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# MALIGNANT FIBROUS HISTIOCYTOMA OF THE LOWER LIP: A CASE REPORT

Primljen/Received: 05. 04. 2012. god.

Prihvaćen/Accepted: 08. 05. 2012. god.

**Summary:** Malignant fibrous histiocytoma (MFH) represents the diagnosis that is still commonly used by both patients and physicians although in 2002., the World Health Organization (WHO) declassified MFH as a formal diagnostic entity, renaming it as an undifferentiated pleomorphic sarcoma not specifying it further. MFH is extremely rare in the oral cavity. The aim of this article was to describe a newly diagnosed case of primary MFH in a 78-year-old male presented after 4-months history of rapidly increasing swelling of lower lip. Histopathology of the lesion showed highly malignant cell infiltration with epithelioid/spindle cell type morphology. Immunohistochemical study was positive for vimentin (++), S-100 protein (++) and negative for desmin and cytokeratin. The authors discussed the possibilities of immunohistochemical study and necessity of individual treatment plan in patients with MFH, comparing their point of view with other studies.

**Key words:** malignant fibrous histiocytoma, undifferentiated pleomorphic sarcoma, oral cavity.

## INTRODUCTION

Malignant fibrous histiocytoma (MFH) was first introduced in 1961. by Kauffman and Stout. Three years later MFH was described by O'Brien and Stout. In 2002., the World Health Organization (WHO) declassified MFH as a formal diagnostic entity and renamed it as an undifferentiated pleomorphic sarcoma not otherwise specified (1). This new terminology has been supported by a compelling body of evidence to suggest that MFH represents a final common pathway

in tumors that undergo progression towards . However, the appellation of MFH is still considered to be an acceptable classification by WHO. Malignant fibrous histiocytoma (MFH) represents the diagnosis that is still commonly used by both patients and physicians.

Fibrous histiocytic tumors, benign or malignant, include spindle-celled neoplasms in which fascicles of cells exhibit a “storiform” arrange. The term “storiform”, derives from the ancient Greek language, and is, denoting a matted, irregularly whorled pattern, somewhat resembling that of a straw mat. The concept of fibrous histiocytic tumors is derived from the questionable assumptions in which histiocytes may act as facultative fibroblasts or those primitive mesenchymal elements and may give rise to both fibroblasts and histiocytes. Up to date, the posture view is that histiocytes or tissue macrophages are derived from blood monocytes originating in the bone marrow. Fibrous histiocytic tumors are composed of fibroblasts and histiocyte-like cells, as well as other cellular components such as myxoid, foam, inflammatory, and giant cells (2).

Five histological subtypes of MFH have been described including (I) storiform/pleomorphic (most common), (II) myxoid, (III) giant cell, (IV) inflammatory (usually retroperitoneal), and (V) angiomatoid (often located more superficially than other varieties) (3). MFH has also been associated with hematopoietic diseases such as non-Hodgkin lymphoma, Hodgkin lymphoma, multiple myeloma, and malignant histiocytosis. The malignant form of fibrous histiocytoma has attracted significant interest because of its relative frequency, can be present in numerous locations and having generally dim prognosis. The most common clinical presentation of MFH is an enlarging painless intramuscular soft-tissue mass, typically 5–10 cm in diameter. Rare signs and symptoms include episodic hypoglycemia and rapid tumor enlargement during pregnancy. The most frequent location of MFH is in the soft tissue-

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es, showing a higher incidence in the lower extremities, especially the thighs followed by the upper extremities and the retroperitoneum. The second most prevalent tissue for MFH is bone. MFH has been reported to occur in the lung, kidney, bladder, scrotum, vas deferens, heart, aorta, stomach, small intestine, central nervous system, paraspinal area, dura mater. Only 3% of cases have been reported in the head and neck. MFH is extremely rare in the oral cavity.

The aim of this article was to describe a newly diagnosed case of primary MFH with unusual localization.

### CASE REPORT

A 78-year-old male presented with 4-months history of rapidly increasing swelling of lower lip. There was no history of pain, fever, headache or dysphagia. On examination, the patient was a healthy looking individual with normal vital signs. Local examination of the swelling of lower lip showed soft to firm, non fluctuant, non tender and not adherent lesion to the overlying normal looking skin and mucosa (fig.1). The size of the formation was 3,0 x 3,0 cm (fig. 2a, fig. 2b). No



