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Dermatofibrosarcoma protuberance: A case series of 15 cases in a tertiary care centre for a period of 2 years

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Abstract

Background: Dermatofibrosarcoma protuberans (DFSP) is a rare superficial fibroblastic cutaneous tumour of low malignant potential characterized by storiform architecture on histopathological appearance with local recurrence rate which varies from 20-50% mostly due to lack of wide tumour free margins excision. Usually DFSP never metastasizes but in 10-15% patient with Fibro sarcomatous DFSP shows distant metastasis, most often in Lungs. Typically, DFSP shows intense positive staining for CD 34 and the Cornerstone of treatment for DFSP is wide surgical excision.

Aim and objective: To study the different clinicopathological presentation of Dermatofibrosarcoma protuberance.

Materials and Methods: This study was conducted in the biopsies with features of DFSP which was later confirmed by Immunohistochemical (IHC) staining by CD 34. The cases studied was sent to the Department of Pathology, Assam Medical College during 2019 and 2020 from the Department of Dermatology and Department of Surgery.

Results: The mean age of diagnosis was 39 years (range, 19-60 years). Among this group of patients, most commonly affected part were the Upper extremities than the Lower extremities with 03 cases (20%) experienced local recurrence.

Conclusions: Our study showed that the patients, who have tumours arranged in a storiform pattern and confirmed by CD34 immunohistochemical staining provides the definitive diagnosis of Dermatofibrosarcoma protuberance. The exact histopathological categorization is also important to select the appropriate treatment and predict the clinical outcome.

Keywords: Dermatofibrosarcoma protuberance, Recurrence, CD-Cluster of Differentiation.

Introduction

Dermatofibrosarcoma protuberans (DFSP) is a superficial, locally aggressive fibroblastic neoplasm, having a cellular storiform appearance and carrying a COL1A1-PDGFB or related fusion with 0.8 – 4.5 new cases per million persons annually which accounts for about 4% of all sarcomas.[1] The World Health Organization classifies it as a low-grade sarcoma.[2] However, it is the most common soft tissue sarcoma which is infiltrative with a high rate of local recurrence, ranging from 24 to 60%.[3] DFSP clinically is characterized by a plaque-like lesion with a sclerodermiform, telangiectatic surface and color ranging from skin-color to red-tinged which generally confined to the dermis and can freely grow into relatively deeper tissue but only invades deep tissue at late stage of the disease and after years of growth, DFSP can eventually form large bulging nodules. However, tumor margins in DFSP are often difficult to define because their irregular antennae can invade surrounding tissues and even deep into the fascia and muscle, which makes it difficult to completely remove the tumor and leads to high recurrence rates after surgery.[4] Histopathologically, it is composed of uniform spindle cell fascicles growing in a storiform pattern with multiple variants with strong and diffuse CD34 immunoreactivity. However, its spindle cell morphology and CD34 immunostaining pattern are overlapped which must be differentiated from other benign and malignant lesions.[5] Further, the early clinical symptoms of DFSP are non-specific which makes the diagnosis difficult, leading to a high incidence of misdiagnosis. Thus, Pathological and immunohistochemical examinations are currently the gold standard for diagnosing DFSP while surgical resection remains the main treatment option.[6]

Aims And Objectives

To study the different clinicopathological presentation of Dermatofibrosarcoma protuberance in a Tertiary Care Centre.

Materials And Methods

A Descriptive study was performed on 15 cases showing DFSP features diagnosed from excisional biopsies obtained from the Department of Pathology, Assam Medical College for a period of 2 years, during 2019 and 2020. The histopathological diagnosis of DFSP was reviewed and later confirmed by CD 34. The patients clinicopathological characteristics were assessed and the patient's follow-up information was obtained from hospital database, medical records, and telephone interviews. The median follow-up duration after initial diagnosis was 1.5 years. The detailed data including age of disease onset, size of the tumour, history of trauma, reoccurence, and related symptoms were collected and analyzed. The approval to the study protocol was given by the institutional ethics committee of the study institute.

