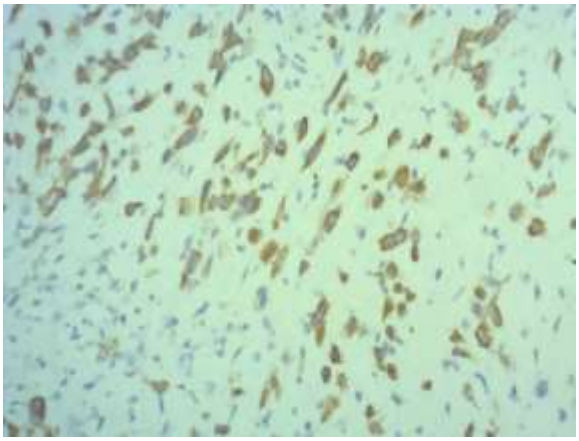

**Figure 5.** Spindle tumor cells showing positivity in Cytokeratin examination, which is characteristic in immunohistochemical staining

## 3. Discussion

Spindle cell carcinoma, or sarcomatoid carcinoma, is a highly malignant variant of squamous cell carcinoma, accounting for 2-3% of all laryngeal cancers. It is considered a biphasic tumor consisting of squamous cell carcinoma (in situ or invasive) and sarcomatous-looking spindle cell carcinoma (1-4). Histological classification of malignant tumors is crucial for prognosis and treatment. Spindle cell (sarcomatoid) carcinoma of the larynx is a biphasic tumor with epithelial and mesenchymal-appearing components. Due to its complex phenotypic features, it is a high-grade tumor that is difficult to diagnose by light microscopy alone.

It is thought that there are different sarcoma type areas within the same tumor due to the metaplasia of the spindle cell tumor component. Due to the morphological complexity of this tumor, many terms such as pseudosarcoma, pseudo carcinoma and pseudo carcinosarcoma have been used to describe the lesion. In contrast, the tumor location in our 31 patients was glottis, 52%; hypopharynx, 29%; supraglottis, 13%; and subglottis, 6%. The typical tumor is a polypoid, bulky, gray tesion, generally located on the anterior portion of the vocal cord (5). Because most of these tumors are polypoid or pedunculated, they tend to cause obstructive symptoms. Since these tumors are symptomatic, they are usually detected at an early stage and have a good prognosis (6). Hoarseness, dysphagia dispnea and cough are the most common symptoms. Typically, the onset of complaints is shorter than one year (7, 8).

Although, arcomatoid carcinoma has a good prognosis and is usually detected and treated at an early stage, it has a poor prognosis in the advanced stage. In the related the literature, it is mostly seen in male patients and generally in the 6th and 7th decades of life. Smoking, alcohol use, and undergoing radiotherapy for the neck play a role in the etiology (6-8).

Since superficial mucosa is usually ulcerated in sarcomatoid carcinoma, there is no indication of the epithelial component of the tumor in this area. Therefore, it is possible to diagnose with samples taken from the base of the tumor. In the histopathological examination of the biopsy material belonging to the vocal cord, two tissue samples matching the superficial areas of the lesion were found to have a classical invasive squamous cell carcinoma area with partial keratinization. In addition, in other tissue samples, the presence of tumoral infiltration consisting of scattered individual cells with

large hyperchromatic nuclei, spindle and pleomorphic-looking cells in the edematous, myxoid and hypocellular stromal areas under the epithelial region was noted. Cytokeratin AE1/AE3 immunohistochemical examination was performed to determine the epithelial origin of these cells in the hypocellular stromal areas and it was found that these cells were stained. With these histopathological and immunohistochemical findings, the case was evaluated as spindle cell "sarcomatoid" squamous cell carcinoma with hypocellular and myxoid stromal changes.

In cases of spindle cell "sarcomatoid" squamous cell carcinoma, sometimes the classical squamous cell carcinoma area may not be present in the biopsy samples. This is especially important in cases of tumor with hypocellular stroma. In these cases, the tumor is characterized by a very small number of cellular infiltrates with individual distribution. This situation can make the diagnosis of histopathological tumor very difficult and even impossible in biopsies. Clinical and histopathological correlation is critical. If deemed necessary by the pathologist, a biopsy can be sampled from the lesion site again and pathological diagnosis can be achieved. Sarcomatoid carcinoma has a tendency for local recurrence, regional lymph node involvement, and distant metastasis. In a study with 187 laryngeal sarcomatoid carcinomas, Thompson et al. reported local metastasis at a rate of 45% (9). Metastasis of sarcomatoid carcinoma can be epithelial, sarcomatous or mixed types. The first metastasis usually occurs in the cervical lymph nodes. Early systemic metastasis has been reported, although it is small-sized and limited in the glottis regions. Metastases of supraglottic region are more observed than glottic region. Sarcomatoid carcinoma is usually diagnosed and treated at an early stage; consequentially, it has a good prognosis. However, it has a poor prognosis in the advanced stage. It has been reported that the major prognostic factor in sarcomatoid carcinoma depends on the depth of tumor invasion. The 5-year survival rate is 64.4% (9). Nonglottic location of the tumor, vocal cord fixation, tumor size > 3 cm, epithelial tissue density on histopathological examination, metastasis to regional lymph nodes, and distant metastasis and radiotherapy are among the poor prognostic factors (5-8).

The treatment is determined by the location and stage of the tumor. The Tumor Node Metastasis (TNM) classification of American Joint Committee is used in tumor staging. Therefore, endoscopic larynx examination, computed tomography or MRI of the patient is required. Tumor size and invasion depth can be evaluated in radiological imaging. Since symptoms are evident in the early-stage tumors, tumors detected in their early stages provide a better prognosis. Most cases with laryngeal sarcomatoid carcinoma are diagnoised at the T1 stage. Moreover, distant metastasis is rarely observed. The treatment aim should be to preserve the laryngeal structure and voice quality optimally in the postoperative period. The main method of the treatment is surgery ranging from wide excision to total laryngectomy and neck dissection according to the tumor stage. Some researchers have also stated that radiotherapy is effective in the early stages (6-8). A wide local excision is mostly sufficient for a definitive treatment in the early period, and adjuvant radiotherapy should also be evaluated in the treatment. Stage 3–4 tumors can be treated with local resection, partial laryngectomy, and total laryngectomy with or without lymph node dissection, followed by a combination of radiation therapy and chemotherapy. It is known that sarcomatoid carcinoma is traditionally aggressive and radiation-resistant, and radiotherapy has a limited role in the treatment (6).

## 4. Conclusion

Sarcomatoid carcinoma is usually diagnosed and treated at an early stage. So, it has a good prognosis. However, it has a poor prognosis in the advanced stage.

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