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Pleomorphic Nasopharyngeal Liposarcoma: An Aggressive Tumor

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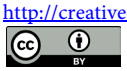


Abstract

Nasopharyngeal liposarcoma is an extremely rare clinical entity. Less than ten cases have been reported in the literature. It mainly occurs in humans. The non-specificity of the clinical and radiological signs explains the difficulty of the diagnosis confirmed by anatomopathological and immunohistochemical examinations. Its pathogenesis remains obscure and often correlates with genetic abnormalities. Pleomorphic liposarcoma is its rarest subtype, remarkable for its aggressiveness and rapidity of growth. Complete surgical excision remains the treatment of choice. We report a case of liposarcoma whose diagnosis was made during histological examination coupled with immunohistochemistry.

Keywords

Liposarcoma, Nasopharynx, Pathology, Immunohistochemistry

1. Introduction

Nasopharyngeal liposarcoma is an extremely rare clinical entity. Fewer than a dozen cases have been reported in the literature. It occurs mainly in men. Its pathogenesis remains obscure and is often correlated with genetic abnormalities. Liposarcoma pleomorphic (LSP) is its rarest subtype, remarkable for its aggressiveness and rapid growth. Complete surgical excision remains the treatment of choice [1].

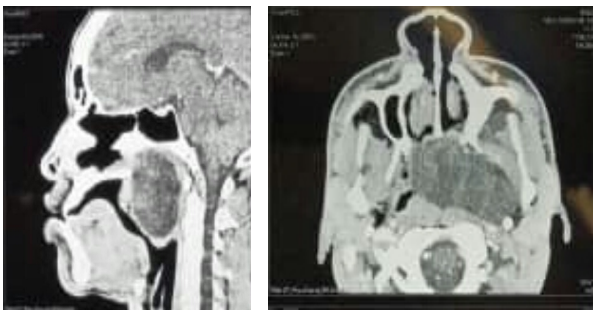
The aims of our study were to review the otorhinolaryngological manifestations of liposarcoma in an unusual location through an observation and to evaluate its management.

2. Clinical Case

A 30-year-old man with no particular pathological history had been presenting for 1 year with intermittent bilateral epistaxis of progressive aggravation associated with nasal obstruction and closed rhinolalia.

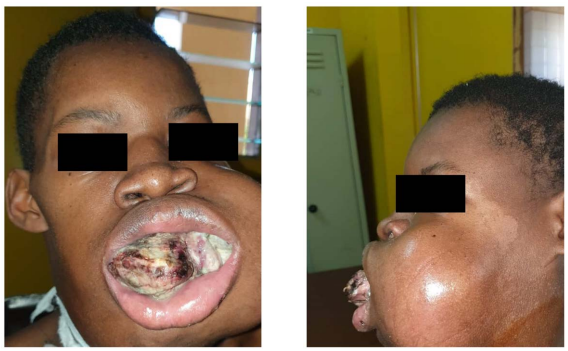
ENT examination revealed a left endonasal mass, limited mouth opening and retrovelar bulging. The lymph nodes were free. Cervicofacial CT scan with contrast injection revealed a calcified nasopharyngeal mass of mixed fluid and fat density extending into the oropharynx and left infratemporal fossa (Figure 1).

Panendoscopy revealed a voluminous, firm mass in the nasopharynx exteriorizing through the oropharynx and adhering to the posterior surface of the soft palate. Biopsies were taken. Histological examination of the biopsy specimen showed a grade III pleomorphic liposarcoma, confirmed by immunohistochemistry. No metastases were found in the extension study. Given the inoperability of the lesion, the patient underwent Carboplatin-Taxol-based chemotherapy and radiotherapy. After 5 months, the evolution was unfavorable, with a flare-up of the lesion externalizing through the mouth (Figure 2).



(a) (b)

Figure 1. Sagittal (a) and frontal (b) CT images showing the mixed density mass in the nasopharynx.



(a) (b)

Figure 2. Mass flare-up with expression in the oral cavity.

3. Discussion

Liposarcomas are malignant soft-tissue tumours with adipocytic differentiation. They arise mainly in the deep soft tissues of the limbs and retroperitoneum, in the form of slow-growing, painless masses. Very few cases have been described in the head and neck [1] [2]. Histologically, liposarcoma is divided into 4 groups: well-differentiated liposarcoma, dedifferentiated liposarcoma, myxoid liposarcoma and pleomorphic liposarcoma [3]. In our case, the patient presented with a pleomorphic liposarcoma discovered on histological examination and confirmed by immunohistochemistry. This is very rare, accounting for only 5% to 10% of liposarcoma cases [4].

The most common sites in the cervicofacial region are the cheek (14%), the anterior cervical region (13%), the larynx and piriform sinus (13%), the orbit (8%) and the supraglottic region (7%) [5].

A review of the English literature revealed only a few publications relating to liposarcomas of the head and neck, generally representing a limited series of patients. Nasopharyngeal liposarcoma is very rare; fewer than five cases have been described in the literature.

Diagnosis of liposarcoma is rare, and usually takes the form of a painless mass progressively increasing in volume. Signs of compression may be encountered, depending on the location and size of the tumor. In reported cases of nasopharyngeal lesions, the main symptoms were progressive in onset and marked by nasal obstruction accompanied by persistent unilateral seromucous otitis with reflex otalgia [6] [7]. Our patient presented with nasal obstruction associated with progressively closed rhinolalia, with an endonasal mass on examination.

