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MALIGNANT FIBROUS HISTIOCYTOMA – A CONFUSING ENTITY

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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.

INTRODUCTION

Malignant fibrous histiocytoma (MFH) was first introduced in 1961. by Kauffman and Stout. 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] 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 hematopoetic 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 tissues, 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.

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 noticed 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.

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