Spontaneous Regression and Recurrence in a Case of Nodular Fasciitis

Eva N. Hüter[1], Chyi-Chia R. Lee[2], Richard M. Sherry[3] and Mark C. Udey[4]*
*[1]Laboratory of Immunology, National Institute of Allergy and Infectious Diseases, [2]Laboratory of Pathology, [3]Surgery Branch and [4]Dermatology Branch, National Cancer Institute, National Institutes of Health, Bethesda, MD 20892, USA. *E-mail: udey@helix.nih.gov*
Accepted February 26, 2009.

Sir,

Nodular fasciitis is a sporadic tumor that results from a benign proliferation of myofibroblasts in soft tissues. It presents clinically as a rapidly-growing firm nodule, most often localized in the subcutaneous tissue (1). While it may arise in any part of the body, there is a predilection for the upper extremities, especially the forearms (1–3). Nodular fasciitis occurs most commonly in younger adults, affecting males and females with equal frequency (2–4). Histologic diagnosis of these rapidly growing lesions can be difficult owing to their dense cellularity and high mitotic activity; features shared with some sarcomas (2, 5, 6). Standard care consists of complete excision, and local recurrences occur very rarely (1–3). Spontaneous regression of nodular fasciitis has been reported, but there are only a few cases documented in the literature, and most often regression followed biopsy or incomplete excision of the lesion (7, 8). We describe here an unusual case of nodular fasciitis that regressed spontaneously and recurred after a 5-year interval.

CASE REPORT

A 30-year-old female physician presented for evaluation of a solitary nodule on her left forearm. The lesion had grown to a maximal diameter of 1.5 cm over several days. There was no history of trauma. On clinical examination the nodule was firm, non-tender, well circumscribed, but seemed adherent to muscle. The lesion was not fixed to the overlying skin. The patient was otherwise healthy and there was no family history of dermal or connective tissue tumors. Interestingly, the patient described occurrence of a similar lesion at the exact same location on her left forearm 5 years earlier. This lesion had grown to a maximal diameter of ~2 cm over a several week period and had exhibited similar clinical features. The earlier tumor had regressed spontaneously over a several-week period, leaving an asymptomatic residual firm nodule ~2 mm in diameter. A clinical diagnosis of nodular fasciitis was made. The residual lesion did not change in size or character over a subsequent 5-year period.

The rapid regrowth of this lesion was cause for concern and complete excision was recommended. The tumor was subsequently excised completely, together with surrounding subcutaneous tissue and an overlying ellipse of skin. Histologic evaluation of the surgical specimen revealed a small, circumscribed, but unencapsulated, nodular lesion comprised predominantly of spindle-shaped myofibroblasts located in the subcutaneous fat (Fig. 1A). The lesion was confined to the subcutis and did not involve the overlying dermis or epidermis. Some myofibroblasts showed reactive changes, including plump nuclei and conspicuous nucleoli, but increased mitotic activity was not observed (Figs 1B and C). Immunohistochemical staining revealed that the tumor cells were positive for smooth muscle actin (SMA) (Fig. 1D), but negative for S-100, desmin and CD34 (not shown). The histologic diagnosis of a benign myofibroblastic proliferation confined to the subcutis was consistent with our clinical diagnosis of nodular fasciitis. The postoperative course was uneventful and local recurrence was not observed during a 6-month follow-up period.

DISCUSSION

The clinical course of our patient reveals two interesting features of nodular fasciitis: the spontaneous near-complete regression of the tumor without any antecedent manipulation, and subsequent local recurrence after a 5-year quiescent period. Regression of nodular fasciitis has been reported after fine needle aspiration biopsy, incisional biopsy or incomplete excision (7, 8). In these instances, it is difficult to determine if involution was "spontaneous" or if it was triggered by surgical intervention. Since gene expression profiles of nodular fasciitis lesions reveal upregulation of genes encoding inflammatory cytokines and chemokines in comparison with other types of fibroblastic entities (5), one could speculate that these lesions are poised to regress, or that they are inflamed and would eventually regress in the absence of definitive treatment. In our patient, since there was no manipulation of the original lesion, it is likely that the initial regression was truly spontaneous.

