

## Atypical Fibrous Histiocytoma of the Skin with Necrobiotic Granuloma-like Features

Yosuke Ishitsuka<sup>1</sup>, Kunuaki Ohara<sup>2</sup> and Fujio Otsuka<sup>1</sup>

Department of Dermatology, <sup>1</sup>Graduate School of Comprehensive Human Sciences, University of Tsukuba, 1-1-1 Ten-nodai, Tsukuba, Ibaraki 305-8575, and <sup>2</sup>Toranomon Hospital, Toranomon, Japan. E-mail: c0930412@md.tsukuba.ac.jp  
Accepted December 16, 2010.

Fibrous histiocytoma (FH) is a common benign cutaneous neoplasm. It is also known as dermatofibroma, sclerosing haemangioma, or nodular subepidermal fibrosis, depending on the microscopic appearance of the lesion. Many histological variants of FH, such as aneurysmal, cellular, epithelioid, keloidal, and palisading subtypes, have been identified. Atypical fibrous histiocytoma (AFH) is a rare variant of FH that was first described in 1983 by Fukamizu et al. (1). AFH is also known as dermatofibroma with monster cells (2) or atypical benign fibrous histiocytoma (3). This neoplasm is characterized by a relatively benign clinical course despite having pseudosarcomatous histological features (1–3); however, in comparison with FH, recurrence of AFH is common, although metastases are rare (4). The clinicopathological spectrum of AFH is currently undergoing reclassification because of the rarity of this condition and the concern that it may have been previously misdiagnosed as pleomorphic sarcoma (4). Although both FH and AFH have wide variations in their histological profiles (4, 5), no cases of necrobiotic granuloma-like features have been reported. We report here a case of AFH with necrobiotic granuloma-like features.

### CASE REPORT

A 52-year-old Japanese man attended our clinic with a tumour on his left elbow, which had first been noticed 6 months previously. The lesion was accompanied by slight tenderness. The patient had no history of trauma or any other significant medical problem.

On examination, the tumour was found to be 20 mm in diameter with a pale red-coloured elevation with crust formation in the centre. On palpation, it was found to be a firm, mobile, dermal-to-subcutaneous mass (Fig. 1). Although the initial clinical differential diagnosis included an epidermal inclusion cyst or a granulomatous lesion, establishing a conclusive diagnosis was difficult. The lesion was excised for treatment and diagnostic purposes.

Histopathological examination revealed a tumour existing mainly in the dermis, with focal extension into the subcutaneous tissue (Fig. 2a). The epidermis was hyperplastic and the rete ridges were elongated. In the centre of the lesion, a large area composed of degenerated collagen, which was surrounded by fibroblast-like cells, histiocyte-like cells, and inflammatory cells interspersed with hyaline collagen bundles, was observed (Fig. 2b). The fibroblast-like cells showed a partial sto-



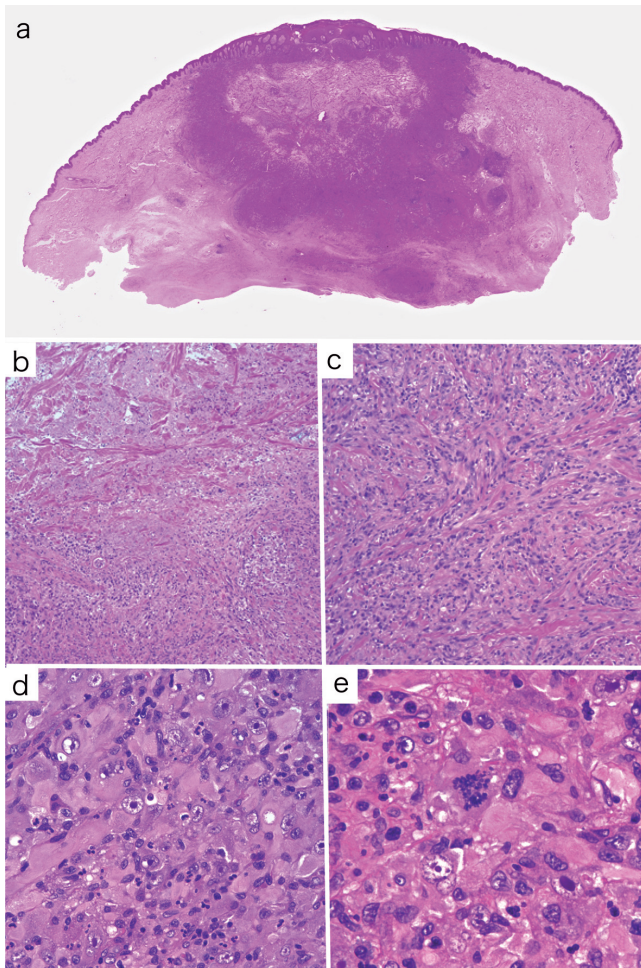
Fig. 1. Tumour on the left elbow. A firm dermal-to-subcutaneous tumour (20 mm in diameter) with a pale red-coloured elevation with crust formation in the centre.

reform pattern, which was more evident at the periphery of the lesion (Fig. 2c). In the deep part of the lesion, a predominance of histiocyte-like cells was observed, among which bizarre pleomorphic cells with foamy cytoplasm, marked nuclear atypia, and multinucleated giant cells were included (Fig. 2d). These cells had large nuclei with prominent nucleoli and comprised mitotic figures (8/HPF), including some atypical forms (Fig. 2e). Immunostaining of the tumour cells revealed diffuse positivity for vimentin and negativity for alpha-smooth muscle actin, CAM5.2, CD68, CD34, and S-100. AFH was diagnosed on the basis of these findings.

