Primary parotid liposarcoma

Košec, Andro; Zurak, Krešo; Čupić, Hrvoje; Bedeković, Vladimir

Source / Izvornik: **European Annals of Otorhinolaryngology, Head and Neck Diseases, 2019, 136, 405 - 408**

Journal article, Accepted version
Rad u časopisu, Završna verzija rukopisa prihvaćena za objavljivanje (postprint)

https://doi.org/10.1016/j.anorl.2018.01.013

Permanent link / Trajna poveznica: https://urn.nsk.hr/urn:nbn:hr:105:651623

Primary Parotid Pleomorphic Liposarcoma

Andro Košec, MD[1], Krešo Zurak, MD[1], Hrvoje Čupić, MD, PhD[3], University Professor Vladimir Bedeković, MD, PhD[2]

[1] Department of Otorhinolaryngology and Head and Neck Surgery, University Hospital Center Sestre milosrdnice, Vinogradska cesta 29, Zagreb, Croatia

[2] School of Medicine, University of Zagreb, Šalata 3b, Zagreb, Croatia

[3] Department of Pathology Ljudevit Jurak, University Hospital Center Sestre milosrdnice, Vinogradska cesta 29, Zagreb, Croatia

Author correspondence:

Andro Košec, MD

Department of Otorhinolaryngology and Head and Neck Surgery, University Hospital Center Sestre milosrdnice, Vinogradska cesta 29, Zagreb, Croatia

Phone: +385 1 3787108, Fax: +385 13769067

e-mail: andro.kosec@yahoo.com

Abstract

Introduction: High-grade liposarcoma is very rare in the head and neck. To date, only 8 cases of primary parotid gland liposarcoma have been reported, with only 2 previous cases being of the pleomorphic subtype. These are the only reported subtypes resulting in mortality. The histopathologic differential diagnoses should consider intramuscular myxoma, spindle cell or pleomorphic lipoma, lipogranuloma, and glioblastoma.

Case report: We report a case of pleomorphic parotid liposarcoma arising in the left parotid gland of a 66 year-old man, causing local morbidity, recurrence, repeated surgical treatment and death 5 months after initial treatment.

Discussion: This specific subtype is marked by a high probability of local recurrence of up to 70% and is prone to distant metastatic spread, as was the case in our patient. Based on limited experience from published literature, optimal treatment entails radical surgery with negative margins. High-grade tumors have a worse outcome despite the addition of surgery and postoperative radiotherapy.

Keywords: parotid neoplasms; head and neck; surgery; pleomorphic liposarcoma

Introduction

Liposarcoma is the most common soft-tissue sarcoma arising from adipose tissue in adults and comprises 20% of all soft-tissue malignancies, affecting men more frequently than women. [1] Of all the specific histologic subtypes, the pleomorphic is the rarest. Primary tumors arising in the head and neck region are very rare, with only several previous cases arising in the parotid glands. The histopathologic differential diagnoses should consider intramuscular myxoma, spindle cell or pleomorphic lipoma, lipogranuloma, and glioblastoma. [2]

Case Report

A 66 year-old man presented with a painless and fast-growing tumor in his left parotid area over the period of several months. Facial nerve paresis with a House – Brackmann score of III/VI was noted. Fiberlaryngoscopy and contralateral neck palpation were unremarkable, and contrast enhanced MSCT imaging showed an inhomogeneous mass in his left parotid gland measuring 1.5 cm, with no evidence of extraglandular spread, but marked postcontrast imbibition, possible necrosis and border irregularity (Figure 1). Fine needle aspiration biopsy showed a high-grade mesenchymal tumor or carcinoma. The tumor was resected through a left-sided selective neck dissection (regions I, II, III and Va), total parotidectomy, digastric muscle, exterior carotid artery and facial nerve resection due to tumor infiltration with great auricular nerve graft reconstruction (Figures 2 and 3). A wide resection margin was planned with respect to the orbit and the internal carotid artery and the tumor measuring up to 4 cm in diameter was removed en bloc. Frozen tissue samples were analyzed, with results confirming a malignant mesenchymal high-grade tumor. Definitive histopathology confirmed a primary parotid liposarcoma of the pleomorphic subtype. The tumor infiltrated the parotid gland and extensive sampling failed to reveal any epithelial tumor component. Histopathologic results

showed perineural infiltration of the facial nerve, without tumor infiltration of the 25 lymph nodes identified by the pathologist in regions I-III and Va and clear margins. (Figure 4) The patient's findings were reviewed by an interdisciplinary tumor board, but postoperative chemoradiotherapy based on doxorubicin and ifosfamide was declined by the patient. After four months of follow-up, the tumor recurred in the left preauricular region. A left-sided radical neck dissection was performed, removing the sternocleidomastoid muscle, internal jugular vein and accessory nerve with multiple positive lymph nodes in the left neck and histopatological characteristics identical to the primary tumor. Two months later, the patient developed cutaneous metastatic disease in his right pectoral region and died 5 months after the initial surgery due to haemorrhage associated with metastatic infiltration and tumor cachexia. This case report followed the Declaration of Helsinki on medical protocol and ethics and its submission was approved by the University Hospital Center Ethical Review Board.