Local recurrence of nodular fasciitis has rarely been reported. One of 250 patients in a large series experienced a local recurrence (1), and another study reported none in 15 patients (3) with an average follow-up period of 5.7 and 7.2 years, respectively. In contrast, a study describing 50 cases of nodular fasciitis of the external ear revealed a higher rate of local recurrence (9.3%), which was attributed to difficulties in obtaining clear surgical margins in the region around the ear (9). Of note, a detailed clinical-pathologic review of 134 cases of nodular fasciitis published more than 25 years ago reported that lesions recurred (18/134) only in patients who had been misdiagnosed initially (2). This study strongly supports the view that nodular fasciitis

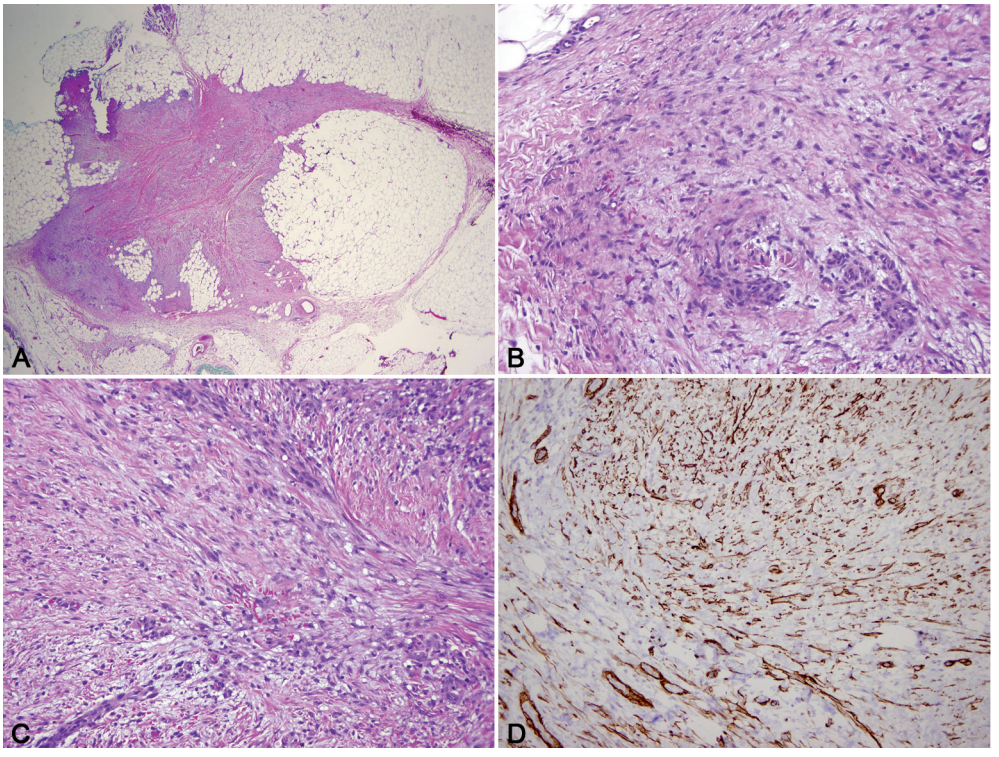

Fig. 1. (A) A small, circumscribed, but unencapsulated, nodular proliferation of predominantly spindle cells is present in the subcutis. The nodular lesion is contiguous with the subcutaneous fibrous septae and does not involve the overlying epidermis or the dermis. (B) There is a mild to moderately dense cellular proliferation of predominantly spindle shaped myofibroblasts with pale, eosinophilic/myxoid stroma. This histologic appearance resembles so-called "tissue-culture fibroblasts". (C) A moderately dense cellular proliferation of myofibroblasts is arranged in loose fascicles, resulting in a feathery appearance. Throughout the lesion are numerous thin-walled blood vessels lined by somewhat prominent endothelial cells. There are also microscopic foci of hemorrhage and scattered lymphocytes within the lesion. (D) Immunohistochemical staining for smooth muscle actin (SMA) shows distinct cytoplasmic staining of lesional myofibroblasts and the smooth muscle layer of the blood vessel wall. H&E stain; A: ×20, B: ×200, C: ×200, D: ×200.

is a non-recurrent lesion, and that recurrence of a lesion that was originally classified as nodular fasciitis should prompt a critical evaluation (2).

Some authors are of the opinion that incisional biopsy or complete excision may be considered for suspected nodular fasciitis lesions. Because nodular fasciitis is a benign condition, one might regard complete excision as overtreatment and favor incisional or fine needle biopsy, especially since these procedures may lead to lesional regression. On the other hand, a suspected nodular fasciitis lesion should be regarded as a potentially malignant tumor (sarcoma) until the final histologic diagnosis is made. In addition, lesion growth is rapid and regression is unpredictable, as there is a report of cranial fasciitis, a rare variant of nodular fasciitis in children, in which rapid tumor growth occurred after fine needle biopsy (10).

