

CASE REPORT

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A unique case of triple malignancy of the head and neck region managed successfully in a single surgery

Aliaksandr Aksionau, Rusella Mirza, Ashley Flowers, Nestor Dela Cruz

ABSTRACT

Introduction: Head and neck skin tumors usually occur with increasing sun exposure. The most common types are squamous cell and basal cell carcinomas (BCCs). Soft tissue sarcomas of the head and neck region are rare malignancies, especially the leiomyosarcoma (LMS) type. Leiomyosarcoma may appear nonspecific, making diagnosis difficult. **Case Report:** We present a unique and interesting case of simultaneous LMS, BCC, and squamous cell carcinoma (SCC) of the head and neck region in a 96-year-old patient. All lesions were excised in the same surgery day with negative surgical margins and diagnosed according to standard protocol. **Conclusion:** Basal and SC carcinomas are the most common nonmelanoma skin cancers of the head and neck. Leiomyosarcoma is a rare entity for this region. On extensive literature search, eight cases of the external ear LMS were reported from 1964 to 2019. As a rare type of cancer, it can be misdiagnosed and poorly managed at the beginning, leading to a worse prognosis. In our

case, the first line of treatment was surgery with a fairly wide excision of all three tumors. The diagnosis was confirmed by histomorphological evaluation of excised specimens followed by ancillary tests. A synchronous case of LMS, BCC, and SCCs is an exclusive and rare entity that is worth to be studied and reviewed.

Keywords: Basal cell carcinoma, Head and neck tumors, Leiomyosarcoma, Squamous cell carcinoma

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INTRODUCTION

Based on the Union of International Cancer Control, SCC of the head and neck is the sixth leading cancer by incidence worldwide. About 300,000 deaths occur among more than 550,000 new cases of head and neck cancer each year [1]. Based on the American Cancer Society, the actual number of the most common type of nonmelanoma skin cancer (NMSC), such as basal cell and squamous cell, is difficult to estimate as these cases do not have to be recorded in cancer registries [2].

Soft-tissue sarcomas (STSS) are relatively rare malignancies with approximately 12% occurring in the head and neck region [3]. Based on the National Cancer Institute's Surveillance Epidemiology and End Result for 2012–16, LMSs represent 12.8% among all STSS [4]. Head and neck STSS are associated with a higher

local recurrence rates and lower overall disease-specific survival rates of all STSs [3, 5]. Leiomyosarcomas are malignant tumors derived from the smooth muscle and are rarely found in patients aged less than 40 years [6, 7]. The outer ear location of these tumors affects mainly white individuals, with a small predominance of males [3, 7]. The gross appearance of these tumors can be misleadingly benign and can be mistaken for nonmalignant conditions. Early diagnosis and aggressive initial treatment remain the basis of therapy for a good prognosis [8]. In the head and neck, LMSs have been reported in the sinonasal tract, hypopharynx, cervical esophagus, and trachea [9], which makes the external ear location unique.

In the majority of LMS, SCC, and BCC cases, surgery with negative surgical margins is the first line of treatment [1, 3, 7–9]. Each patient should be regularly monitored for a tumor recurrence by a medical team.

CASE REPORT

A 96-year-old Caucasian male was referred by his primary care physician to our hospital for the surgery on three new skin lesions in the head and neck region. He had a history of SCC of nasal dorsum seven months prior to his admission, which was excised with negative surgical margins. The patient complained of mild ear soreness and noted fast growth of the ear “bump,” but he did not remember when it appeared. He did not worry about his cheek lesions and did not remember when they appeared as well. Among radiological methods, a computed tomography (CT) scan with intravenous contrast was used a month before the surgery. It showed an enhancing soft tissue mass that involved the lateral surface of the right auricle and measured $2.2 \times 1.9 \times 2.3$ cm (in transverse, anteroposterior, and craniocaudal diameters, respectively). There was infiltration of the underlying subcutaneous fat with a depth of invasion approximately 1.6 mm with focal involvement of the underlying cartilage. No abnormal lymphadenopathy, other masses, or abnormal enhancement were revealed by CT criteria. Two weeks before the surgery day, a biopsy of the ear mass favored LMS. Lesions of the cheeks were not biopsied; based on the appearance and history of previously treated SCC, they were determined for surgical excision simultaneously with the ear mass with subsequent histopathological evaluation after treatment. The remainder of the clinical examination was normal. There was no history of radiation exposure/therapy to the head and neck. Lymph nodes were not palpable. The patient complied with his oral medication for hypertension and diabetes mellitus. On the surgery day, all three lesions were excised with wide margins. Only the ear mass was evaluated for margins in the frozen section room with margins free of tumor cells.