Discussion

Liposarcoma is regarded to be one of the most common soft-tissue sarcomas in adults, arising most often in the extremities, with the least number of reported cases being in the head and neck area. [3] The World Health Organization classifies liposarcomas into 5 major histological subtypes; (1) well differentiated - adipocytic, sclerosing, and inflammatory subtypes; (2) dedifferentiated; (3) myxoid; (4) round cell; and (5) pleomorphic. with the pleomorphic variant being the rarest. [2,4] When compared to the low frequency of primary parotid malignancies (1% to 3% of head and neck malignancies), and sarcomas of the major salivary glands (0.3% to 1.5% of all salivary gland malignancies), the combination of this particular histological subtype and location is the least likely of all head and neck liposarcoma cases. [1,4,5,6] The criteria for diagnosis of a primary salivary sarcoma are the absence of

sarcoma elsewhere, the absence of metastatic spread from a mucosal or skin malignancy, the gross appearance consistent with glandular origin and a carcinosarcoma excluded by multiple microscopic sections. [5,6] In this case, all of the criteria were satisfied.

Magnetic resonance imaging (MRI) and computed tomography (CT) are the most common imaging techniques in assessing lipomatous tumours. Of those, MRI is the preferred technique in establishing relations to adjacent structures of tumors considered suspicious for well-differentiated liposarcoma, but is not sufficienty accurate in distinguishing between liposarcoma and many benign lipoma variants. The most significant MRI features related to liposarcoma are the presence of thickened septa over 2 mm, nodular and globular areas of non-adipose tissue within the lesion and a total amount of non-adipose tissue making up more than 25% of the lesion. Computed tomography scans better display calcifications in the lesion and its relation to bone structures. Fludeoxyglucose positron emission tomography (FDGPET) may also help in diagnosing different liposarcoma types, but its accuracy is not sufficient to support the omission of diagnostic biopsy. [6]

Only 11 reported cases of primary salivary gland liposarcoma were identified to date, with two studies listing the parotid and the neck as one indiscriminant area. Gerry D et al. reported 13 cases of salivary gland liposarcoma and Davis EC et al. reported 1 case of parotid gland liposarcoma, but without specific data on localization, histology, follow up or treatment. (Table 1). [7,8,9,10,11] That is often caused by a grouping head and neck liposarcomas with all liposarcomas in most studies due to their small number, making region-specific analyses difficult. Little emphasis has been put on understanding liposarcomas in specific head and neck localizations, making treatment planning challenging. [11] Several review studies have listed cases with primary salivary gland liposarcomas, but none have attempted summarize the cases affecting the parotid gland only. [7,8,9] Surgical treatment of parotid liposarcoma differs significantly in comparison to other smaller salivary glands owing to its specific

position, surrounding complex anatomy and inherent individual lymphatic drainage pattern. All of these characteristics should be taken into consideration when planning a comprehensive resection of primary parotid liposarcomas. [8,9] Out of 8 individual cases, only 2 were the of the pleomorphic subtype, one patient being disease-free after 3 years of follow-up, and one with local recurrence, distant metastatic spread and death several months after initial treatment. [5,7,8] It has been reported that histologic subtype correlates with disease prognosis, with other factors including tumor size, site and distant metastatic spread. Published 5-year survival rates show 77% to 85% for mxoid and well-differentiated liposarcomas, but only 18% for pleomorphic liposarcomas, who also have a 73% rate of local recurrence. [10] The overall 5-year survival rate for all histologic subtypes is only 20% to 50%. [11] Available data for the parotid gland show that only 2 of the 8 reported patients have died of disease spread within the first year of diagnosis, with both of them affected by the pleomorphic subtype. One large review study suggests that patients with smaller tumors are more likely to have local recurrence but less likely to die of liposarcoma. In contrast, the same study found that patients with smaller tumors were more likely to have positive margins and no adjuvant therapy as part of initial treatment. Limited evidence suggests that oral and salivary gland, and head and neck primary liposarcomas in general have a somewhat better prognosis than other soft-tissue liposarcomas. Surgical excision with wide surgical margins is the primary therapy for liposarcomas and currently recommended as the most preferable treatment. The fascia surrounding the tumor is not a true enveloping layer, and excision margins should be at least 2 cm from the palpable tumor margin to avoid leaving microscopic residual disease behind. [9,10] Chemo and radiotherapy seem to have limited value, especially in cases where achieving adequate wide excision margins is difficult, such as in the head and neck area, but with responses rates limited to as low as 20% for doxorubicin and ifosfamide. Nonetheless, neoadjuvant chemotherapy has been advocated in patients with high-grade

tumors involving complex anatomical subsites. Adjuvant postoperative chemotherapy could be beneficial in patients with high-grade tumors. Postoperative radiotherapy remains an option for patients with high-grade tumors, positive margins, large tumors and involvement of complex anatomic subsites. [11,12]

