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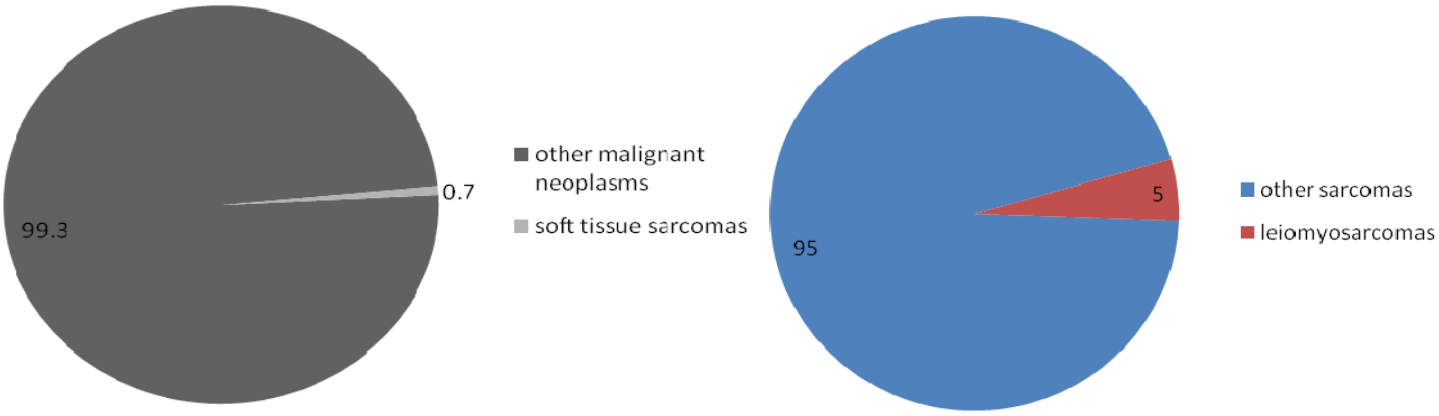
內文：

Introduction

Soft tissue sarcomas compromise approximately 0.7% of all malignant neoplasms. leiomyosarcomas (LMSs) have been reported to account for 3-7% of soft tissue sarcomas. [1]



Superficial LMSs arising in the dermis with or without extension into the subcutis are referred to as cutaneous LMSs while tumors arising in the subcutis are termed subcutaneous LMSs. [2]

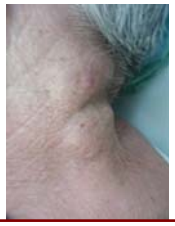
Leiomyosarcomas ⇔ deep / peripheral (superficial LMS) ⇔ cutaneous LMSs / subcutaneous LMSs

Subcutaneous LMS

- Be thought to arise from small to medium-sized blood vessels in the subcutaneous tissue[3]
- associated with higher rates of local recurrence, metastasis and death from disease, compared with lesions arising from cutaneous structures
- painless or tender solitary subcutaneous nodule or group of nodules
- arise on the head and neck, back, thigh and beneath radiation dermatitis[4,5]

Case report

A 67-year-old Greek woman, presented in the General Hospital of Volos with a painless neck mass on the left.



She reported that the lesion appeared in the last year and had a slow growth.

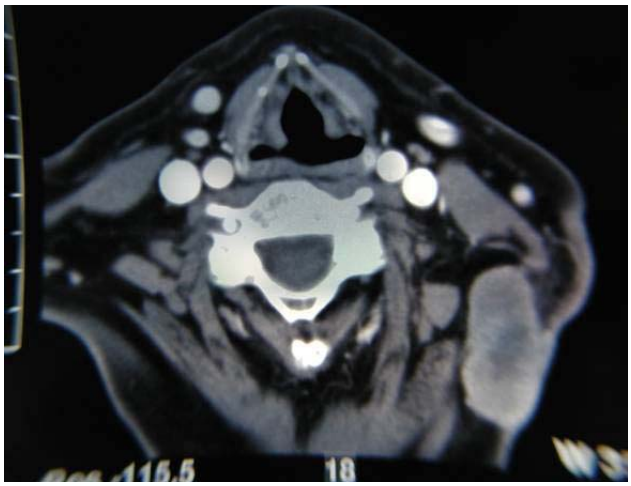
No previous operations were recorded in patient's medical history.

Fine needle aspiration biopsy (FNA)

Fine needle aspiration biopsy (FNA) of the lesion revealed a small amount of inflammatory cells mainly lymphocytes and several neoplastic cells with malignant features advocating undifferentiating carcinoma. These cells were either scattered or aggregated with large, hyperdense nuclei, nuclear membrane grooves and several abnormal mitoses. Several neoplastic cells were spindle shaped with solid cytoplasm and multiple nuclei.

Computed tomography

Computed tomography with contrast of the neck region revealed a bi-lobe lesion of 3.2 × 2 × 4 cm dimensions with peripheral enhancement and hypodense necrotic center in the subcutaneous tissue of the posterior neck triangle with no involvement of the deeper muscle tissue.



