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Case report

Primary parotid liposarcoma

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ABSTRACT

Introduction: Primary liposarcoma is very rare in the parotid gland. To date, only 8 cases of primary parotid liposarcoma have been reported. The aim of this study is to report on a case of primary parotid liposarcoma highlighting the complexity of its treatment and analyze treatment outcomes of other reported cases.
Case summary: We report a case of 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: Parotid liposarcoma 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. Postoperative radiotherapy is an option for patients with large high-grade tumors, positive margins and involvement of complex anatomic subsites. High-grade tumors have a worse outcome despite the addition of surgery and postoperative radiotherapy.

1. 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 parotid region are very rare, with only several previous cases reported. The histopathologic differential diagnoses should consider intramuscular myxoma, spindle cell or pleomorphic lipoma, lipogranuloma, and glioblastoma [2].

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 (Fig. 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 gross tumor infiltration, followed by great auricular nerve graft reconstruction (Figs. 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 (Fig. 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

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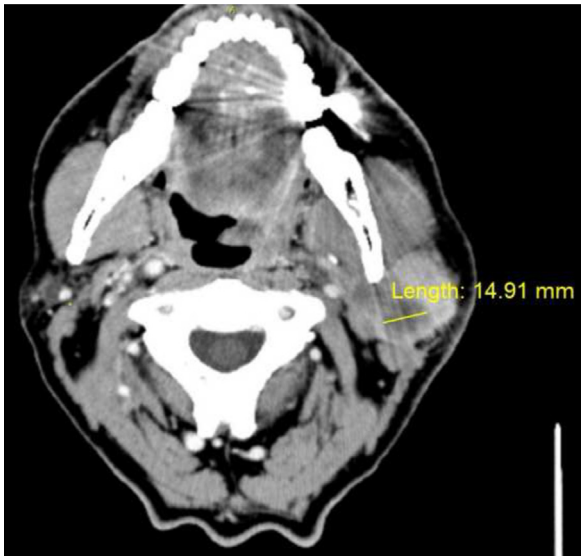

Fig. 1. Contrast enhanced computed tomography imaging showing an inhomogeneous infiltrative mass in the left parotid gland.

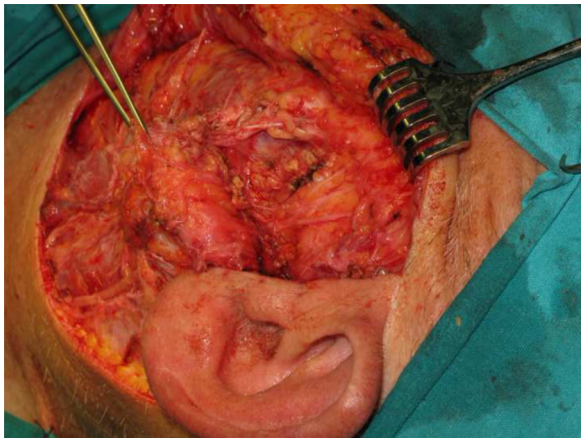

Fig. 2. Infiltration of the facial nerve, both superficial and deep lobes of the parotid can be seen, with a surgical field planned for further wide margin resection.

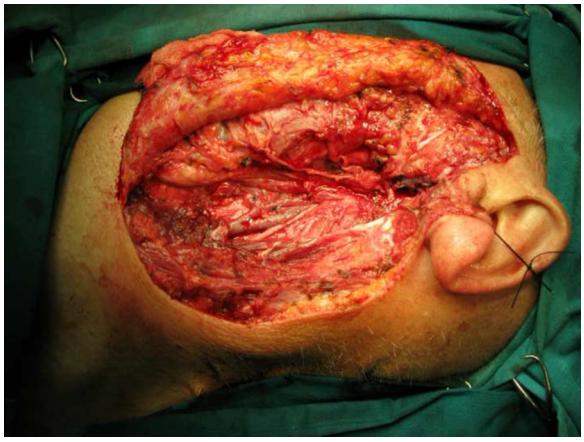

Fig. 3. Surgical field after a left-sided selective neck dissection encompassing neck regions I, II, III and Va, total parotidectomy, digastric muscle, exterior carotid artery and facial nerve resection and reconstruction using a great auricular nerve graft.