Conclusion

Liposarcomas rarely develop in the head and neck region even more so in the parotid gland. The mainstay of treatment is a wide margin surgical excision, and the prognosis is determined by the histological grade and tumor size. Surgery alone could be sufficient to cure low-grade tumors, but high-grade tumors have a worse outcome despite the addition of surgery and postoperative radiotherapy.

Declaration of Conflicting Interests and Financial Disclosure

The authors declare no potential conflicts of interest with respect to the research, authorship, and publication of this article. All of the authors have read and approved the submission of the manuscript.

This research did not receive any specific grant from funding agencies in the public, commercial, or not-for-profit sectors.

References

1. Golledge J, Fisher C, Rhys-Evans PH. Head and neck liposarcoma. Cancer 1995:76:1051-1058.
2. Hornick JL, Bosenberg MW, Mentzel T, McMenamin ME, Oliveira AM, Fletcher CD. Pleomorphic liposarcoma: clinicopathologic analysis of 57cases. Am J Surg Pathol 2004:28:1257-1267.
3. Chandan VS, Fung EK, Woods CI, de la Roza G. Primary pleomorphic liposarcoma of the parotid gland: a case report and review of the literature. Am J Otolaryngol 2004:25:432-437.
4. Fanburg-Smith JC, Furlong MA, Childers EL. Liposarcoma of the oral and salivary gland region: A clinicopathologic study of 18 cases with emphasis on specific sites, morphologic subtypes, and clinical outcome. Mod Pathol 2002:15:1020-1031.
5. Korentager R, Noyek AM, Chapnik JS, et al. Lipoma and liposarcoma of the parotid gland: high-resolution preoperative imaging diagnosis. Laryngoscope 1988:98:967-971.
6. Gaskin CM, Helms CA. Lipomas, lipoma, and variants well-differentiated liposarcomas (atypical lipomas): results of MRI evaluations of 126 consecutive fatty masses. AJR Am J Roentgenol 2004;82:733–739.
7. Jones JK, Baker HW. Liposarcoma of the parotid gland: report of a case. Arch Otolaryngol 1980:106:497-499.
8. Gerry D, Fox NF, Spruill LS, Lentsch EJ. Liposarcoma of the head and neck: analysis of 318 cases with comparison to non-head and neck sites. Head Neck 2014:36:393-400.

9. Gritli S, Khamassi K, Lachkhem A, Touati S, Chorfa A, Ben Makhlouf T, El May A, Gammoudi A. Head and neck liposarcomas: a 32 years' experience. Auris Nasus Larynx 2010:37:347-351.
10. Davis EC, Ballo MT, Luna MA, Patel SR, Roberts DB, Nong X, Sturgis EM. Liposarcoma of the head and neck: The University of Texas M. D. Anderson Cancer Center experience. Head Neck 2009:31:28-36.
11. Seynaeve C, Verweij J. High-dose chemotherapy in adult sarcomas: No standard yet, Semin Oncol 1999:26:119-133.
12. de Bree E, Karatzanis A, Hunt JL, Strojan P, Rinaldo A, Takes RP, Ferlito A, de Bree R: Lipomatous tumours of the head and neck: a spectrum of biological behaviour. Eur Arch Otorhinolaryngol 2015;272:1061-1077.

Tables

Table 1. Demographic characteristics, localization, histopathologic subtype and follow-up of previously reported parotid gland liposarcomas.

Source		Fanburg-Smith et al.[4]					
	Chandan et al.[3]	Case 1	Case 2	Case 3	Korentager et al.[5]	Jones et al.[7]	Our Case
Age (years)	80	51	63	67	69	25	66
Sex	Female	Female	Male	Female	Male	Male	Male
Localization	Left parotid	Left parotid	Right parotid	Left parotid	Left parotid	Left parotid	Left parotid
Liposarcoma size	3.5 cm	3.5 cm	1.8 cm	5.5 cm	6.5 cm	10 cm	6 cm
Histology	Pleomorphic	Well differentiated	Myxoid	Dedifferentiated	Not specified	Pleomorphic	Pleomorphic
Follow up	Disease-free, 3 years	Disease-free, 2 years	Lost to follow-up	Disease-free, 17 years	Disease-free, 3 years	Died of metastatic disease after 2 years	Died of metastatic disease after 5 months
Treatment	Surgery	Surgery	Not available	Not available	Surgery	Sugery, Chemoradiotherapy	Surgery

