INVOLVEMENT OF EXTRAOCULAR MUSCLES IN MYCOSIS FUNGOIDES

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Extracutaneous involvement in mycosis fungoides is not uncommon as evidenced by recent studies. Involvement of various parts of the eye by mycosis fungoides has been reported previously, though that of the extraocular muscles has not been described. We report here an unusual case of involvement of the lateral rectus muscle by mycosis fungoides.

Key Words : Mycosis fungoides

Introduction

Mycosis fungoides is a low grade cutaneous T-cell lymphoma. It pursues an indolent but progressive course. Dissemination of mycosis fungoides to extracutaneous sites was once considered uncommon, but visceral involvement has been observed in 61-82% of autopsied patients.[1] There are conflicting reports regarding the precise incidence and involvement of extracutaneous sites. The involvement of extraocular muscles is extremely rare. We are presenting an unusual case of superior and lateral rectus muscle involvement in mycosis fungoides. To the best of our knowledge, it has not been reported earlier.

Case Report

A 52-year-old male was seen with complaints of scaly skin rash on his legs since 12 years. He had noticed new elevated lesions and ulceration of some of them on the right chest wall since 9 months.

Physical examination did not reveal any significant findings except for the skin lesions. Examination of the skin showed large, scaly erythematous patches. In addition, there were multiple red and brown plaques and nodule some with central ulceration involving t trunk and extremities. Haematologic radiological, biochemical and an abdomin ultrasound examination did not reveal a abnormality.

Incisional biopsy from the tumo revealed a picture typical of mycosis fungoide with the epidermis showing epidermotropis and Pautrier microabscess (Fig. 1). Patient w staged according to the mycosis fungoide cooperative group (MFCG) classificatio system as stage IIB.[2]

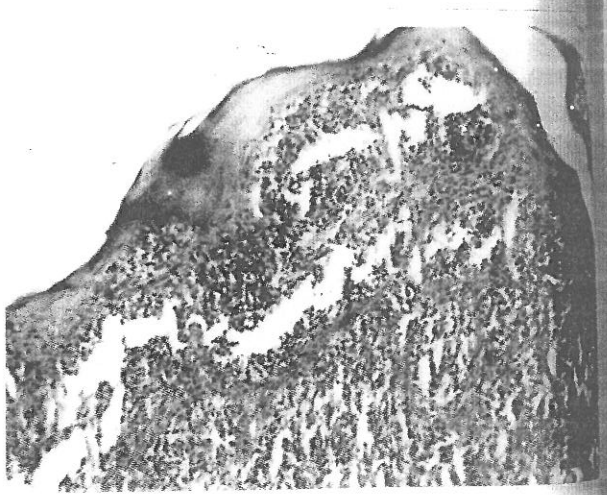

Fig. 1. Microphotograph illustrating epiderm tropism and Pautrier microabscess (H E x 125).

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The skin nodules and large tumours on the chest wall were treated with telecobalt-60 therapy, delivering a dose of 36GY fractionated over 9 weeks with a weekly fraction of 4GY. There was complete regression and healing of the ulcers. In the meantime, chemotherapy with cyclophosphamide, vincristine and prednisolone was started and administered at 3 weekly pulses. He received a total of 5 course of chemotherapy and remained free of all skin lesions. He was lost for follow up for a period of 4 months.

He again presented with nodules at regions other than the sites treated earlier. He also complained of diplopia. Examination revealed restricted movements of the right eye ball. Physical examination revealed only a mild hepatomegaly which was confirmed by an ultrasound scan. Routine blood tests, bone marrow aspiration and chest roentgenogram were normal. CT scan of the orbit and brain revealed infiltration of the lateral and superior rectii muscles of the right eye (Fig. 2). The cranial nerves were normal and there was no intracranial involvement. A biopsy of the lateral rectus muscle showed normal epidermis with areas of atrophic epithelium. Subepithelial connective tissue including the skeletal muscle fibres show diffuse infiltration by monomorphous cells. Some cleaved cells and a few larger cells with prominent nuclei were seen suggestive of cutaneous lymphoma involving the muscle. With external radiotherapy to the right orbit and chemotherapy with cyclophosphamide, vincristine and prednisolone eyeball movements improved but patient died within 6 weeks.

Comments

The clinical course of mycosis fungoides begins typically with a premycotic eruption

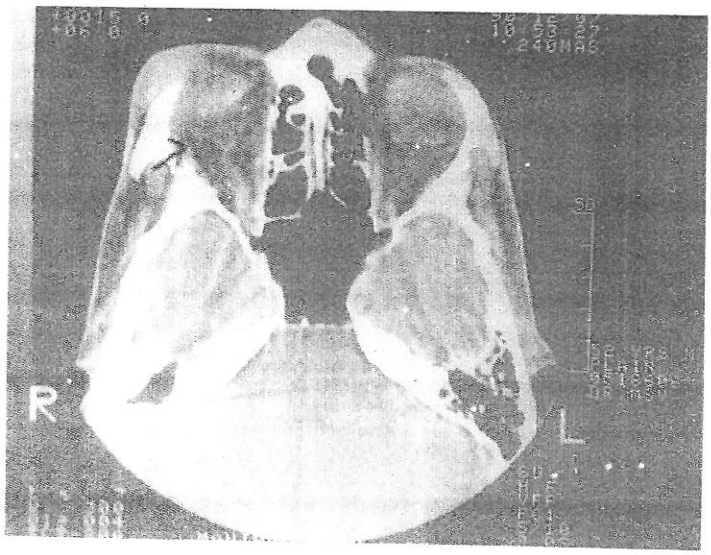

Fig. 2 CT scan showing infiltration of the lateral and superior rectii muscles of the right eye

which waxes and wanes over a period of many years. Subsequently cutaneous plaques and tumours occur. Visceral involvement is found frequently (72%) after the appearance of the tumour stage.

Extracutaneous presentation of mycosis fungoides is usually seen in the terminal stages of the disease. It is most commonly observed in lymph nodes, spleen, lung, liver and kidney.[1] Although the disease is commonly observed in various parts of the eye such as lid, cornea, conjunctiva, choroid and rarely retina; the available literature has not shown the involvement of extraocular muscles.

The initial diagnosis of mycosis fungoides in our patient was established based on the microscopic appearance showing the presence of epidermotropism, formation of Pautrier microabscess and the presence of mycosis

cells. As the disease progresses from the superficial to the tumour stage there will be a natural evolution resulting in the absence of epidermotropism and Pautrier microabscesses and the infiltrate mainly consists of mycosis cells.[3] In our case the histological diagnosis of the lateral rectus muscle involvement was made based on the presence of infiltration of the skeletal muscle fibres by monomorphous cells and cleaved cells.

In general, patients with extracutaneous involvement of mycosis fungoides carry a grave prognosis. The average duration of survival from time of diagnosis of extracutaneous involvement to death ranged from 7.5 months to 18 months.[1,3-5] The survival for the subgroup of patients with ocular involvement has not been reported separately from patients with extracutaneous mycosis fungoides in other sites.

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