Two months after the first operation, an extended resection and skin graft was performed. To date, no local recurrence or metastasis has been observed in the patient since his first visit 4 years earlier.

### DISCUSSION

According to a report on the clinicopathological analysis of 59 patients with AFH (4), all tumours in these patients had features of classical FH, at least focally, although the proportion of pleomorphic cells was reported to be



**Fig. 2.** Histopathological features observed with haematoxylin and eosin staining. (a) Dermal-to-subcutaneous tumour with focal extension into the subcutaneous tissue. A thickened epidermis and the main lesion located in the dermis (1 $\times$ ). (b) A large area composed of degenerated collagen surrounded by fibroblast-like cells, histiocyte-like cells, and inflammatory cells interspersed with hyaline collagen bundles (40 $\times$ ). This finding was suggestive of necrobiotic granuloma. (c) At the periphery of the lesion, fibroblast-like cells showing a storiform pattern are evident (100 $\times$ ). (d) A region showing a predominance of histiocyte-like cells. Bizarre pleomorphic cells with foamy cytoplasm and multinucleated giant cells are present. The bizarre cells contain large nuclei with prominent nucleoli (400 $\times$ ). (e) Normal and atypical mitotic figures are observed (800 $\times$ ).

variable. In the present case, histological features typical of classical FH, such as a thickened epidermis, the main lesion being located in the dermis, and the presence of fibroblast-like cells showing a storiform pattern, were observed. These unique features enabled the tumour to be diagnosed as AFH, rather than pleomorphic sarcoma, despite the presence of bizarre pleomorphic cells in regions where the histiocyte-like cells were predominant, which is a feature of the latter tumour.

In addition to the pseudosarcomatous findings, a salient histopathological feature was the large central area composed of degenerated collagen surrounded by fibroblast-like and histiocyte-like cells. This finding was suggestive of necrobiotic granuloma, and such a finding has led to debate and controversy for decades as to whether FH should be considered a reactive inflammatory process or a neoplasm (6–8). Some authors have studied this question, and have proved the existence of monoclonality in at least some FHs (9, 10). Based on the accepted understanding that FH progresses from a histiocyte-like form to a fibroblast-like form over time, Hui et al. (10) postulated that histiocyte-like cells represent a neoplastic process, and in contrast, fibroblast-like cells represent a reactive process. Although FH shows diverse histological features and has many variants, to our knowledge, FH (or AFH) with such features has not been documented previously.

Although most cases of AFH pursue a benign clinical course, AFH has a higher tendency to local recurrence than common FH (1–3, 5) and it rarely metastasizes (4, 5). We therefore performed an extended resection after the final diagnosis.

## REFERENCES

1. Fukamizu H, Oku T, Matsumoto K, Okayama H, Tagami H. Atypical ("pseudosarcomatous") cutaneous histiocytoma. *J Cutan Pathol* 1983; 10: 327–333.
2. Tamada S, Ackerman AB. Dermatofibroma with monster cells. *Am J Dermatopathol* 1987; 9: 380–387.
3. Beham A, Fletcher CDM. Atypical 'pseudosarcomatous' variant of cutaneous benign fibrous histiocytoma: report of eight cases. *Histopathology* 1990; 17: 167–169.
4. Kaddu S, McMenamin ME, Fletcher CDM. Atypical fibrous histiocytoma of the skin. Clinicopathologic analysis of 59 cases with evidence of infrequent metastasis. *Am J Surg Pathol* 2002; 26: 35–46.
5. Luzar B, Calonje E. Cutaneous fibrohistiocytic tumours – an up date. *Histopathology* 2010; 56: 148–165.
6. Vanni R, Fletcher CD, Sciort R, Dal Cin P, De Wever I, Mandahl N, et al. Cytogenetic evidence of clonality in cutaneous benign fibrous histiocytomas: a report of the CHAMP study group. *Histopathology* 2000; 37: 212–217.
7. Calonje E. Is cutaneous benign fibrous histiocytoma (dermatofibroma) a reactive inflammatory process or a neoplasm? *Histopathology* 2000; 37: 278–280.
8. Zelger BG, Zelger B. Dermatofibroma (fibrous histiocytoma): an inflammatory or neoplastic disorder? *Histopathology* 2001; 38: 379–381.
9. Chen T, Kuo T, Chan H. Dermatofibroma is a clonal proliferative disease. *J Cutan Pathol* 2000; 27: 36–39.
10. Hui P, Glusac EJ, Sinard JH, Perkins AS. Clonal analysis of cutaneous fibrous histiocytoma (dermatofibroma). *J Cutan Pathol* 2002; 29: 385–389.