For the right ear lesion, the partial auriculectomy was performed with wide local excision 6×3 cm. Its clinical appearance was a large red ulcerated nodular mass

involving the upper 2/3 of the right anterior ear. Grossly, the mass was $3.7 \times 3.5 \times 3$ cm, soft, and covered by tan-brown crust. The erythema and abnormal skin extended into the conchal bowl but did not extend into the external auricular canal (Figure 1A). It was illustrated by the results of CT (Figure 1B, C), and increased dimensions of the mass confirmed fast growth. Histologic examination showed a spindle, moderately differentiated, cell line characterized by cigar-shaped and blunt-ended nuclei with marked polymorphism. Some cells demonstrated mitotic atypia up to 5–8 per high power field. The tumor had a distinct fascicular growth pattern and a well-defined border (Figure 1D, E). The overlying squamous epithelium was attenuated and focally ulcerated. Squamous dysplasia or carcinoma in situ was not identified. Ancillary immunohistochemistry (IHC) stains showed tumor cells positive for smooth muscle actin (SMA) and muscle-specific actin (MSA) confirming smooth muscle origin. Negative IHC for pancytokeratin (AE1/AE3), 34BetaE12, CK 5/6, p40, p63, desmin, myogenin, S100, and SOX10 ruled out squamous, epithelial, skeletal muscle, neural, and melanocytic origin (Figure 2). The diagnosis of LMS, grade 2 was made.

The second lesion was excised 2×3 cm followed by the transfer of adjacent tissue. It was an ulcerated skin lesion, located on the right cheek, and measured 0.4×0.7 cm (Figure 3A). Microscopically on H&E stain, it represented basaloid cells with scant cytoplasm and elongated hyperchromatic nuclei. The cells invaded the underlying stroma by 0.2 cm, lined up in parallel arrays and were surrounded by myxoid stroma diagnostic of BCC (Figure 3B).

The third lesion was excised 2×3 cm followed by the transfer of adjacent tissue. It was a white-gray nodular skin lesion with central hyperpigmentation, located on

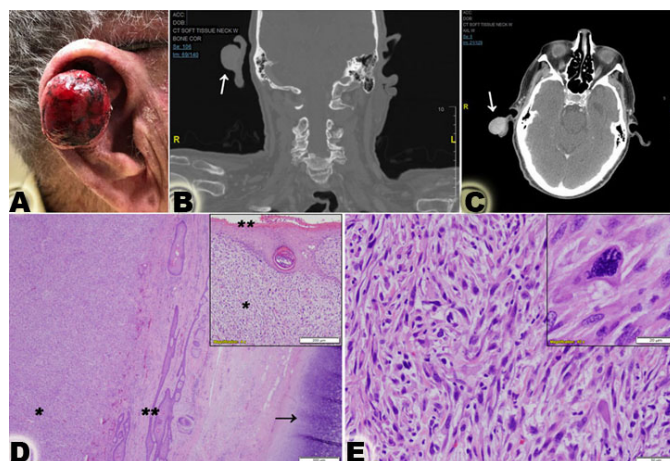


Figure 1: The right ear lesion. Gross appearance of a nodular mass with an ulcerated surface (A). CT of the head and neck, frontal (B) and axial sections (C) demonstrate the ear mass. H&E stain shows a spindle cell neoplasm (*) with relation to the auricular cartilage (→) and skin (**) (D). Marked nuclear pleomorphism and increased mitotic activity are present (E). The magnification for D and E is $\times 20$, $\times 40$ (inset), $\times 200$ and $\times 400$ (inset), respectively.

the left cheek, and measured $0.7 \times 0.7 \times 0.2$ cm (Figure 4A). Microscopically, it represented squamous cells growing below the level of overlying epidermis (0.3 cm in depth) with keratin pearl formation diagnostic of SCC (Figure 4B).

