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MALIGNANT SPINDLE CELL TUMOR – CARCINOMA OR SARCOMA?

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Summary

Spindle cell carcinoma (SpCC) is a rare subvariant of upper aerodigestive tract mucosal squamous cell carcinoma.

The histological appearance of SpCC is characterized by dual composition of malignant squamous carcinoma cells and malignant sarcoma-like spindle cells. The spindle cells are not specific for SpCC, but are present in various other malignant, benign and inflammatory lesions. Subsequently, accurate pathological diagnosis of SpCC is challenging. Although there are no clear treatment guidelines, it is widely regarded that SpCC should be treated as SCC of the corresponding localization and disease stage.

KEY WORDS: *spindle cell carcinoma, carcinosarcoma, collision tumor, head and neck squamos cell carcinoma*

MALIGNI TUMOR VRETENASTIH STANICA - KARCINOM ILI SARKOM?

Sažetak

Karcinom vretenastih (spindle) stanica rijetka je podvarijanta pločastog karcinoma sluznice gornjeg aerodigestivnog trakta.

Histološka slika tumora je specifična i karakterizira je dualna građa. Dio tumora građen je od malignih pločastih stanica poput karcinoma , dok je ostatak tumorske mase građen od malignih vretenastih stanica i pokazuje sliku sarkoma. Vretenaste stanice nisu patognomonične za „spindle cell" karcinom već ih nalazimo u nizu drugih malignih, benignih i upalnih lezija te je postavljanje prave dijagnoze izazov za patologa. Iako nema jasnih terapijskih smjernica prevladava stav da takve tumore treba liječiti istovjetno pločastom karcinomu jednakog stadija i lokalizacije.

KLJUČNE RIJEČI : *spindle cell karcinom, karcinosarkom, kolizijski tumor, planocelularni karcinom glave i vrata*

INTRODUCTION

Spindle cell carcinoma (SpCC) is a rare malignant neoplasm that occurs mainly in the upper aerodigestive tract mucosa. According to the most recent WHO classification it is a subtype of squamous cell carcinoma (SCC) (1,2,3). SpCC is most frequently encountered in larynx, but can also be found in nasal cavity, hypopharynx, oral cavity or esophagus, most commonly affecting people in their 60s and 70s with male predominance (4,5,6). Smoking, alcohol consumption, and previous irradiation of the head and neck region are predisposing factors (3,4,5). Due to relatively low incidence of SpCC and insufficient knowledge about its clinical and pathological characteristics, there

is limited data on this type of carcinoma. Consequently, a wide spectrum of nomenclature, such as carcinosarcoma, pseudosarcoma, sarcomatoid carcinoma and collision tumor, are used to describe the same clinical entity. (2,3,4,5).

CASE REPORT

A 53 year-old male patient diagnosed with a laryngeal tumor was hospitalized. He underwent previous treatment in another institution, where an emergency tracheotomy due to onset of acute respiratory insufficiency was performed. During the same procedure, a biopsy of a suspicious lesion of the right vocal cord was performed. The biopsy results described a malignant mesenchymal tumor, probably leomyosarcoma. The patient complained of hoarseness and occasional sore throat that lasted for about a year. Before treatment, he smoked up to 10 cigarettes a day for over 30 years. He was not an alcohol consumer.

After admittance, additional diagnostic procedures were performed. Flexible laryngoscopy revealed a large exophytic, polypoid mass, 4 cm in diameter, which immobilized the right vocal cord, nearly obliterating the larynx entrance. Neck ultrasound examination revealed several lymph nodes measuring up to 10 mm, located in the upper and middle third of the neck. Fine needle aspiration biopsy was performed, and there was no evidence of regional lymph node infiltration. CT scan showed an irregular, hyper-vascularized infiltrative lesion on the right side of the larynx, indistinguishable from the area of the anterior commisure and the opposite vocal cord (Figure 1). Chest x-ray and liver ultrasound showed no signs of metastatic disease. The tumor was classified as T3N0M0, stage III according to the AJCC (7). The patient underwent total laryngectomy according to the recommendations for the treatment of sarcoma limited to the larynx with no signs of locoregional and distant metastasis (Figure 2). The final