Results

Patient characteristics: In total, 15 cases consisting of 6 male and 9 female patients, with an age range of 19-60 years old, and a mean age of 39 years, respectively. All patients presented with primary disease without evidence of metastasis but local reoccurrence was present in 3 cases. Among all patients, the tumors mostly affected were the Trunk(n=08), followed by head and neck (n=04) extremities (n=03). The cases were most commonly upper extremities than the lower extremities. The tumor sizes ranged from 0.5-10 cm. Two patient experienced mild pain when the tumour grew large.

Surgical treatments and complications: A total of 05 patients underwent wide large excision followed by local

excision in 07 cases, 01 patients underwent a wide excision followed by radiotherapy and about 02 patients it was unknown.

Pathological finding: The most common first pathological diagnosis was Classical DFSP showing neoplastic cells which are spindle shaped with plump to elongated nuclei arranged in storiform to whorled pattern. It was often a nonencapsulated subcutaneous tumour but were microscopically infiltrative. Focal dermal, and subcutaneous tissue along with fats invasion was observed in some cases. There were myxoid changes seen in three cases and 1 case with fibro sarcomatous changes.

Follow up: Among the 15 patients with DFSP, follow-up were lost in 3 patients. The median follow-up duration after initial diagnosis was 1.5 years. Among all the 12 patients completed follow-up, 03 patients had local recurrence in 1-1.5 years and 1 of the 3 patients did not undergo WLE after local resection had recurrence and 3 patients succumbed to death.

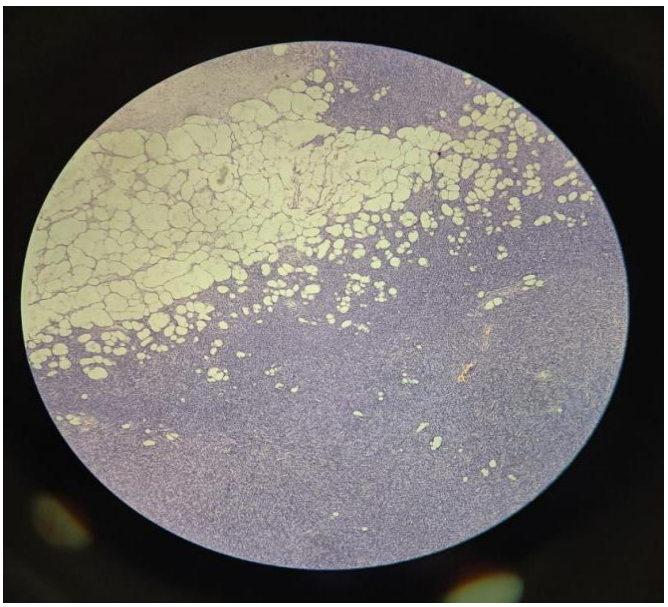

Figure 1: H n E (10x) Proliferation of neoplastic cells arranged in storiform pattern showing individual spindle shaped with ovoid to elongated nuclei and moderate amount of cytoplasm infiltrating around adipocytes.