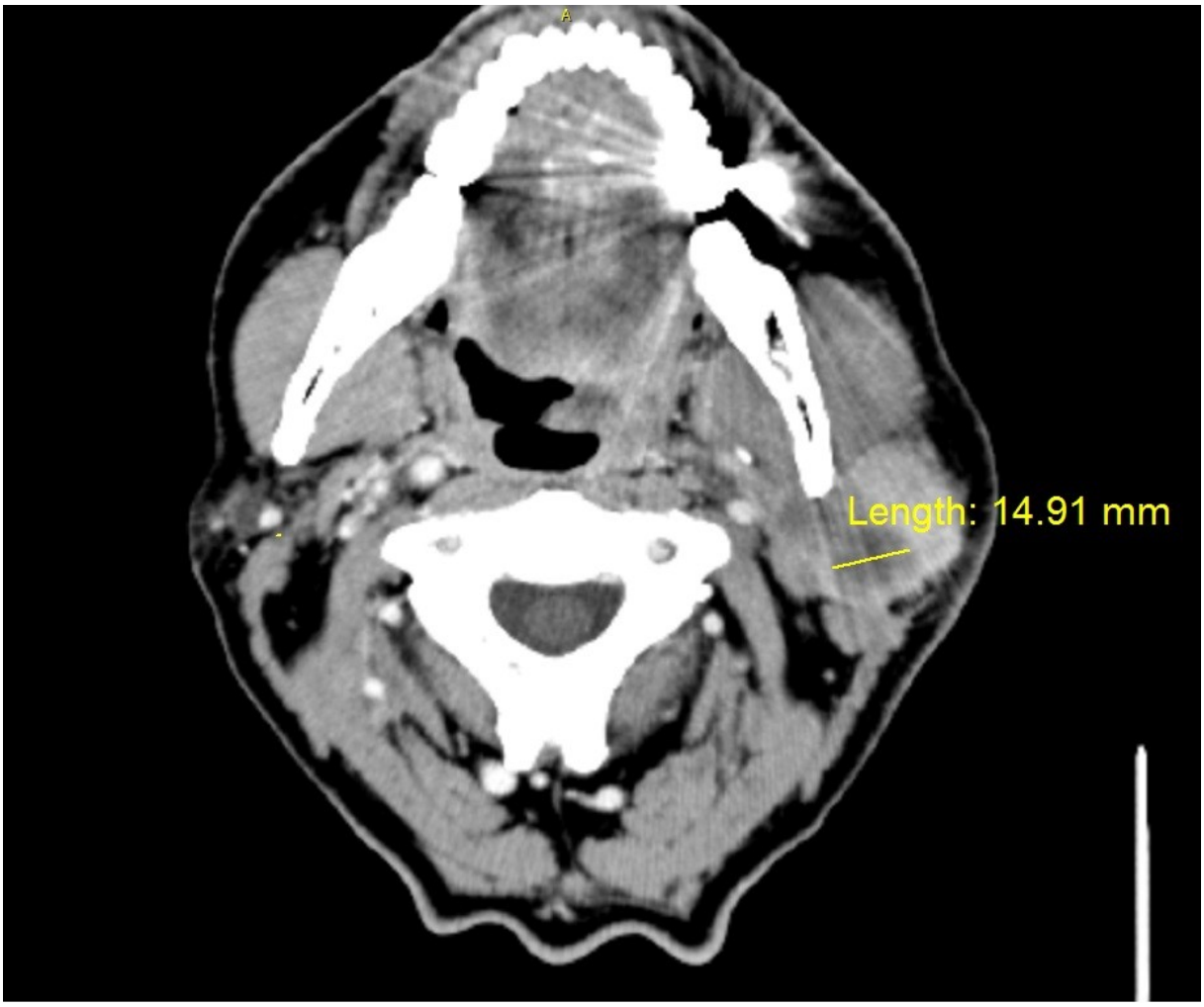
Figure Legends

Figure 1. Contrast enhanced computed tomography imaging showing an inhomogeneous infiltrative mass in the left parotid gland.

Figure 2. Infiltration of the facial nerve, both superficial and deep lobes of the parotid can be seen, with a surgical field planned for further wide margin resection.

Figure 3. Surgical field after a left-sided selective neck dissection encompassing neck regions I, II, III and Va, total parotidectomy, digastric muscle, exterior carotid artery and facial nerve resection and reconstruction using a great auricular nerve graft.

Figure 4. Primary parotid pleomorphic liposarcoma showing proliferation of pleomorphic malignant cells with evidence of lipomatous differentiation. Characteristic oval and multinucleated giant cells with vacuolated cytoplasm are present. (HE, 200x)


Length: 14.91 mm