Most authors agree that the imaging modality of choice is MRI [6] [8]. However, computed tomography (CT) is still recommended, particularly when data concerning bone or cartilage erosion are required. Although radiographic findings generally support the diagnosis, the imaging features of these lesions can vary considerably depending on the histological subtype and macroscopic appearance of the tumor. It is difficult to distinguish LSP from other liposarcomas and sarcomas on CT. LSP usually presents as a heterogeneous, poorly defined mass, containing fatty areas of density ranging from −50 to −150 Hounsfield units, coexisting with tissue islands of higher density, enabling them to be recognized [9]. A CT scan in our patient showed a mixed-density image.

Definitive diagnosis of LSP always depends on pathological examination, which reveals a proliferation of spindle-shaped cells, often with a fasciculated architecture, sometimes associated with epithelioid cells, round cells and multinucleated giant cells. Cytonuclear atypia is marked, with hyperchromasia and nuclear monstrosity. In places, specific differentiation such as osteosarcoma, chondrosarcoma and/or rhabdomyosarcoma may be observed, which may pose a differential diagnostic problem with these sarcomas. Pleomorphic lipoblasts with multivacuolar cytoplasm and bizarre notched nuclei are seen in variable numbers. Mitotic activity is usually high; most LSPs show extensive necrosis [4]

[9]. In our patient, the diagnosis was confirmed by histology and immunohistochemistry.

The management of liposarcoma of the head and neck has been largely based on experience with these tumors affecting the extremities and retroperitoneum [1] [2]. Wide surgical excision is considered appropriate for most low-grade liposarcomas. However, as it may be difficult or impossible to achieve complete excision of low-grade liposarcoma at depth, adjuvant radiotherapy and/or chemotherapy is used in these cases to reduce the risk of local recurrence [4] [6]. Treatment strategies for liposarcoma of the head and neck are generally deferential to those for similar lesions in other regions. This reflects anatomical limitations, as well as expected morbidity [1] [2]. The localization of the lesion in the head and neck region can lead to serious difficulties in surgical treatment. Radical excision, with sufficient healthy margins, is generally quite difficult to achieve. In addition, the expected postoperative morbidity is considered significant in many cases. Our patient benefited from chemotherapy.

Survival rates at 1, 3 and 5 years for LSP were 93%, 75% and 29% respectively [4]. Patients treated with surgery alone appear to have the best outcomes compared to patients treated with combined therapeutic modalities (5-year survival rate 83% vs. 63%) [1]. Golledge has argued that local recurrence is also less frequent in patients treated with surgery alone [1]. This observation obviously reflects the fact that the most aggressive tumors are often treated with combined strategies. However, in Davis' study [10], there was no statistically significant difference between patients treated with surgery alone and those treated with surgery and adjuvant therapies, either in terms of local recurrence or distant metastases.

Local recurrence is considered to be fairly frequent (reported recurrence rates range from 36% to 63%) [1] [2].

4. Conclusion

Pleomorphic nasopharyngeal liposarcoma remains an extremely rare and aggressive tumour. It poses therapeutic difficulties due to the impossibility of carcinological surgical excision, which remains the treatment of choice. Radiochemotherapy was ineffective in this patient.

Conflicts of Interest

The authors declare no conflicts of interest regarding the publication of this paper.

References

[1] Golledge, J., Fisher, C. and Rhys-Evans, P.H. (1995) Head and Neck Liposarcoma. *Cancer*, **76**, 1051-1058. https://doi.org/10.1002/1097-0142(19950915)76:6<1051::AID-CNCR2820760620>3.0.CO;2-4

[2] Gritli, S., Khamassi, K., Lachkhem, A., *et al.* (2010) Head and Neck Liposarcomas: A

32 Years Experience. *Auris Nasus Larynx*, **37**, 347-351. https://doi.org/10.1016/j.anl.2009.08.003

[3] World Health Organization, International Agency for Research on Cancer (2006) WHO Classification of Tumors of Soft Tissue and Bone. 4th Edition, IARC Press, Lyon.

[4] Wang, L., Luo, R., Xiong, Z., *et al.* (2018) Pleomorphic Liposarcoma: An Analysis of 6 Case Reports and Literature Review. *Medicine*, **97**, e9986. https://doi.org/10.1097/MD.0000000000009986

[5] Yueh, B., Bassewitz, H.L., Eisele, D.W., *et al.* (1995) Retropharyngeal Liposarcoma. *American Journal of Otolaryngology*, **16**, 331-340. https://doi.org/10.1016/0196-0709(95)90063-2

[6] Chakraborty, S., Kassel, E.E. and Yu, E. (2007) Nasopharyngeal Liposarcoma MRI Imaging Features and a Review of the Literature. *The British Journal of Radiology*, **80**, e145-146. https://doi.org/10.1259/bjr/49643955

[7] Nageris, B., Feinmesser, M., Brama, I., *et al.* (1991) Liposarcoma of the Nasopharynx: A Case Report. *Ear, Nose and Throat Journal*, **70**, 520-522.

[8] Papacharalampous, G.X., Kikidis, D., Vasileiou, A., *et al.* (2012) Liposarcoma of the Nasopharynx: Diagnosis and Management of a Rare Diagnostic Entity. *Case Reports in Medicine*, **2012**, Article ID: 314697. https://doi.org/10.1155/2012/314697

[9] Msaad, S., Yangui, I., Ayedi, L., *et al.* (2007) A Rare Primary Tumor of the Mediastinum: Pleomorphic Liposarcoma. *Revue de Pneumologie Clinique*, **63**, 373-378. https://doi.org/10.1016/S0761-8417(07)78424-9

[10] Davis, E.C., Ballo, M.T., Luna, M.A., *et al.* (2009) Liposarcoma of the Head and Neck: The University of Texas M. D. Anderson Cancer Center Experience. *Head and Neck*, **31**, 28-36. https://doi.org/10.1002/hed.20923