According to the current recommendations, all lesions were excised with negative surgical margins. For the ear

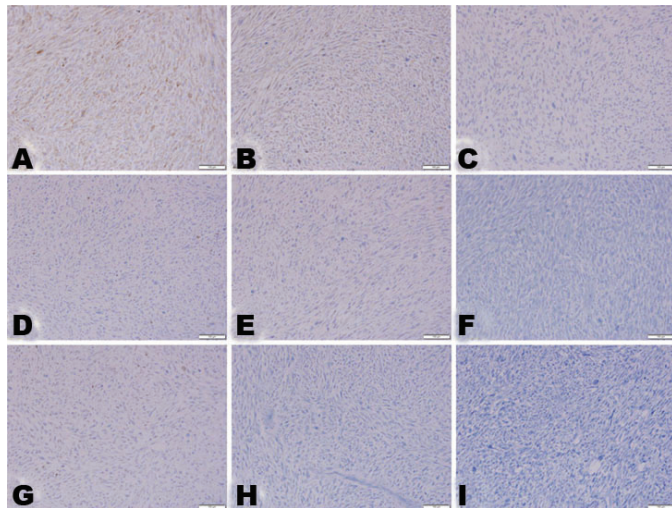


Figure 2: Ancillary methods for the ear lesion. Tumor cells are positive for MSA and SMA (A and B, respectively), and negative for AE1/AE3 (C), desmin (D), myogenin (E), p40 (F), p63 (G), S100 (H), and SOX-10 (I). All images are taken at $\times 100$ magnification.

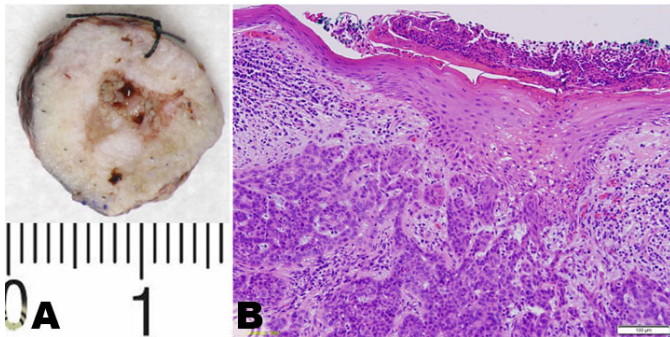


Figure 3: The right cheek lesion. Gross appearance of an ulcerated skin lesion (A) with nested basaloid cells on microscopy (B) (H&E stain, magnification is $\times 100$).

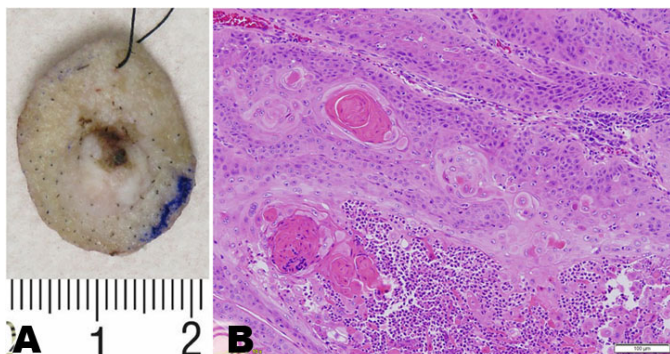


Figure 4: The left cheek lesion. Gross appearance of an elevated skin lesion (A) with infiltrating nests of keratinized squamous cells on microscopy (B) (H&E stain, magnification $\times 100$).

lesion, the distance from the tumor cells to the closest surgical margin was 1.4 cm, for BCC and SCC—0.3 and 0.5 cm, respectively.

