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Case Report Open Access

Myxoid Liposarcoma of the Skin-Rare Case with Multifocal Presentation

Shiv Chopra*, Aman Chopra, Asok Aggarwal and Reshma Yadav

Inderprastha Apollo Hospital, New Delhi, India

***Corresponding author:** Dr. Shiv Chopra, Senior consultant Surgeon, Department of surgery, Inderprastha Apollo Hospital, New Delhi, India, E-mail: drshivchopra@apollohospitalsdelhi.com; sbchopra2004@yahoo.com

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Abstract

The dermis is an exceptionally rare primary site of presentation of liposarcomas. Only two cases of the myxoid variety have been reported in the literature. The lesions present as dome shaped or polypoidal masses that frequently show high grade morphological changes, but carry a comparitively a good prognosis. We report a 35 year old male who presented with multiple "ginger tuber" like lesions on the upper half of the body. The clinical behaviour was extremely aggressive with a short history of short duration.It is important to recognise the fact that though dermal liposarcomas by and large carry a good prognosis there exists a rare subset with a "multifocal" presentation that suggests a poor clinical course even though the histopathological features may show a high grade pathology suggesting a good prognosis. These cases require urgent treatment with radiotherapy and chemotherapy, where surgery may have no role in therapeutics. It is for this reason that we are presenting this rare subset of “multifocal myxoid dermal liposarcoma” which should alert all clinicians to act with speed.

Keywords: Cutaneous myxoid liposarcoma; Signet ring variant; Dermal liposarcoma; Ginger-tuber like; Multifocal dermal liposarcoma; Wildfire Hurricane

Introduction

Cutaneous myxoid liposarcoma are rare tumours presenting as painless swellings in the extemeties and scalp. They have an indolent clinical behaviour, with no metastasis and good prognosis. In our case however the tumour metastized rapidly from the outer canthus right eye and the patient presented with multiple painful “ginger tuber “like nodules and papules in the face, neck, chest and back. The clinical history was of a short duration and a downhill course with cachexia prompted an urgent referral to oncology/radiotherapy department of tertiary care hospital for urgent further management.

Case History

A 35 year old male presented with multiple painful lesions “ginger tuber “like all over the upper half and fading out over the lower half of the body, (Figures 1 and 2) since 4 months. These were accompanied by severe arthralgias, fever & weakness since 2½ months, loss of appetite 2 months, weight loss of 25% over 4 months (62.0 kg to 47.0 kg on 19/08/2008). The first lesion started out as a small nodule over outer canthus right eye, which increased to its present shape of a ginger tuber 5 × 3 cm size. The lesions varied from 0.5 to 6.0 cm and showed rapid growth. Lesions were yellowish pink with areas of ulceration, haemorrhage with irregular bossellated surfaces. There was pallor, chest was clinically clear; per abdominal examination including per rectal examination was within normal limits. The patient took homeopathic treatment 3 months ago after which the disease spread. H/0 Bidi/cigarettes, Tobacco chewer × 20 years, no intake of alcohol. The patient gave a clinical appearance typical of “von recklinghausens disease”, but the short history suggested malignancy with metastasis. HIV1 and 2 negative, VDRL-non-reactive. He underwent excision biopsies from two different sites. On histopathological examination, the sections showed a tumour mass that occupied most of the dermis, composed of groups of cells with round to oval hyperchromatic nuclei surrounded by a rim of cytoplasm. Majority of these cells showed a single intra-cytoplasmic vacuole resulting in the typical signet-ring appearance. The stroma was myxomatous with a delicate framework of plexiform capillaries with mild nuclear atypism, but no mitotic activity was present. Sparse lymphocytic infiltrate was seen in surrounding tissues. Seeing his rapid hurricane like spread, multifocal presentation suggestive of cutaneous metastasis, he was referred to the oncologist/radiotherapist for further urgent treatment by radiotherapy/chemotherapy.

Figure 1: Clinical photograph-front view.

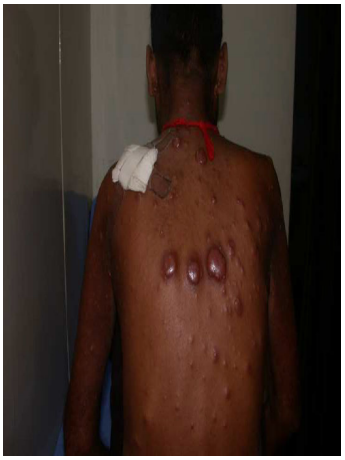

Figure 2: Clinical photograph-back view with dressing over the biopsy site over left shoulder.