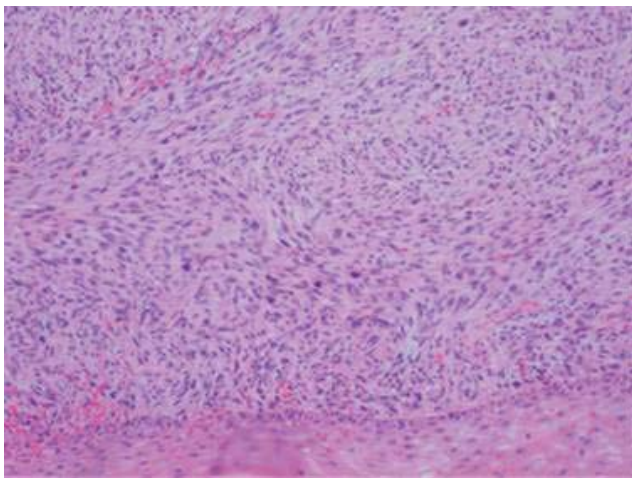
Endoscopy

fter a thorough clinical examination, including endoscopy of the nasopharynx, larynx, and hypopharynx, and with no signs of metastatic disease, the lesion was excised under topical anesthesia.

Histology

Histology showed a low grade nodular malignant mesenchymal neoplasm consisting of perpendicularly arranged fascicles of spindle cells with eosinophilic fibrillary cytoplasm, scattered pleomorphic nuclei and irregular mitosis with a rate of 6 to 8 per 10 high power-fields.

The lesion was localized in the subcutaneous tissue, while medially it was in contact with striated muscle without infiltrating it.



Immunohistochemical staining

On immunohistochemical staining, the tumor expressed focal smooth muscle actin (a-SMA) and vimentin, while a small number of tumor cells were also weakly positive to HHF-35, desmin and S-100 stains.

註：The Speical Protein Express of The Leiomyosarcoma (收集自網路)
smooth muscle actin (a-SMA)：平滑肌中的肌動蛋白
HHF-35： muscle-specif actin的一種
Smooth muscle myosin (SMMS)：平滑肌中的肌凝蛋白
Vimentin ：真核細胞重要的細胞骨架結構，中間蛋白絲(intermediate filament) 。平常細胞呈穩定狀態，但在fibroblast內為動態狀態。
desmin： 肌小節(sarcomeres)中靠近Z線的中間蛋白絲
S-100 stains：可染神經脊中melanocytic origin的細胞，及某些種類的histiocytes。

According to the histopathology and immunohistochemistry, the final diagnosis was subcutaneous LMS of the neck and the specimen's surgical margins were negative. Brain magnetic resonance as well as computed tomography of lungs and abdomen did not reveal any distant metastatic disease.

Due to the local aggressive nature of the disease, the patient underwent a further wider local excision of the dermis and subcutaneous tissue of approximately 3 cm around the first excision and the defect was reconstructed with a regional rotational flap.

No adjuvant therapy was recommended and three years postoperatively the patient is without signs of local recurrence or metastasis.

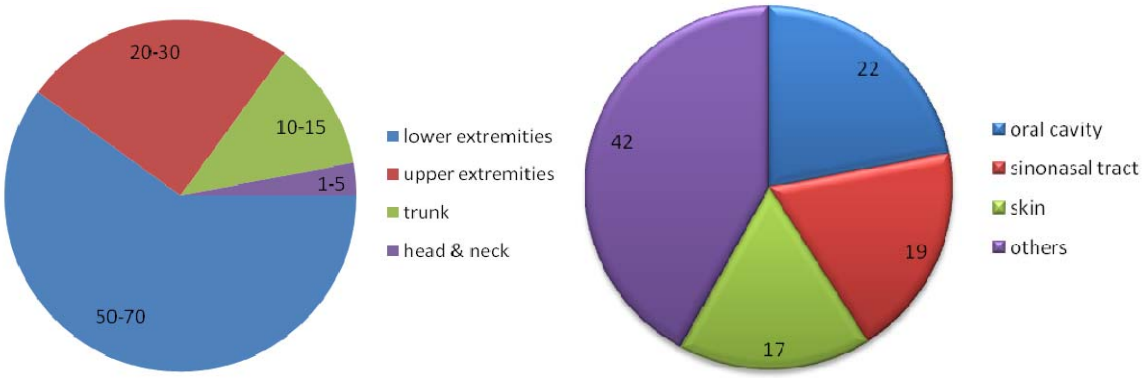
Discussion

Soft tissue sarcomas are relatively rare neoplasms that may arise in any anatomic region.
Occurrence in the head and neck accounts for less than 1% of all malignant tumors in this site. [5]

Sarcomas of the head and neck most commonly present as painless submucosal or subcutaneous mass of uncertain duration.

Superficial LMS is presented in middle age with a median age of diagnosis 45 to 50 years of age. A male-to-female predominance is reported of 2:1 to 3:1.