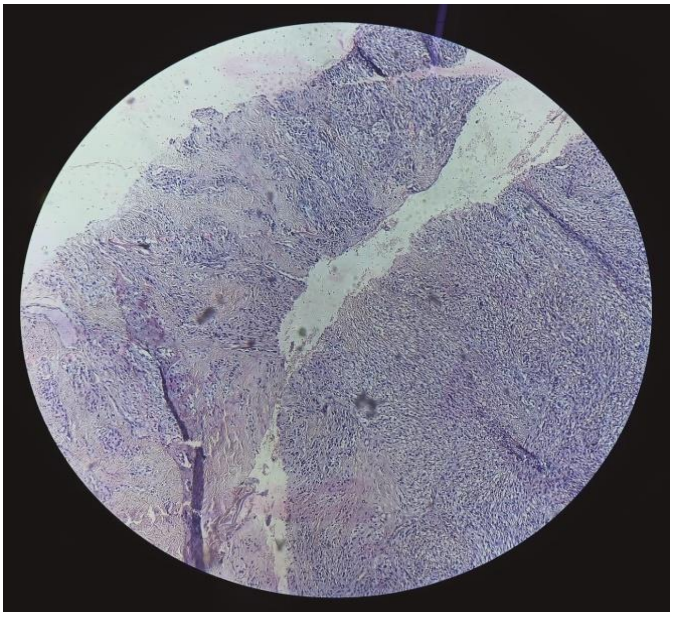

Figure 2: H n E (10 x) Proliferation of neoplastic cells arranged in storiform pattern alongwith areas of myxoid changes.

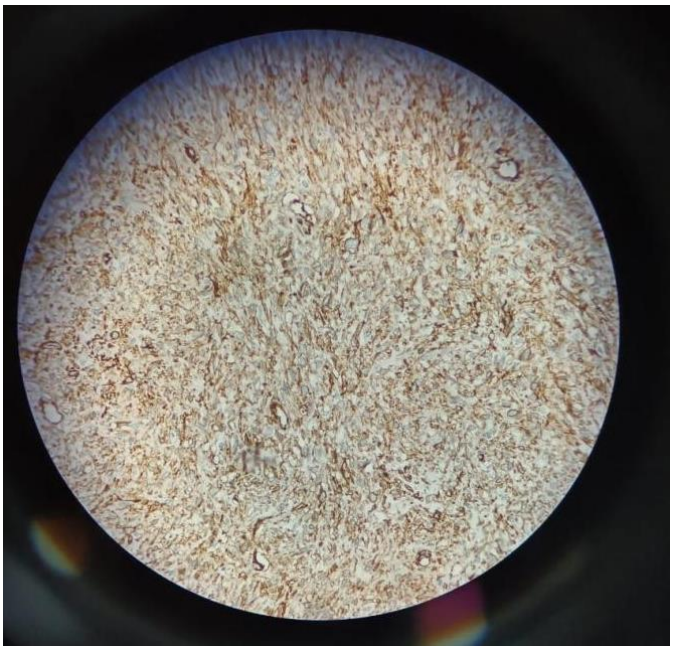

Figure 3: Immunohistochemistry (IHC) (40X) of DFSP Showing CD 34 Positivity.

Table 1: Clinical characteristics of patients diagnosed with Dermatofibrosarcoma protuberans.

Patients Characteristics	Number (%)
Gender	
Male	06 (40%)
Female	09 (60%)
Tumor Size (Cm)	
<2	01 (6.67%)
2-5	06 (40.00 %)
>5	08 (53.33%)
AGE(Y)	
<20	01 (6.67%)
20-50	10 (66.67%)
>50	04 (26.67%)
Location	
Head And Neck	04 (26.67%)
Trunk	07 (53.33%)
Extremities	03 (20.00%)
Tumour Presentation	
Firm	07 (46.67%)
Mobile	00 (00.00%)
Well-Circumscribed	03 (20.00%)
Irregular	04 (26.67%)
Unknown	01 (6.67%)
Symptoms	
Painful	02 (13.33%)
Not Painful	13 (86.66%)
Subtype	
Classical	12 (80.00%)
Variant	03 (20.00%)
Premier Or Recurrence	
Premier	05 (33.33%)
Recurrence	04 (26.67%)
Unknown	06 (40.00%)
Treatment	
Local Excision	07 (46.67%)
Wide Large Excision	05 (33.33%)
Wide Excision with Radiotherapy	01 (06.67%)
Unknown	02 (13.33%)
Post-Operative Radiotherapy	
Yes	01 (06.67%)
No	08 (53.33%)
Unknown	06 (40.00%)

Table 2: Histologic Characteristics of patients diagnosed with Dermatofibrosarcoma Protuberans.