DISCUSSION

Nonmelanoma skin cancer is the most common cancer in the United States with an estimated incidence of more than 3.5 million each year. Basal cell carcinoma and cutaneous SCC account for 99% of all NMSC [10] with BCC being in the lead, accounting for approximately 65% [11]. Soft-tissue sarcomas are a rare type of head and neck tumors with a broad range of differential diagnoses. These tumors require careful microscopic examination and ancillary tests to determine origin since cells can demonstrate a wide range of differentiation, such as skeletal muscle, adipocytes, smooth muscle, vascular and neural tissues, while some cases lack specific differentiation [12]. One type of pathology does not exclude the coexistence of another type, even if they are located in the same area or in the same organ; the same applies to tumors.

Basal cell carcinoma and SCC develop from epidermal keratinocytes; the incidence is associated with increased sun exposure. For SCC, the cause may also be infections caused by human papillomavirus and human immunodeficiency virus, chronic inflammatory skin diseases, and burn scars [10]. Basal cell carcinomas most often are found in the nose (32.3%), orbital (19.1%), and cheek (18.1%) areas [11]. Basal cell carcinoma rarely metastasizes, unlike SCC, which is often locally aggressive and metastasizes to regional lymph nodes [10]. In most cases, H&E stain is used only. Being often found in clinical practice, SCC and BCC are regularly treated by family practice physicians or surgeons of various subspecialties, which is not applicable for LMSs.

Leiomyosarcoma has been classified into site-specific subgroups due to remarkable clinical and biological dissimilarity [12]. Leiomyosarcomas are divided into superficial (of the skin) and deep [13, 14], in turn, superficial tumors include cutaneous (dermal) and subcutaneous types [7, 13]. Generally, LMSs of the skin represent a solitary, slowly growing lesion, most often on the proximal extremities, which coincide with the hair-bearing areas [15]. The majority of LMS of the head and neck occur in the oral cavity, superficial soft tissues, such as scalp, jaws, and paranasal sinuses [16]. The etiology of these tumors is relatively unknown, although antecedent traumatic injury, sunlight, chemicals, ionizing irradiation, and lupus vulgaris have been associated with this type of tumor [15–17]. When the tumor is located in the dermis, its origin lies in the pilar follicle erector muscles or smooth muscles surrounding the sweat glands. If the tumor affects the subcutaneous tissue, its origin is in the smooth muscle of veins and arteries [7, 13, 15, 18–20]. While the arrector pili muscle is considered to be the most likely source of cutaneous LMS, some authors have

suggested that the tunica media of the blood vessels is the source of deep primary LMS [21].

Clinically, the overlying skin of cutaneous LMSs is red in color while it appears normal in subcutaneous forms. In addition, cutaneous LMSs are fixed to epidermis while subcutaneous LMSs are mobile. Moreover, ulceration and crusting of the skin support the diagnosis of cutaneous types [13].

Since LMS is the least common head and neck sarcoma, its diagnosis is often missed or delayed leading to a more

advanced stage of disease at the time of diagnosis [19, 22], which is confirmed by histopathological evaluation of an excised specimen after IHC [7, 20, 22, 23]. Differential diagnosis includes benign and malignant tumors of soft tissue, poorly differentiated epithelial malignancy, and melanoma (Table 1) [20, 24, 25]. On microscopy, LMS shows poorly oriented atypical spindle cells with mitoses. The number of mitoses per high power field is an important criterion and ≥ 2 mitoses per high power field almost certain to be malignant [23].

Table 1: Immunohistochemical differential diagnosis of some spindle-cell tumors

Neoplasm	CK/EMA	S100	SOX-10	SMA	Desmin
Leiomyoma	-/rare F+	-	-	+	+
LMS	-/rare F+	-	-	+	+
Rhabdomyosarcoma	-	-	-	-	+
Neurfibroma	-	F+	+	-	-
Schwannoma	-	D+	+	-	-
Malignant peripheral nerve sheath tumor	-/F+	+/- (50% F+)	+/- (50% F+)	-	-
Angiosarcoma, Kaposi's sarcoma	-/F+	-	-	-	-
Fibromatosis	-	-	-	+	-/+
Nodular fasciitis	-	-	-	+	-
Inflammatory myofibroblastic tumor	-/rare F+	-	-	+	+/-
Dermatofibroma	-	-	-	-	-
Dermatofibrosarcoma protuberance	-	-	-	-	-
Liposarcoma	-	-/+	-	F+	+/-
Spindle cell melanoma	-	+	+	-	-
Solitary fibrous tumor	-	-	-	-	-

Abbreviations: F – focal, D – diffuse.

