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Dercum's Disease: A Rare Clinical Presentation of Multiple Lipomas

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Abstract: *Lipoma is a painless soft tissue tumour of the mesenchymal origin, which is slow growing and well circumscribed. They are usually asymptomatic. Dercum's disease is characterised by multiple lipomatous growth which are painful in nature. The classical treatment includes surgical resection, though liposuction can be a viable option of treatment. This report describes a case of multiple lipomas in a young female, who has been offered a trial of medical management, in accordance with her economic background.*

Keywords: Dercum's disease

I. INTRODUCTION

Lipoma is a benign adipocyte metabolic disorder, which is most common in soft tissue mesenchymal neoplasm. They can be found anywhere in the body with approximately 15 to 20% located in the head and neck region and the majority of the rest in the shoulder and back [1]. The lipoma usually occurs as a solitary lesion that may be sessile, pedunculated or submerged [2]. Dercum's disease is characterised by multiple such lipomatous growth which are painful in nature, associated with obesity.

1.1 Case Presentation and Treatment

A 27-year-old married female of Jharkhand origin, presented to the OPD with multiple swelling in both arm, forearm, shoulder, abdomen and inner thighs, some of which were painful. The history revealed that the swellings gradually increased over the course of last couple of years. The swellings were not associated with any itching, pus discharge, redness or scaling. The patient denied a history of unintentional weight loss. Her menstrual cycles are uneventful. On palpation, multiple well-delineated, mildly tender, mobile and soft masses were felt, most evident in the arm & forearm. The overlying skin was normal. Since plain anteroposterior radiograph of the forearm showed no invasion of the bone (thus excluding parosteal lipoma from the differential diagnosis) and palpation was inconclusive, ultrasound was ordered for further evaluation, under the care of Dr Mayukh Bhattacharya. The ultrasonographic evaluation revealed multiple clearly-defined well organised areas of hetero-echogenicity, strongly in accordance with lipoma. Two specimen smears were prepared from the left lower arm and right forearm nodule, and were sent for histopathological analysis. The histopathological examination of specimens from the left lower arm revealed fat droplets and occasional fat cells, and that from the right forearm revealed fat droplets, occasional fat cells and very scanty mature adipose tissue fragments. Overall cytomorphology of both specimens were suggestive of lipoma. Her biochemical parameters like prolactin, TSH, FT4, sodium, potassium, creatinine and lipid profile, were tested and found within normal physiological range.

All the possible surgical methods of treatment have been properly explained to her, but her economic background refrained her from surgical interventions. In accordance with her wishes and economic background, the patient has been offered an initial trial of medical management with a **high dose of Atorvastatin, Vitamin E and Ezetimibe.** She has been asked to follow up every third month to check the progression of the disease.

II. DISCUSSION

Lipomas are benign adipose tumours of mesenchymal origin secondary to hamartomatous proliferation of mature fat cells. Lipomas are classified as subcutaneous type, subfascial type or intermuscular type [3]. Lipids unavailable for metabolism coupled with autonomous growth of lipoma have rendered to be a true benign neoplasm. Occasionally, the

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lipoma may invade muscles or grow between them, the so-called infiltrating lipoma. It is an uncommon mesenchymal neoplasm that characteristically infiltrates adjacent tissues and tends to recur after excision. This type of lipoma is extremely rare in the head and neck region, and its congenital type is rare. They are usually non-tender, soft and almost 'cheesy' in the consistency, but may be fluctuant. It is typically superficial in depth; but may infiltrate the muscle, become fixed to surrounding tissues and therefore unmovable. Deeply occurring lesions may produce only a slight surface elevation and may be well encapsulated, more diffuse and less delineated than the superficial variety. Thus, more diffuse form generates the clinical impression of a fluctuant tumour [4]. The size, location, and pace of development of the lesion all have an impact on the clinical characteristics. Like the majority of benign tumours, they first appear as painless, mobile, palpable lumps that patients frequently ignore until they develop into a noticeable mass, as in our case. Rapid progressive growth should always highlight the possibility of malignancy. The onset of Dercum's disease can be insidious, progressive and mostly associated with obesity and rapid weight gain. Some cases of Dercum's disease may occur in one or more people in a family with familial multiple lipomatosis.

The first imaging modality used to diagnose lipomas is ultrasonography, with histopathological examination being the gold standard to confirm the diagnosis. Given that lipomas frequently pose a challenge during surgery and necessitate scrupulous skills due to their proximity to vital structures, this procedure should only be performed on patients who are experiencing cosmesis and pressure effects. To stop a recurrence, a full excision with a capsule should be done.

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