Figure 1. MFH of lower lip



Figure 2a. MHF after surgical excision

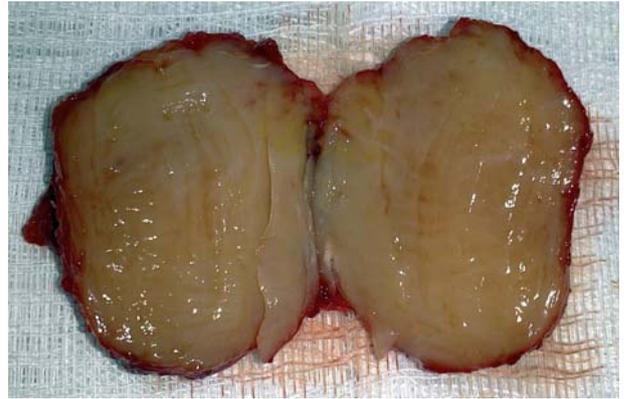


Figure 2b. MFH after surgical excision



Figure 3a. Intraoral status after surgical excision



Figure 3b. Extraoral status after surgical excision

regional lymph nodes were palpable. The treatment was wide local excision by intraoral approach, with surgical board extension of 1 cm (fig. 3a, fig. 3b). Histopathology of the lesion showed highly malignant cell infiltration with epithelioid/spindle cell type morphology, having abundant eosinophilic cytoplasm with hyperchromatic, pleomorphic nuclei (fig. 4a). Scattered multinucleate giant cells, abundant mitoses and necrosis were seen. Immunohistochemical study was positive for vimentin (++), S-100 protein (++) (fig. 4b) and negative for desmin and cytokeratin. The patient

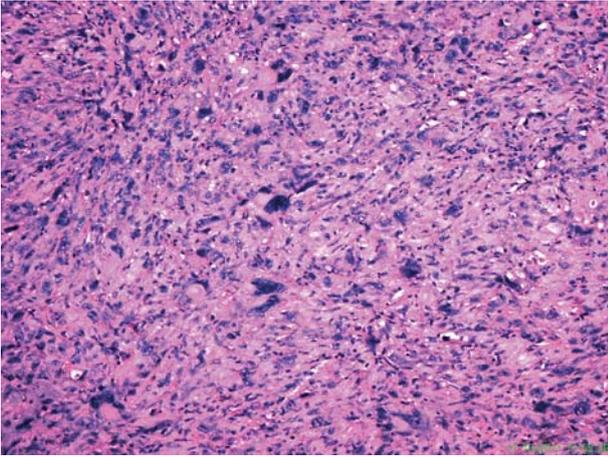


Figure 4a. Pathological view (HEx100)

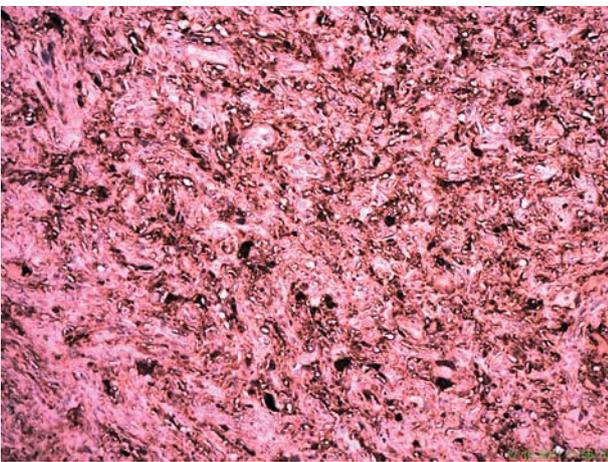


Figure 4b. Pathological view (S-100x100)

had an uneventful recovery after surgery and was clinically free of tumor at 18 months of follow-up.

## DISCUSSION AND CONCLUSION

MFH accounts for 20–24% of soft-tissue sarcomas in adults. MFH most commonly occurring between age 50–70. A review of the literature yielded only few cases of primary MFH to the soft tissues of the oral cavity (4, 5, 6).

Lawson et al. studied 10 ultra structurally -defined cases of MFH and found that all labeled for vimentin. Additionally six cases showed expression of desmin and neurofilaments (7). In 1989 Hirose et al. examined the expression of intermediate filaments in 34 cases of MFH and found 30/34 to be vimentin positive, 12/34 to be desmin positive, 2/34 to be neurofilament positive and 1/34 to be cytokeratin positive (8).

Kamatani et al. (9) reported strong and diffuse immunohistochemically positivity for CD34 and vimentin within the cytoplasm of the tumor cells. They noti-

ced positive expression for S-100 protein. Even some of the acinar cells were immunopositive for S-100 protein, although the peripheral cells of acinar structures were simultaneously actin-immunopositive, suggesting their myoepithelial differentiation. The authors explained these unusual staining patterns for S-100 protein as indicator that the entrapped salivary gland components in the tumor were neither normal in shape nor in function.

There are essentially three main types of treatment that will need to be coordinated to treat the MFH: surgery, radiation, chemotherapy. Sabesan et al. (10) defined the management of MFH of the head and neck as a technical challenge that is dictated by the anatomical and functional complexity of the head and neck, and the rarity of MFH in this region intensifies this challenge. They suggest that the initial operation for this tumour in the head and neck should be as radical as possible. Biopsy is often necessary to make a diagnosis. Early and complete surgical removal using wide or radical resection is indicated because of the aggressive nature of the tumor. Radiation therapy decrease the incidence of local recurrence and become an integral part of the treatment for MFH. Barker et al. (11) reported that preoperative radiotherapy or chemotherapy can render the marginally operable tumours respectable with tumour-free margins, and postoperative radiotherapy improves local control, particularly in patients with close or involved surgical margins. In a prospective randomized trial, 91 patients with MFH were randomized to surgery alone or surgery with post-operative external beam radiation therapy (XRT) (12). Patients in the surgery alone group experience 20% local recurrence rates compared to 0% for the surgery plus XRT group. For both groups, there was no difference in overall survival. In general, patients treated with adequate limb-sparing surgery supplemented with radiation have a likelihood of experiencing over 85% local control. The role of chemotherapy in the treatment of MFH is not entirely clear. Prognostic factors correlate with survival in patients with MFH include tumor grade, depth, size, metastatic status, patient's age, and histologic subtype. Favorable prognostic factors include age less than 60 years old, tumor size less than 5 cm, superficial location, low grade, the absence of metastatic disease, and a myxoid subtype. The literature data suggest that overall linear trend of mortality is increasing (13).

