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Case Report

Clinical Mimicry and Dermatofibrosarcoma Protuberans: A Case Report with a Point to Ponder

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Abstract: Dermatofibrosarcoma protuberans (DFSP) is a very rare cutaneous neoplasm which usually affects the torso. The clinical presentation is nonspecific and can mimic many benign as well as malignant skin lesions. We present a case with clinical diagnostic dilemma which was located over an unusual site to be suspected as DFSP with deceiving feature of cookie cutter sign. Moreover, the skin colored lesion was above a superficial bone, nearly immobile and hard in consistency; which even was mimicking underlying bony lesion on first impression. We have also reviewed the recently published DFSP case presentations and discussed with context to the mimicking nature of the disease.

Keywords: Dermatofibrosarcoma Protuberans, Clinical Mimicry, Non Specific Presentation

1. Introduction

Dermatofibrosarcoma protuberance (DFSP) is a superficial, cutaneous, locally invasive tumor characterized by high rate of recurrence. It affects 4.2 cases / million per year and has been shown increase in incidence in recent years [1]. It is pain less slow growing neoplasm which usually affects torso. As it is a rare neoplasm and slow growing, this skin carcinoma is rarely suspected and thus clinicians can easily mistake it with other common benign skin lesions [2]. In this paper we present a case which was also mistaken to be a benign skin as well as bony lesion. It is it's this ability of mimicry that leads us to a point to ponder: whether clinical mimicry is the rule for DFSP? In the present report we also briefly review the literature related to this.

2. Case

A 35 year old gentleman presented to outpatient department (OPD) with a complaint of small swelling over the left side of face. The swelling was present for last 2 years. It was a painless and very slow growing nodular lesion, located over the maxillary sinus surface marking area. He had one primary care consultation few months ago where computerized tomography (CT) scan was done suggesting cellulitis. As the disease was not resolving, the patient attended our instate OPD. On inspection, an ill defined, solitary, nodular, skin colored, non scaly lesion of approximately 7 x 3 cm over the left maxillary area of the face was seen (figure 1). The lesion was non tender, woody hard on palpation and skin over the lesion was found to be non pinchable. However, the lesion was almost immobile over the underlying tissue (bone).

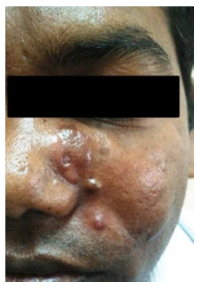

Figure 1. *Gross clinical appearance of the lesion.*

A clinical differential diagnosis of Scleromyxedema, Mucinosis and Dermatofibroma was thought of, while an underlying maxillary bony tumor was also not possible to rule out. When punch biopsy was taken from the lesion, it showed cookie cutter sign / square punch biopsy, thus mimicking Morphea. Ear, Nose and Throat referral and x-ray were done to rule out bony lesion. The histopathology showed horizontally oriented non epithelial neoplasm with fibrocytic differentiation extending from upper reticular dermis to subcutis. The monomorphous fibrocytes are at places spindle shaped and are arranged in horizontal orientation in short interweaving fascicles giving a storiform appearance with hyperplastic epidermis suggestive of DFSP (figure 2). He was then referred for surgical treatment of the lesion.

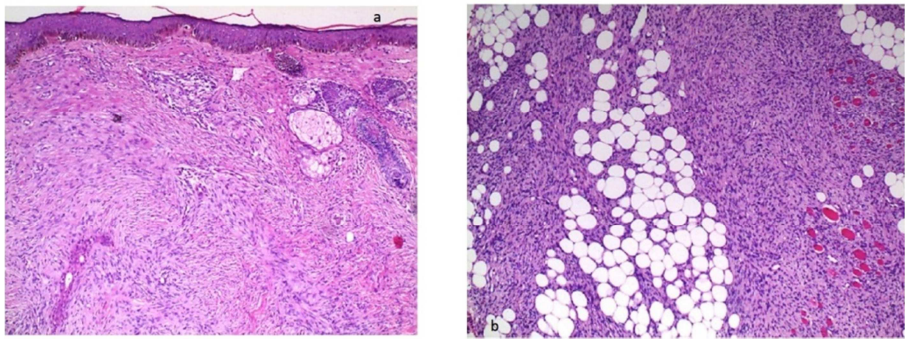

Figure 2. *Histopathologic image (H and E, x40).*

3. Discussion

Dermatofibrosarcoma protuberans is a very rare cutaneous neoplasm. Although it was possibly known before, it was first described in 1924 [3]. It is a clinical dictum and teaching that rare diagnosis is rarely correct. As DFSP is very rare, clinician usually tend to overlook it or rarely consider it as a differential diagnosis and considers the other commoner diseases of same of nearly same clinical presentation. DFSP usually occurs on the trunk, often the chest and shoulders; however it can also affect the limbs, head and neck [2, 4]. The gross presentation is usually a painless, thick or discoloured patch of skin, an indented area or a lump increasing in size or even an irregular nodules or plaques whose color can range from skin coloured to pink / brown and occasionally can have a blue appearance [2]. As the presentation is quite non specific and is a very rare disease, the diagnosis is often delayed and may be mistaken for common harmless skin conditions [2]. The current case was present over face, an unusual site, skin was not discolored, nearly immobile lesion with hard feel just above maxillary bone and deceiving feature of cookie cutter sign which suggested morphea lead to a clinical diagnostic dilemma and was mistaken for commonly encountered benign skin problems.

Published case reports also support this notion and DFSP has been reported to mimic many clinical entities. DFSP of breast has been reported to mimic benign breast lesion [5]. A report of 20 centimeter large DFSP mimicking a breast malignant tumor and abscess have also been reported [6]. Perianal DFSP has been reported to mimic mucinous adenocarcinoma [7]. It has also been reported to mimic with cutaneous sarcoidosis and even meningioma, keloids, scar tissue, etc. [8, 9, 10, 11].

These case reports along with the present case suggest that DFSP is a very good mimicker of common clinical entities like keloids, scar tissue etc. All these features and presentations clearly take us to a question: whether clinical mimicry is the rule for DFSP? Although it is not wise to generalize it but it can also not be denied that DFSP is a great mimicker and clinicians should consider it in differential diagnosis for non-healing and slow growing skin lesion with similar presentations.

As the lesion is often without symptoms other than a slow growing skin lesion, which may even be of skin color; the presentation and diagnosis is usually delayed. High degree of clinical suspicion in a skin lesion which is nonresponsive to treatment over weeks or lesion or scars whose color changes is probably essential in early diagnosis. Such skin lesions can be subjected to biopsy to confirm the diagnosis. The diagnosis of DFSP depends on histopathology and immunohistochemistry. Histopathology of the lesion shows dermal bland spindle cell lesion in storiform islands pattern (spokes of the wheel pattern) containing adipose tissue within the tumor described as honeycomb / Swiss cheese pattern [12]. Immunohistochemistry shows diffuse expression of CD34 [12]. Early suspicion and biopsy will help in diagnosing localized diseases and can be treated with wide surgical resection or Mohs micrographic surgery which is the preferred treatment.

4. Conclusion

The present case and discussion highlights that DFSP is a great mimicker. Mimicking clinical presentation is probably the rule for it. As it very rare disease and non specific in presentation, early suspicion and biopsy in a non-healing similar presenting lesion should be considered.

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