Variable	Number (%)
Infiltration	
Dermis	08 (53.33%)
Subcutis	04 (26.67%)
Adipose Tissue	03 (20.00%)
Muscular Layer	00 (00.00%)
Histology	
Spindle Cells	11 (73.33%)
Mxoid Changes	03 (20.00%)
Fibro sarcomatous	01 (06.67%)

Discussion

According to current WHO 5th Edition, DFSP is defined as a superficial, locally aggressive fibroblastic neoplasm, having cellular storiform appearance and carrying a COL1A1-PDGFB or related fusion. The present study collected data on DFSP was uniformly diagnosed based on the expression of CD34 by immunohistochemistry. It is mostly common in adults between 20 and 50 years of age[7], but recently there is a trend of increasing incidences in children. According to reported cases, the proportion of pediatric DFSP cases ranges between 6 and 20%.[8] In our study, 10 patients (66.67%) were in their 20–50s, 04 (26.67%) patients were in more than 50 years and 01 case (6%) under 20s with patients age ranging from 19-60 years.

In the present study, local recurrence was seen in 04 cases (26.67%), whereas few studies shows DFSP has a tendency for local recurrence of up to 25% and most

recurrences tend to develop within 3 years, after ressection and for late recurrences, even after 15 years, have been also reported. [9,10,11.]

The most affected anatomical locations in our study were the trunk (n=07), followed by the head and neck (n=04 and the lower extremities (n=03). The lower extremities were more commonly affected than the upper extremities which is similar to Mariaarchontaki et al. study.[12] However, Larbcharoensub et al. in their study found the most affected part were body.[13]

On other hand, sarcomatous change in DFSP represents a form of tumor progression, which occurs in 10-15% of DFSP and is associated with a prognosis worse than ordinary DFSP. [14–19] In the present study, sarcomatous transformation occurred in 1 cases (06.67%) of DFSP cases and Larbcharoensubet al. in their study also found sarcomatous change but in 06 cases.[13] Further, in current series it was seen, 3 patients were succumbed to death within the follow-up time of 1.5 years.

The main limitation of the present study was small sample and compared with other studies, the follow-up time was also short and, with time, there may be different results. Therefore, a large prospective randomized study should be done along with multi-centre study to demonstrate the possible differences.

Conclusion

In conclusion, the present study found that the factors that predict the recurrence of DFSP were mostly common in the patients with trauma history. The tumours with spindle cell arranged in a storiform pattern and CD34 immunohistochemical staining provides the pathological diagnosis of DFSP. However, a combination of clinicopathological features, immunohistochemistry and, in specific cases, molecular or cytogenetic testing, is essential for definitive diagnosis. On the otherhand, the exact histopathological categorization is important for selecting the appropriate treatment and for predicting the clinical outcome.

References

1. Molina, A. S., Duprat Neto, J. P., Bertolli, E., da Cunha, I. W., Fregnani, J. H., Figueiredo, P. H.,. & de Abranches Oliveira Santos Filho, I. D. (2018). Relapse in dermatofibrosarcoma protuberans: A histological and molecular analysis. Journal of surgical oncology, 117(5), 845-850.
2. Shah, K. K., McHugh, J. B., Folpe, A. L., & Patel, R. M. (2018). Dermatofibrosarcoma protuberans of distal extremities and acral Sites. The American Journal of Surgical Pathology, 42(3), 413-419.
3. Malhotra, B., & Schuetze, S. M. (2012). Dermatofibrosarcoma protruberans treatment with platelet-derived growth factor receptor inhibitor: a review of clinical trial results. Current opinion in oncology, 24(4), 419-424.
4. Zhou, X., Sun, D., Liu, Y., Sun, Q., Yuan, Z., Luo, X.,. & Chen, J. (2020). Dermatofibrosarcoma protuberans: our 10-year experience on 80 patients. Journal of Dermatological Treatment, 31(6), 554-558.
5. Hao, X., Billings, S. D., Wu, F., Stultz, T. W., Procop, G. W., Mirkin, G., & Vidimos, A. T. (2020). Dermatofibrosarcoma protuberans: update on the diagnosis and treatment. Journal of clinical medicine, 9(6), 1752.
6. Fletcher, C. D. (2014). The evolving classification of soft tissue tumours–an update based on the new 2013 WHO classification. Histopathology, 64(1), 2 11.