Superficial leiomyosarcomas occur on the lower extremities (50-70%), the upper extremities (20-30%), the trunk (10-15%), and head and neck (1-5%). [3]
The most common site for head and neck LMSs is the oral cavity (22%), followed by sinonasal tract (19%) and skin (17%). [6]



LMSs may originate from undifferentiated mesenchymal cells or it may be metastatic from other body regions, mainly as a late event associated with systemic metastasis and poor prognosis. [6]

The low incidence of LMSs in the head and neck region is attributed to the scarcity of smooth muscle in this area which is limited to vessel walls, erector pili muscle of the hair follicles, esophagus and the posterior wall of the trachea. [6,7]

Subcutaneous LMS may be covered with intact skin or may sometimes invade the overlying dermis.

Management of these lesions should begin with a thorough clinical examination, followed by imaging studies, CT and/ or MRI.

MRI has the advantage of delineating vascular involvement and is valuable for lesions located in the neck and parapharyngeal region.

In case of a neck mass the appropriate work-up has to exclude the possibility of metastatic carcinoma or lymphoma. FNA in experienced hands can rule out metastatic squamous cell carcinoma, thyroid carcinoma or lymphoma. FNA results may also be suspected for soft tissue sarcoma, although it is difficult to diagnose a particular subtype of sarcoma. False negative results may be attributable to necrotic center of the tumor. [5,7]

Subcutaneous LMS shows a higher primary growth rate than cutaneous LMS and a rate of local recurrence of 40 to 60%. Distant metastases affect commonly the lungs, bones, central nervous system and liver in 30 to 60% of patients.

Differential diagnosis includes myofibrosarcoma, fibrosarcoma, malignant nerve sheath tumor, malignant fibrous histiocytoma and rhabdomyosarcoma and can be made only after detailed histopathologic analysis. [9]

Treatment of subcutaneous LMS is dictated by stage, location, size and patient age. The primary modality of therapy is surgery. A wide excision with 3-5 cm lateral margins and a depth that includes subcutaneous tissue and fascia is recommended. Lymph node metastasis in head and neck sarcomas account for 10 to 15% of cases and neck dissection is not required for staging or treatment.

Radiotherapy has been used as an adjuvant therapy in order to minimize the incidence of local recurrence although sarcomas are considerable radio-resistant. Two indications for postoperative radiotherapy exist: 1) high grade lesions and/or positive surgical margins, and 2) lesions larger than 5 cm and/or recurrence. [10]

The outcome of chemotherapy in soft tissue sarcomas in head and neck is similar with that of the extremities, with adriamycin being the most important chemotherapeutic agent. [7]

Although adjuvant therapy in LMSs is still controversial as no sufficient statistical evidence exists regarding their efficacy, chemotherapy combined with radiotherapy seems to improve local control of the disease especially when wide resection cannot be achieved. [7,11]

A 42% recurrence rate for oral cavity and skin lesions has been reported [6]. Although tumor location and depth affect the recurrence rate and metastatic risk, the mitotic activity seems not to have a similar effect. [11]

Head and neck sarcomas show worse survival outcomes compared with sarcomas of the extremities, with a 5-year survival rate between 49 and 55%. [5] Since lymph node metastasis is uncommon, the survival outcome depends mainly on local recurrence and distant metastasis control. Patients should be observed for a minimum of 5 years after surgery, as recurrence rates are variable, depending on the site of the tumor. Most of the patients present local recurrence within 2 years after initial management and those with recurrent disease are at risk for developing distant metastasis. [8]

Conclusion

Management of a neck mass may surprisingly reveal a rare lesion such as subcutaneous leiomyosarcoma. A thorough clinical and laboratory examination is essential in order to achieve fast diagnosis and curative excision of this aggressive tumor. The primary modality of therapy of subcutaneous LMS is surgery, which must be designed to be curative. Adjuvant radiotherapy or chemotherapy may be used for control of local recurrence in case of positive surgical margins, high-grade or large tumors.

Abbreviations

LMS: leiomyosarcoma; FNA: fine needle aspiration; BMI: body mass index; a-SMA: smooth muscle actin; HHF-35: muscle-actin-specific monoclonal antibody; S-100: specific protein tumor-marker; CT: computed tomography; MRI: magnetic resonance imaging; HPF: high-power fields

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題號	題目
1	下列關於軟組織肉瘤(sarcoma)，何者為是？ (A) 軟組織肉瘤是頭頸部常見的癌症 (B) 最常見的軟組織肉瘤是平滑肌肉瘤(leiomyosarcoma) (C) HAART 對 Kaposi's sarcoma 的治療影響不大 (D) 遠端轉移最常發生口內軟組織的牙齦，其次為舌頭
答案(D)	出處：P.552, 553, 557, 563
題號	**題目**
2	下列關於平滑肌肉瘤(leiomyosarcoma)，何者為非？
	(A) 平滑肌肉瘤多出現在子宮壁及腸胃道，口腔內甚少出現 (B) 生長在口腔顏面的平滑肌肉瘤，好發年齡在年輕人 (C) 免疫組織化學染色可以發現有 desmin 標記的存在 (D) 容易復發及遠端轉移
答案(B)	出處：P.559