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Lethal midline granuloma: A case of T-cell lymphoma

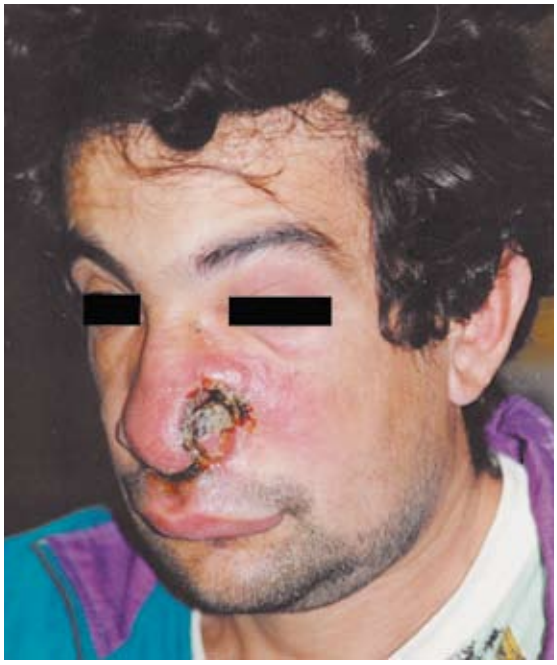

Fig. 1

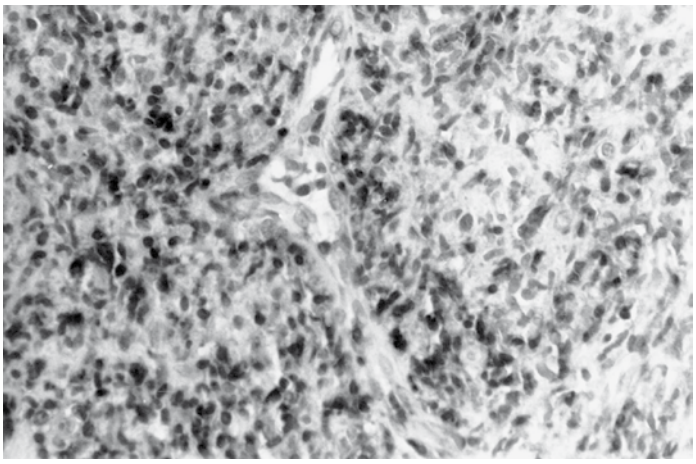

Fig. 2

A 34-year-old man referred to our hospital with necrotic granulation tissues in the nasal cavities and ulcerative destructive change of the palate with necrotic tumor (Fig. 1). Anterior rhinoscopic examination showed necrotic granulation tissues in the nasal cavities with destruction of the columella and septum nasi.

Computed tomography of the paranasal sinuses showed osteolisis of ethmoid anterior cells, nasal septum, vomer, and concha inferior and penetration to the left orbita. Immunhistologic study of the specimens from the nasal lesion revealed that immature or atypical cells had phenotypes of T-cells. Polymorphous cellular infiltrates were found containing large atypical cells with positive reaction to CD3 and negative reaction to markers for CD20 (Fig. 2). The presence of Epstein-Barr virus genome in the tumor cells was assessed by monoclonal antibodies (EBV-LMP 1, Dako).

The patient was irradiated with 4-MV X-rays using a three field isocentric technique to treat a target volume including the nasal cavity, nasopharynx, paranasal sinuses, oropharynx, larynx and palate. Irradiation therapy with a total dose of 56 Gy yielded an extremely good therapeutic result.

Lethal midline granuloma (LMG) is a clinical entity characterized by progressive, unrelenting ulceration and necrosis of the nasal cavity and midline upper respiratory structures. The term LMG designates a syndrome and not a specific form of disease. With respect to histopathology the following entities have been grouped under the term LMG: Wegener granulomatosis, non-Hodgkin lymphoma, polymorphic reticulosis and idiopathic midline destructive disease [1,2].

Neven Skitarelic

References

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2. Barker TH, Hosni AA (1998) Idiopathic midline destructive disease – does it exist? J Laryngol Otol 112: 307–309

Key words: T-cell lymphoma, lethal midline granuloma.

Correspondence: Neven Skitarelic, MD, PhD, Department of Otorhinolaryngology Head and Neck Surgery, Put Murvice 33, 23 000 Zadar, Croatia, E-mail: neven.skitarelic@zd.htnet.hr