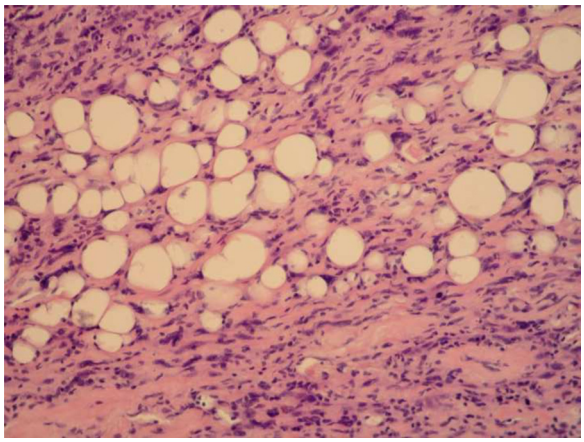

Fig. 4. Primary parotid pleomorphic liposarcoma showing proliferation of pleomorphic malignant cells with evidence of lipomatous differentiation. Characteristic oval and multinucleated giant cells with vacuolated cytoplasm are present (HE, 200 ×).

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.

3. 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 parotid area [3]. The World Health Organization classifies liposarcomas into 5 major histological subtypes:

- well differentiated – adipocytic, sclerosing, and inflammatory subtypes;
- dedifferentiated;
- myxoid;
- round cell;
- 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 [1,4–6]. 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 tumors. Of those, MRI is the preferred technique in establishing relations to adjacent structures of tumors considered suspicious for well-differentiated liposarcoma, but is not sufficiently 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

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

Source	Chandan et al. [3]	Fanburg-Smith et al. [4]			Korentager et al. [5]	Jones et al. [8]	Agarwal et al. [9]		Our case
		Case 1	Case 2	Case 3			Case 1	Case 2	
Age (years)	80	51	63	67	69	25	60	56	66
Sex	Female	Female	Male	Female	Male	Male	Male	Male	Male
Localization	Left parotid	Left parotid	Right parotid	Left parotid	Left 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	3.2 cm	1.1 cm	6 cm
Histology	Pleomorphic	Well differentiated	Myxoid	Dedifferentiated	Not specified	Pleomorphic	Pleomorphic	Well differentiated	Pleomorphic
Treatment	Surgery	Surgery	Surgery, incomplete excision	Surgery	Surgery	Surgery Chemotherapy Radiation	Surgery Radiation	Surgery Radiation	Surgery
Facial nerve	Frontal and zygomatic branches preserved	Preserved	Unknown	Preserved	Preserved	Preserved	Preserved	Preserved	Resected
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	Disease-free, 1 year	Disease-free, 1 year	Died of metastatic disease after 5 months

non-adipose tissue making up more than 25% of the lesion. CT scans better display calcifications in the lesion and its relation to bone structures. Fludeoxyglucose positron emission tomography (FDG-PET) may also help in diagnosing different liposarcoma types, but its accuracy is not sufficient to support the omission of diagnostic biopsy [7].

Little emphasis has been put on understanding primary parotid liposarcomas, making treatment planning challenging [8]. Several review studies have listed cases with primary salivary gland liposarcomas, but none have attempted to summarize the cases affecting the parotid gland only [8–12]. 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 [10,11].

It has been reported that the 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 myxoid and well-differentiated liposarcomas, but only 18% for pleomorphic liposarcomas, who also have a 73% rate of local recurrence [11]. The overall 5-year survival rate for all histologic subtypes is only 20 to 50% [12].

Only 8 reported cases of primary parotid liposarcoma were identified to date, with two additional studies listing the parotid and the neck as one indiscriminant area (Table 1). Out of 8 individual cases, 6 patients were disease-free, in regular follow-up. One patient was lost to follow-up soon after initial surgical treatment, and one patient had local recurrence, distant metastatic spread and died several months after initial treatment. Available data for the parotid gland show that only 1 of the 8 reported patients has died of disease spread. Both of the patients with metastatic disease were diagnosed with the pleomorphic liposarcoma subtype, which is consistent with known survival data. Ours is the only case that required complete facial nerve resection due to gross tumor infiltration. The majority of cases described the tumor in the infra-auricular, caudal or peripheral areas of the parotid gland, which allowed for facial nerve preservation. The average size of the primary tumor was 5.1 cm. It was most frequently localized in the left parotid gland. Postoperative radiotherapy was used in two cases with large high-grade tumors. In addition, highlighting varying treatment preferences, it was used in one patient with a small, well-differentiated tumor as well [3–5,8,9].

Gerry et al. reported on 13 cases of salivary gland liposarcoma and Davis et al. reported on 1 case of parotid gland liposarcoma, but without specific data on localization, histology, follow up or treatment [10,12]. That is often caused by grouping of parotid liposarcomas with all liposarcomas in most studies due to their small number, making region-specific analyses difficult.

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 [10]. Limited evidence suggests that oral and salivary gland, and primary parotid 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 [10–12]. 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, 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 [7,12,13].

4. Conclusion

Liposarcomas rarely develop 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.

Disclosure of interest

The authors declare that they have no competing interest.

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