

CASE REPORT

CASE REPORT OF DERMATOFIBROSARCOMA PROTUBERANS

By

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Dermatofibrosarcoma protuberans is a distinct clinical entity. Though the condition was first recognised by Darier and Ferrand in 1929 who described it under the heading of "progressive and recurring dermatofibromata or fibrosarcoma of the skin", there were isolated earlier reports of the condition also in the literature. It was Hoffmann in 1925, who while describing several cases of this disease designated it as "dermatofibroasarcoma protuberans". Since then, there have been many articles, reports, and reviews; and a considerable volume of literature has accumulated on the subject.

Prompted by a recent case-report from Bhopal, (India) by Shrivastava, Tandon and Hafeez, we put on record another case which, as a matter of fact, was diagnosed earlier by us in 1960 as dermatofibrosarcoma protuberans and demonstrated in a staff clinical meeting.

CASE-REPORT

R, a 55 yeares old, Hindu, house-wife came to the outdoor in 1955 for recurrent nodular swellings of 15 years duration on left thigh just below the lateral part of left inguinal ligament. It was of the size of an almond when first noticed, but it gradually enlarged to that of an orange in nearly six months' period. Later it started bleeding, and hence it was got excised. Because of its recurrence at that very site, this excision had to be repeated for three more times before she came under our obseavation in July, 1955.

The presenting lesion, at that time, revealed a superficial scar nearly 3"x2", in size, portion of which was depigmented and a portion was red, elevated and slightly ulcerated. There were also four nodular swellings of sizes varying from pea to an egg, on it. These nodular swellings were painless, firm, some-what lobulated, movable over the under-lying structures and covered over with reddish skin which was intact but firmly adherent to them. Excisional biopsy of one of the nodules was done, which after histopathological examination was reported as "fibroma". After this biopsy report, remaining nodules were also enulceated and these too were subjected to histopathological examination. They, too, were reported as "fibromra". Patient re-visited the department on 27-6-60 with fresh similar nodules in the operated scar. She refused excision at that tima but returned on 27-1-1961 when one of the nodules had attained the size of 3"x1½". Portion of it was fungating and was covered with gangrenous slough; it was also emitting foul smell. Patient was admitted again, and biopsy repeated. There was no evidencee of metastasis anywhere in the body. Later, entire lesion together with

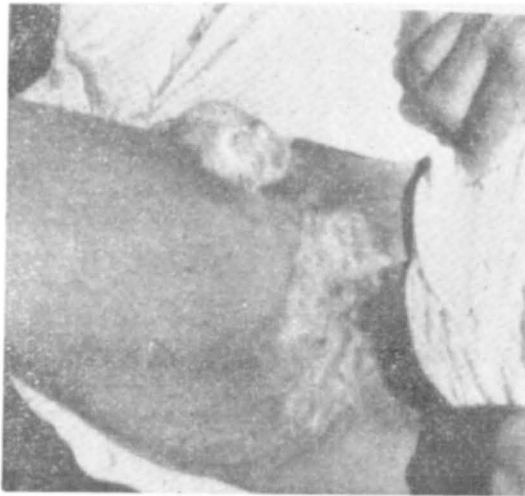
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1 cm of surrounding healthy skin was re-excised. The growth, at the time of excision, was found to be superficial to deep fascia.

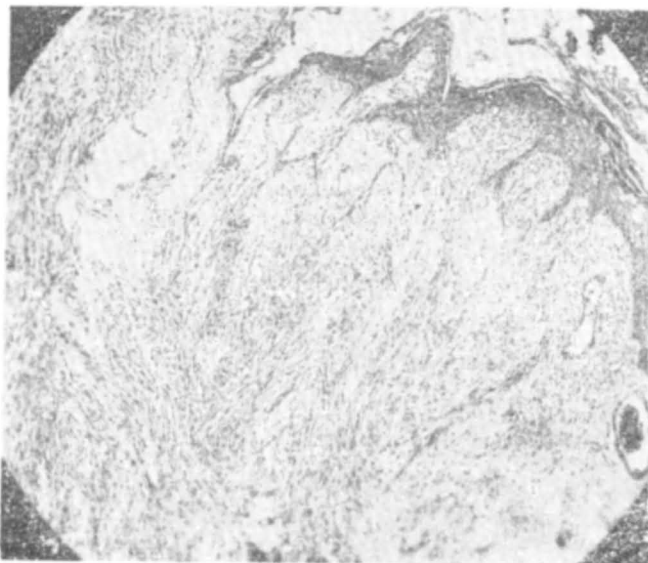
Follow-Up. Patient was sent for on 5-9-1964. Her re-examination at that time revealed fresh 30 nodules in the scar varying from the size of a pea to that of a small almond in that very region. Even now, there was no evidence of metastasis anywhere in the body.