We conclude that nodular fasciitis can exhibit a spontaneously regressing and recurring course and favor complete excision of suspected nodular fasciitis lesions at an early stage to simultaneously provide suitable material for definitive histologic diagnosis and accomplish definitive treatment.

ACKNOWLEDGEMENTS

The authors thank Drs Edward Cowen and Maria Turner for helpful discussions.

This study was supported by Intramural programs of the National Cancer Institute and the National Institute of Allergy and Infectious Diseases.

The authors declare no conflict of interest.

REFERENCES

1. Shimizu S, Hashimoto H, Enjoji M. Nodular fasciitis: an analysis of 250 patients. Pathology 1984; 16: 161–166.
2. Bernstein KE, Lattes R. Nodular (pseudosarcomatous) fasciitis, a nonrecurrent lesion: clinicopathologic study of 134 cases. Cancer 1982; 49: 1668–1678.
3. Samaratunga H, Searle J, O'Loughlin B. Nodular fasciitis and related pseudosarcomatous lesions of soft tissues. Aust N Z J Surg 1996; 66: 22–25.
4. Wang XL, De Schepper AM, Vanhoenacker F, De Raeve H, Gielen J, Aparisi F, et al. Nodular fasciitis: correlation of MRI findings and histopathology. Skeletal Radiol 2002; 31: 155–161.
5. Bacac M, Migliavacca E, Stehle JC, McKee T, Delorenzi M, Coindre JM, et al. A gene expression signature that distinguishes desmoid tumours from nodular fasciitis. J Pathol 2006; 208: 543–553.
6. Nair P, Barrett AW, Theodossy T. Oral nodular fasciitis: case report. Br J Oral Maxillofac Surg 2004; 42: 360–362.
7. Wong NL. Fine needle aspiration cytology of pseudosarcomatous reactive proliferative lesions of soft tissue. Acta Cytol 2002; 46: 1049–1055.
8. Kato K, Ehara S, Nishida J, Satoh T. Rapid involution of proliferative fasciitis. Skeletal Radiol 2004; 33: 300–302.
9. Thompson LD, Fanburg-Smith JC, Wenig BM. Nodular fasciitis of the external ear region: a clinicopathologic study of 50 cases. Ann Diagn Pathol 2001; 5: 191–198.
10. Oh CK, Whang SM, Kim BG, Ko HC, Lee CH, Kim HJ, et al. Congenital cranial fasciitis – ''watch and wait'' or early intervention. Pediatr Dermatol 2007; 24: 263–266.