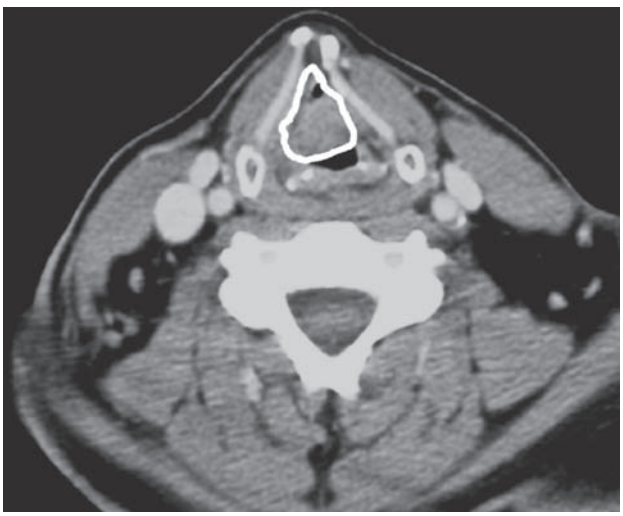

Figure 1

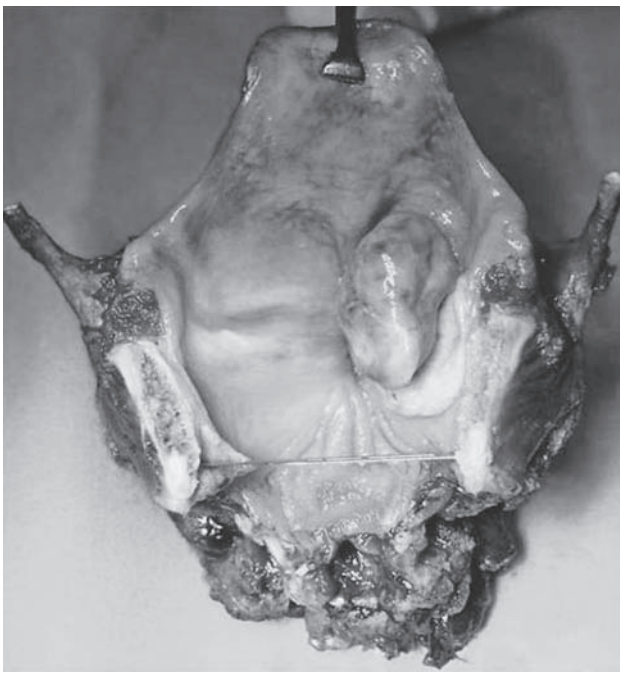

Figure 2

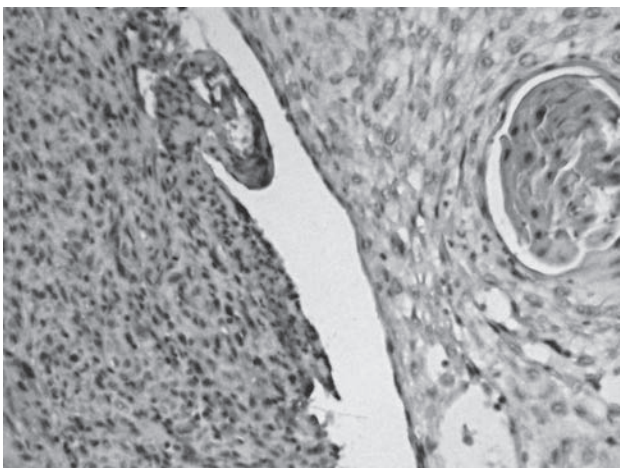

Figure 3

pathology report showed that about 35% of the total tumor mass was composed of atypical, well-differentiated squamous epithelial cells with abundant keratin creation and the histological appearance of a squamous cell carcinoma, grade I. The majority of the tumor was composed of stromal cells resembling pleomorphic sarcoma grade III (Figure 3). Malignant squamous cells were immunohistochemically positive for epithelial markers (cytokeratin and EMA), whereas the stromal cells were positive for mesenchymal markers (vimentin and SMA). The final diagnosis of SpCC was established, and consequently, adjuvant iradiation was applied. The patient showed no evidence of disease during a follow up period of 20 months.

DISCUSSION

SpCC is a biphasic tumor that shows a mixed histology. Morphologically, it is composed of malignant cells of squamous epithelium and a malignant sarcomatoid component. Malignant squamous cells are of epithelial origin and show a histopathological picture identical to other squamous cell carcinomas. Sarcomatous stroma, on the other hand, is composed of spindle like cells that show a mesenchymal appearance.

As mentioned earlier, the etiopathogenesis of SpCC is not fully understood. The epithelial origin of malignant squamous cells is generally accepted. The origin of spindle cells and stromal component is unclear and is subject to several theories (8,9). Three different theories have been proposed to explain the histogenetic nature of spindle cells (10).

According to the first theory, SpCC is a collision tumor where spindle and epithelial cells arise simultaneously from separate stem cells. Another theory explains the origin of the spindle components as atypical reactive stromal proliferation. The third, monoclonal theory, assumes that both components of the tumor arise from one totipotent stem cell that can differentiate into separate epithelial and mesenchymal directions.

Recent studies have shown that, despite of their mesenchymal appearance, spindle cells retain a number of epithelial ultrastructural and genetic characteristics. In the majority of cases, they show combined mesenchymal and epithelial immunoreactivity (4,5,10,11,12). Recently published work supports the monoclonal theory, resulting in its current predominant acceptance. There is a number of other pathological entities of the same localization that are composed of spindle cells. Differential diagnosis includes benign and inflammatory lesions but also malignant tumors such as fibrosarcoma, rhabdomyosarcoma, leiomyosarcoma and spindle cell melanoma (4,5). The treatment of SpCC is a matter of controversy and a limited number of cases results in a lack of clear treatment guidelines. It is widely accepted that these tumors should be treated as squamous cell carcinoma of the same stage and localization (9).

Early T1 and T2 tumors should be treated with either surgery or radiotherapy. Advanced T3 and T4 tumors should be treated with a combined approach, surgery and subsequent adjuvant radiotherapy. Although SpCC has many similar clinical and histopathological features as SCC, the former is more aggressive and hence results in lower overall survival.

CONCLUSION

SpCC is a rare variant of squamous cell carcinoma of the upper aerodigestive tract mucosa. It is characterized by dual structure; keratinized SCC and spindle cell type sarcomatous stroma. Histopathological appearance varies from case to case or within different areas of the same tumor tissue. This can often result in falsely labeling the tumor as sarcoma. Since sarcomas in the head and neck are extremely rare and are treated differently, SpCC should be always considered in diagnosing any lesion with spindle cell component.

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