7. Li, Y., Wang, C., Xiang, B., Chen, S., Li, L., & Ji, Y. (2017). Clinical features, pathological findings and treatment of recurrent dermatofibrosarcoma protuberans. Journal of Cancer, 8(7), 1319.
8. Tsai, Y. J., Lin, P. Y., Chew, K. Y., & Chiang, Y. C. (2014). Dermatofibrosarcoma protuberans in children and adolescents: clinical presentation, histology, treatment, and review of the literature. Journal of Plastic, Reconstructive & Aesthetic Surgery, 67(9), 1222-1229.
9. Stojadinovic, A., Karpoff, H. M., Antonescu, C. R., Shah, J. P., Singh, B., Spiro, R. H.,. & Shaha, A. R. (2000). Dermatofibrosarcoma protuberans of the head and neck. Annals of Surgical Oncology, 7, 696-704.
10. Ballo, M. T., Zagars, G. K., Pisters, P., & Alan Pollack, M. D. (1998). The role of radiation therapy in the management of dermatofibrosarcoma protuberans. International Journal of Radiation Oncology* Biology* Physics, 40(4), 823-827.
11. Haas, A. F., & Sykes, J. M. (1998). Multispecialty approach to complex dermatofibrosarcoma protuberans of the forehead. Archives of Otolaryngology–Head & Neck Surgery, 124(3), 324-327.
12. Archontaki, M., Korkolis, D. P., Arnogiannaki, N., Konstantinidou, C., Georgopoulos, S., Dendrinos, P. & Kokkalis, G. (2010). Dermatofibrosarcoma protuberans: a case series of 16 patients treated in a single institution with literature review. Anticancer research, 30(9), 3775-3779
13. Larbcharoensub, N., Kayankarnnavee, J., Sanpaphant, S., Kiranantawat, K., Wirojtananugoon, C., & Sirikulchayanonta, V. (2016). Clinicopathological features of dermatofibrosarcoma protuberans. Oncology Letters, 11(1), 661-667
14. Mentzel, T., Peeutour, F., Lazar, A., & Coindre, J. M. (2013). Dermatofibrosarcoma protuberans. In: World Health Organization (WHO) Classification of Tumours of Soft tissue and Bone. Pathology and Genetics. Fletcher, C. D. M., Bridge, J. A., Hogendoorn, P., & Martens, F., (eds). Vol 5. 4th edition. IARC Press, Lyon. pp. 77-79.
15. Connelly, J. H., & Evans, H. L. (1992). Dermatofibrosarcoma protuberans: a clinicopathologic review with emphasis on fibrosarcomatous areas. The American journal of surgical pathology, 16(10), 921-922.
16. Sato, N., Kimura, K., & Tomita, Y. (1995). Recurrent dermatofibrosarcoma protuberans with myxoid and fibrosarcomatous changes paralleled by loss of CD34 expression. The Journal of dermatology, 22(9), 665-672.
17. Mentzel, T., Beham, A., Katenkamp, D., Dei Tos, A. P., & Fletcher, C. D. (1998). Fibrosarcomatous (" high-grade") dermatofibrosarcoma protuberans: clinicopathologic and immunohistochemical study of a series of 41 cases with emphasis on prognostic significance. The American journal of surgical pathology, 22(5), 576-587
18. Goldblum, J. R., Reith, J. D., & Weiss, S. W. (2000). Sarcomas arising in dermatofibrosarcoma protuberans: a reappraisal of biologic behavior in eighteen cases treated by wide local excision with extended clinical follow up. The American journal of surgical pathology, 24(8), 1125-1130.

19. Abbott, J. J., Oliveira, A. M., & Nascimento, A. G. (2006). The prognostic significance of fibrosarcomatous transformation in dermatofibrosarcoma protuberans. The American journal of surgical pathology, 30(4), 436-443.