HISTOPATHOLOGICAL EXAMINATION

Fresh sections from blocks of biopsy specimens taken at various times were cut and re-examined. The histopathological appearances were found to be almost identical in all the sections.

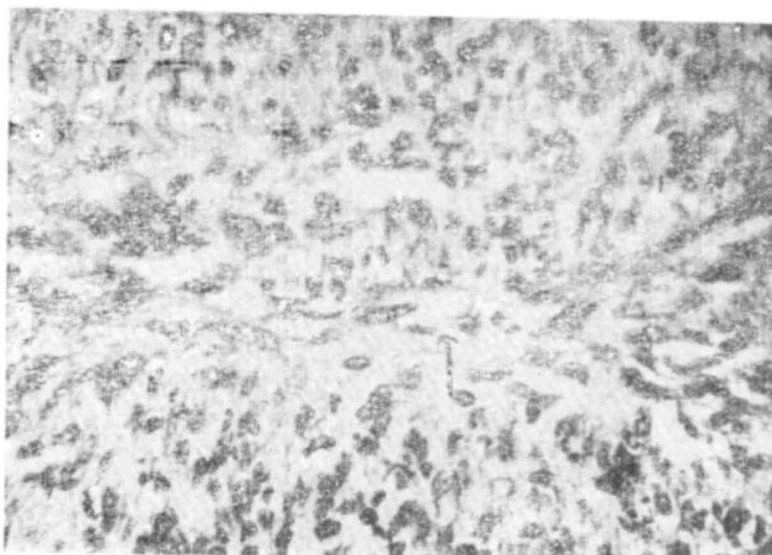


PHOTOGRAPH No. 1.



PHOTOGRAPH No. 2.

Microscopic examination of the sections showed, more or less, a normal epidermis. Underneath this, in the dermis were seen closely set bundles of



PHOTOGRAPH No. 3.

fibroblasts running criss-cross in different directions. There was also seen a tendency to cartwheel arrangement at places. The tumour, in the sections, looked moderately cellular and its nuclei were moderately large and plumpy. Neither there was an appreciable degree of mitosis nor there were any tumour-giant-cells. The tumour was partly myxomatous in the superficial part of the dermis in one of the sections. There was no embolisation of the blood vessels in any of the sections.

COMMENTS

The cases, suffering from this disease, are more likely to seek advice in surgical out-patient department of an institution, but they may occasionally present themselves in a dermatologic clinic also.

The clinical characters of the case and its tendency to recur are quite distinctive. If these features are kept in mind, there is very little likelihood of our missing its diagnosis. As a matter of fact, these characteristic features led Binkley (1939) to believe that the diagnosis of the condition could be made on the basis of clinical evidence alone. He is supported, in this respect, by Degaraciansky et al (1950) and Gerlach (1957).

Considerable controversy existed in regard to histopathology of the lesion. Costa (1946) believed that the histopathology of the disease is not specific but varied from case to case according to the stage of the disease process. Similar views were held by Degracinsk et al (1950). Michelson (1927), Luscombe and Pratt (1954) and Schiff et al (1960). In contrast, Taylor and Helwig (1952) who analysed histopathological aspects of 115 examples of dermatofibrosarcoma protu-

berans from the files of Armed Forces Institute of Pathology came to the conclusion that the presence of the arrangement of tumour cells in a cart-wheel or whirli-gig pattern conferred histological specificity to the lesion and considered it as a characteristic histopathologic feature. Levin Ne et al (1963), while giving the differentiating histological features of the disease from Pseudo-sarcomatous dermatofibrosarcoma, also emphasised cart-wheel arrangement of the predominating cells as an important feature, alongwith uniformity of the cell type and relative absence of mitotic figures. This cart-wheel pattern was present in all the sections of this case.

SUMMARY

1. Another case of dermatofibrosarcoma protuberans is presented.
2. The clinical findings, in general agree with that of the previous report. The characteristic clinical features and its tendency to recur are emphasised.
3. The histopathological appearances are discussed.
4. Our case neither showed any metastasis nor embolisation of the blood vessels in any section of the lesion even after lapse of 24 years after first appearance of the disease.

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