In conclusion, MFH of soft tissues of oral cavity is very rare entity, indistinguishable clinically from other tumors, and diagnosed by wider spectrum of immunostains as well as electron microscopy.

## Sažetak

## MALIGNI FIBROZNI HISTIOCITOM DONJE USNE: PRIKAZ SLUČAJA

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Maligni fibrozni histiocitom (MFH) se kao dijagnoza još uvek često koristi iako je 2002. godine Svetska Zdravstvena Organizacija (WHO) deklasifikovala MFH kao formalni dijagnostički entitet, preimenujući ga u nediferentovani pleomorfni sarkom. MFH je izuzetno redak u usnoj duplji. Cilj ovog rada je da prikaže slučaj primarnog MFH kod muškarca starog 78 godina, otkrivenog nakon četvoromesečnog rasta otoka donje usne. Histopatološka analiza lezije je pokazala malignu ćelij-

sku infiltraciju sa morfolgijom epteloidno/vretenasto ćelijskog tipa. Imunohistoheмиjska analiza je bila pozitivna za vimentin (++) , S-100 protein (++) i negativna za dezmin i citokeratin. Autori analiziraju mogućnosti imunohistoheмиjskih ispitivanja i ističu neophodnost individualnog plana lečenja pacijenata sa MFH upoređujući svoje nalaze i stavove sa drugim studijama.

**Ključne reči:** maligni fibrozni histiocitom, nediferentovani pleomorfni sarkom, usna duplja.

## REFERENCES

1. Fletcher CDM, Unni KK, Mertens F, editors. World Health Organization Classification of Tumors. Pathology and Genetics of Tumors of Soft Tissue and Bone. Lyon: IARC Press; 2002.
2. Akerman M. Malignant fibrous histiocytoma — the commonest soft tissue sarcoma or a nonexistent entity? *Acta Orthop Scand Suppl* 1997; 273: 41–6.
3. Rosenberg AE. Malignant fibrous histiocytoma: past, present and future. *Skeletal Radiol* 2003; 32: 613–8.
4. Nuamah IK, Browne RM. Malignant fibrous histiocytoma presenting as perioral abscess. *Int J Oral and Maxillofac Surg* 1995; 24(2): 158–159.
5. Chen YK, Lin LM, Lin CC. Malignant fibrous histiocytoma of the tongue. *J Laryngol Otol* 2001; 115(9): 763–5.
6. Agnihotri R, Bhat KM, Bhat GS. A rare case of malignant fibrous histiocytoma of the gingiva. *J Periodontol* 2008; 79(5): 955–60.
7. Lawson CW, Fisher C, Gatter KC. An immunohistochemical study of differentiation in malignant fibrous histiocytoma. *Histopathology* 1987; 11: 375–83.
8. Hirose T, Kudo E, Hasegawa T, Abe J, Hizawa K. Expression of intermediate filaments in malignant fibrous histiocytoma. *Hum Pathol* 1989; 20: 871–7.
9. Kamatani T, Horie A, Ishii H, Kasahara H, Hamada Y, Aida Y, Shintani S. Primary malignant fibrous histiocytoma of the mandible: Report of a rare case with an immunohistochemical analysis. *Asian Journal of Oral and Maxillofacial Surgery* 2010; 22(4): 212–215.
10. Sabesan T, Xuexi W, Yongfa Q, Pingzhang T, Ilankovan V. Malignant fibrous histiocytoma: Outcome of tumours in the head and neck compared with those in the trunk and extremities. *British J Oral and Maxillofac Surg* 2006; 44: 209–212.
11. Barker Jr JL, Paulino AC, Feeney S, McCulloch T, Hoffman H. Locoregional treatment for adult soft tissue sarcomas of the head and neck: an institutional review. *Cancer J* 2003; 9: 49–57.
12. Yang JC, Chang AE, Baker AR, Sindelar WF, Danforth DN, Topalian SL, DeLaney T, Glatstein E, Steinberg SM, Merino MJ, Rosenberg SA. Randomized prospective study of the benefit of adjuvant radiation therapy in the treatment of soft tissue sarcomas of the extremity. *J Clin Oncol* 1998; 16(1): 197–203.
13. Belal A, Kandil A, Allam A, Khafaga Y, El-Husseiny G, El-Enbaby A, Memon M, Younge D, Moreau P, Gray A, Schultz H. Malignant fibrous histiocytoma: a retrospective study of 109 cases. *Am J Clin Oncol* 2002; 25: 16–22.

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