For the treatment, most authors recommend wide local excision with no less than 1 cm margins for most cutaneous lesions [7, 13, 16, 20, 22, 26]. Leiomyosarcoma is an aggressive tumor that frequently shows recurrence with regional or distant metastasis [12]. Early-stage cancers have a very favorable prognosis with high cure rates with surgery or radiation alone [1, 17]. Chemotherapy or concurrent chemotherapy/radiation is not indicated. Some authors mention that postoperative irradiation is controversial, since LMSs are reported to be minimally sensitive to chemotherapy or radiation therapy. The frequency of local relapses ranges from 30% to 50%, which indicates the inadequacy of primary treatment [26]. However, head and neck LMSs have a very high incidence of regional recurrence that do not necessarily coincide with the original tumor, even in cases of complete excision and adjuvant therapy. The most common cause

of death is a local relapse, and the five year survival rate without relapse is reported to be only 32%. Metastases are almost exclusively hematogenous, but metastases to the cervical lymph nodes have been documented in some cases [19].

CONCLUSION

As most common NMSC of the head and neck region, SCC and BCC are regularly treated by physicians or various subspecialties using standard surgical excision, which includes the removal of a tumor with negative margins. Leiomyosarcoma is a rare entity in the ear and its early diagnosis and proper treatment are necessary for a better prognosis. Immunohistochemistry is beneficial to differentiate it from other common head and neck

carcinomas. The age of our patient and a synchronous appearance of different types of tumors in the head and neck region are likely associated with excessive sun exposure.

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Author Contributions

Aliaksandr Aksionau – Conception of the work, Design of the work, Acquisition of data, Analysis of data, Interpretation of data, Drafting the work, Revising the work critically for important intellectual content, Final approval of the version to be published, Agree to be accountable for all aspects of the work in ensuring that questions related to the accuracy or integrity of any part of the work are appropriately investigated and resolved

Rusella Mirza – Conception of the work, Design of the work, Acquisition of data, Analysis of data, Interpretation of data, Drafting the work, Revising the work critically for important intellectual content, Final approval of the version to be published, Agree to be accountable for all

aspects of the work in ensuring that questions related to the accuracy or integrity of any part of the work are appropriately investigated and resolved

Ashley Flowers – Interpretation of data, Revising the work critically for important intellectual content, Final approval of the version to be published, Agree to be accountable for all aspects of the work in ensuring that questions related to the accuracy or integrity of any part of the work are appropriately investigated and resolved

Nestor Dela Cruz – Interpretation of data, Revising the work critically for important intellectual content, Final approval of the version to be published, Agree to be accountable for all aspects of the work in ensuring that questions related to the accuracy or integrity of any part of the work are appropriately investigated and resolved

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Conflict of Interest

Authors declare no conflict of interest.

Data Availability

All relevant data are within the paper and its Supporting Information files.

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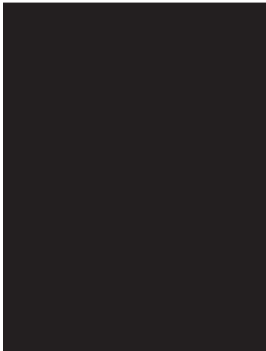
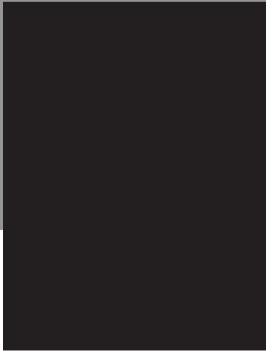
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