REVIEW

A systematic review of outcome data for dermatofibrosarcoma protuberans with and without fibrosarcomatous change

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Background: To our knowledge, no systematic review of dermatofibrosarcoma protuberans (DFSP) outcomes based on the presence or absence of fibrosarcomatous (FS) change has been performed.

Objective: We sought to compare available outcome data for DFSP versus DFSP-FS.

Methods: The literature was searched for DFSP and DFSP-FS reports with outcome data (local recurrence, metastasis, or death from disease). Chi-square tests were calculated to determine whether DFSP and DFSP-FS significantly differed in risk of local recurrence, metastasis, and death from disease.

Results: In all, 24 reports containing 1422 patients with DFSP and 225 with DFSP-FS are summarized. Risk of local recurrence, metastasis, and death from disease in DFSP-FS was significantly higher as compared with DFSP (local recurrence 29.8% vs 13.7%, risk ratio 2.2 [95% confidence interval 1.7-2.9]; metastasis 14.4% vs 1.1%, risk ratio 5.5 [95% confidence interval 4.3-7.0]; and death from disease 14.7% vs 0.8%, risk ratio 6.2 [95% confidence interval 5.0-7.8]). There was no significant difference in DFSP-FS outcomes based on proportion of FS change within tumors.

Limitations: This study is based on previously reported data from different hospitals with no uniform process for reporting FS change. The impact of confounders (age, immune status, tumor location, treatment) could not be evaluated because of limited data.

Conclusion: Based on available retrospective data, risk of metastasis and death is elevated in DFSP-FS as compared with DFSP. Even a low degree of FS involvement portends worse outcomes. (J Am Acad Dermatol http://dx.doi.org/10.1016/j.jaad.2014.03.018.)

Key words: dermatofibrosarcoma protuberans; dermatofibrosarcoma with fibrosarcomatous change; fibrosarcomatous dermatofibrosarcoma protuberans.

Dermatofibrosarcoma protuberans (DFSP) is a rare low-grade malignancy. It is often treated with surgical resection, however a proportion of patients (1%-21%) experience local recurrence.[1,2] Metastasis and death from the disease is rare, with reported metastatic rates of 1% to 4%.[1,3,4] The tumor cells in DFSP have a storiform or cartwheel-like pattern, uniform nuclei, minimal cytologic atypia, absent to low mitotic activity, and are CD-34^{+}. Conversely, tumor cells of fibrosarcoma are monotonously fusiform with enlarged atypical

Abbreviations used:

CI:	confidence interval
DFSP:	dermatofibrosarcoma protuberans
FS:	fibrosarcomatous
RR:	risk ratio

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nuclei, cytologic atypia, a high mitotic count, and are often CD34$^-$. In contrast to the storiform pattern in DFSP, fibrosarcoma cells grow a fascicular (herringbone) arrangement (Fig 1). When fibrosarcomatous (FS) changes are observed in association with that of typical DFSP in a cutaneous lesion, DFSP-FS is diagnosed. It has been reported in various case series that 3% to 20% of all cases of DFSP have foci of FS change and are thus DFSP-FS.[5-11] Studies have attempted to identify independent clinicopathologic risk factors for recurrence and metastasis in DFSP.[5-7] These studies found that cases of DFSP with FS transformation have an increased risk of recurrence. However, not all studies have found an increased risk of recurrence in DFSP-FS as compared with DFSP.[12,13] Such ambiguity regarding the prognostic significance of FS transformation makes it difficult to define optimal management of these cases. It is also unclear whether a higher degree of FS transformation portends worse prognosis or whether there is some level of FS change that is "safe." This systematic review was undertaken to compare the rates of local recurrence, metastasis, and disease-specific death in patients with DFSP and DFSP-FS to provide preliminary prognostic information and define areas for future study.

CAPSULE SUMMARY

- Past series comparing prognosis of patients with dermatofibrosarcoma protuberans (DFSP) versus fibrosarcomatous DFSP have yielded contradictory results.
- Based on available data, risk of metastasis and death is elevated in fibrosarcomatous DFSP as compared with DFSP.
- Fibrosarcomatous DFSP warrants aggressive treatment and close observation for recurrence and metastasis.

METHODS

The MEDLINE database was searched for the period January 1, 1992, to August 1, 2013. No language or study design restrictions were used. Search terms were "dermatofibrosarcoma protuberans," "DFSP-FS," "fibrosarcomatous DFSP," and "transformed DFSP." Reference lists of included reports were also examined to obtain any additional reports that met inclusion criteria. Studies were included if they reported outcomes of interest (local recurrence, metastasis, or disease-specific death) and had a median follow-up of at least 4 years so as not to underestimate the rate of recurrence or metastasis. Studies containing both DFSP and DFSP-FS were included. However, studies in which distinction between DFSP and DFSP-FS was not specified by case were excluded. Other studies were excluded if: they were case reports, the DFSP was not cutaneous in origin, or the primary lesion pathology was unknown. Cases were classified based on the pathology of their primary lesion (ie, a recurrence that transformed to DFSP-FS but original lesion was DFSP would be classified as DFSP). The number of DFSP and DFSP-FS cases was tabulated, as were any outcomes of interest arising from each case. Outcomes in DFSP-FS cases were extracted and analyzed separately by degree of FS change, in addition to overall outcomes. The χ^2 tests and corresponding risk ratios (RR) with 95% confidence intervals (CI) were calculated for each outcome to determine whether DFSP and DFSP-FS significantly differed with regard to risk of local recurrence, metastasis (nodal or distant), and death from disease. P less than or equal to .05 level of significance was used. All statistical analysis was performed using STATA v 12.0 (StataCorp LP, College Station, TX).

RESULTS

A total of 1025 references were identified through MEDLINE search and reference lists; 989 were excluded because of duplicates, lack of outcome data, case reports, unknown pathology of the primary lesion, or noncutaneous origin of the DFSP. Twelve studies were excluded for having less than 4-year median follow-up. Eleven studies containing outcome data on cases of both DFSP and DFSP-FS, 4 studies on DFSP-FS, and 9 studies on DFSP were included. The included studies contained 1422 cases of DFSP (Table I) and 225 cases of DFSP-FS (Table II). All studies were retrospective. Five studies did not report the proportion of FS area. In the remaining reports, the FS area ranged from 5% to 100%. Method of treatment was often not individually reported for each case, so the impact of treatment on outcomes could not be analyzed.

The risk of local recurrence, metastasis, and death from disease in DFSP-FS was significantly higher as compared with DFSP (local recurrence 67/225 [29.8%] vs 195/1422 [13.7%], RR 2.2 [95% CI 1.7-2.9]; metastasis 32/225 [14.4%] vs 16/1422 [1.1%], RR 5.5 [95% CI 4.3-7.0]; and death from disease 33/225 [14.7%] vs 11/1422 [0.8%], RR 6.2 [95% CI 5.0-7.8], respectively).

To examine whether a large degree of FS change is required to adversely impact outcomes, DFSP-FS cases with varying degrees of FS differentiation were

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