Discussion

Myxoid liposarcomas represent approximately 10% of all soft tissue sarcomas comprising the second most common variant of liposarcoma's, presenting in the 4th to 5th decade with a slight male predominance [1,2] tending to occur on the right side of the body [2]. Liposarcomas are generally deep seated tumours most likely in the retroperitoneum, or the deep soft tissues in or below the buttocks and that only very rarely do they arise in the cutis or the dermis [3]. Primary liposarcoma of the skin has been described by Del Tos, in 7 cases, 4 of which arose in the scalp and only one was a myxoid/round cell liposarcoma [3,4]. Newlands in 2003 described the 2nd case of myxoid liposarcoma of the skin occurring on the scalp of a 28 year old lady [5]. The myxoid liposarcoma display various quantities of four elements (a) proliferating lipoblasts, (b) delicate plexiform capillaries, (c) myxoid matrix, (d) mucoplysaccharide lakes- (Figures 3 and 4) [2].

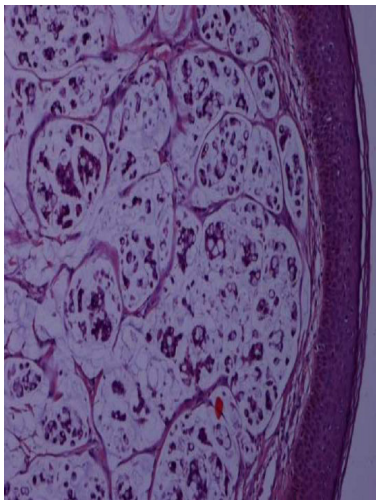

Figure 3: Magnification: 10X-Dermal Myxoid Liposarcoma signet cellvariant.

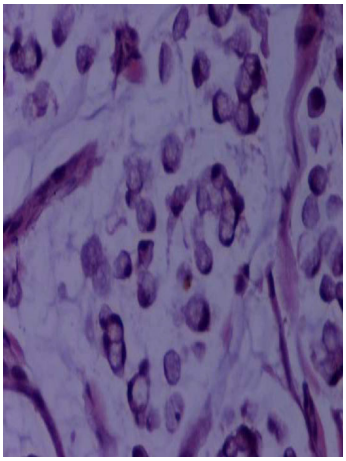

Figure 4: Magnification: 40X Showing signet cells in myxomatous stroma within a delicate framework ofplexiform capillaries.

Mitotic figures are conspicuously absent. These Myxoid tumours are further subdivided into a well-differentiated type with the so called signet ring cells, representing adult lipocytes, relatively indolent course with a good prognosis, little tendency to metastasize and a poorly differentiated type that commonly metastasizes [2-4,6]. The round cell variant, although classified by WHO as separate from myxoid, is often regarded as the poorly differentiated form of the myxoid liposarcoma [3,6]. Dermal liposarcomas may also have unusually high-grade cytological features resembling pleomorphic liposarcomas but metastasis have not yet been recorded [3,4,8]. Tumours with a multifocal presentation are defined as presence of tumour at two or more anatomically separate sites, before the manifestation of disease in sites where sarcomas normally metastasize (eg. lungs) .They occur in 1% of extremity soft tissue sarcomas [1-3,8] and have not been earlier described for dermal liposarcomas starting from the face. Among these multifocal liposarcomas which are now generally regarded as an unusual pattern of metastatic deposits [1,2,8], myxoid liposarcoma is the predominant histological type and this subtype contains a specific t (12:16) chromosomal derrangement [8]. This small series of 7 myxoid liposarcomas with multifocal presentation out of 43 consecutive cases of myxoid liposarcomas showed that in 6 of these 7 multifocal patients, had the same derangements showing thereby the monoclonality of these myxoid liposarcomas [8]. This unusual clinical phenomenon most likely represents haematogenous metastasis to the soft tissue sites, by tumour cells seemingly incompetent to seed the lungs which are the usual site of metastasis [8]. This clinical phenomenon was also seen in our case of these "ginger-tuber" like metastasis, which seemingly spread like a wildfire hurricane !. According to the current literature his clinical presentation did not tally with his histopathological features, which were not adverse [2-4,6,8]. We would like to add that even dermal liposarcomas myxoid type signet ring variant can have a rapidly downhill course when presenting with multiple cutaneous metastasis and chemotherapy/ radiotherapy offer hope of survival [6,7,9]. Our present case highlights the following point-that inspite of histological features suggesting a good prognosis, the clinical presentation of multiple metastatic deposits and rapid onset of disease and short duration are still more important. Because the prognosis is poor, multifocal myxoid liposarcoma should be regarded as a separate subset requiring urgent

diagnosis and speedy treatment [1,2,8,9]. All clinicians must be aware of this dreadful but rare subset.

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