

CASE REPORT

A Case of Dermatofibroma of the Upper Lip

Hye In Lee, M.D., Jin Woong Lee, M.D., Tae Young Han, M.D., Kapsok Li, M.D.,
Chang Kwun Hong, M.D., Seong Jun Seo, M.D., Kye Yong Song, M.D.¹

Departments of Dermatology and ¹Pathology, College of Medicine, Chung-Ang University, Seoul, Korea

Dermatofibroma (DF) is a common benign mesenchymal tumor composed of fibroblastic and histiocytic cells. It occurs anywhere on the body surface but has a propensity for the extremities. To our knowledge, DF arising in the oral cavity, especially on the lip, is quite rare. DFs of the head and neck region have been known to be most often of the cellular type and frequently recur, so a wider initial excision is recommended. Herein we report a case of DF in a 41-year-old female who presented with a deep-seated nodule on her upper lip. (*Ann Dermatol* 22(3) 333~336, 2010)

-Keywords-

Dermatofibroma, Lip

INTRODUCTION

Dermatofibroma (DF), also known as benign fibrous histiocytoma (BFH), is a common benign mesenchymal tumor composed of fibroblastic and histiocytic cells¹. DF occurs anywhere on the body surface but has a propensity for the extremities. To our knowledge, DF arising in the oral cavity or perioral region is quite rare, and its long-term clinical behavior has yet to be elucidated. Herein we reported the clinical, microscopic, and immunohistochemical aspects of a case of DF of the upper lip.

CASE REPORT

A 41-year-old female came to our department for

evaluation of a hard nodule on her upper lip that had been noted 4 months earlier. Her medical and family histories were noncontributory. She denied any history of trauma to the lesion. Clinical examination revealed a rubbery, firm, non-mobile nodule, 1.0 cm in diameter, on the upper lip (Fig. 1). The appearance of the overlying labial mucosa and skin was normal. There was no pain or tenderness. Excision biopsy was performed, and the specimen was stained with hematoxylin-eosin.

Microscopically, a poorly defined, non-encapsulated, nodular tumor was observed throughout the depth of the dermis. Histological findings showed acanthosis of the epidermis and a poorly defined, non-encapsulated deep dermal tumor (Fig. 2A) composed of fibroblastic spindle cells without atypia and pleomorphism in a storiform pattern, separating the dermal collagen, with a few histiocytes in both the superficial (Fig. 2B) and deep portions of the tumor (Fig. 2C). Immunohistochemical stains for CD34, factor XIIIa, CD68 and smooth muscle actin (SMA) were then performed on sections of the tumor. CD34 was negative in fibroblastic spindle tumor



Fig. 1. Rubberly, firm, non-mobile nodule, 1.0×1.0 cm in diameter, on the upper lip with normal overlying labial mucosa and skin.

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Corresponding author: Seong Jun Seo, M.D., Department of Dermatology, Chung-Ang University Medical Center, 224-1 Heukseok-dong, Dongjak-gu, Seoul 156-755, Korea. Tel: 82-2-6299-1525, Fax: 82-2-823-1049, E-mail: drseo@hanafos.com

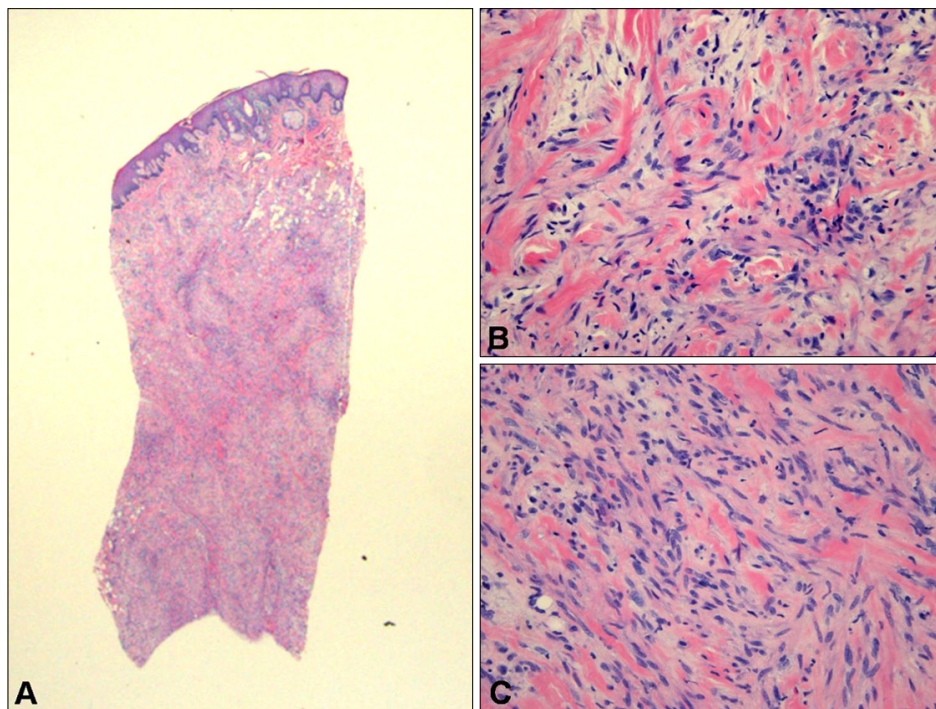


Fig. 2. (A) Histological findings show acanthosis of the epidermis and a poorly defined non-encapsulated deep dermal tumor (H&E, $\times 20$). (B, C) The tumor was found to be composed of fibroblastic spindle cells in a storiform pattern, separating dermal collagen with a few histiocytes both in the superficial (B) and deep portions of the tumor (C) (H&E, $\times 200$).

cells and positive in vessels (Fig. 3A), factor XIIIa was positive in only a few dendritic cells and negative in fibroblastic cells (Fig. 3B), CD68 was weak in spindle cells and strong in histiocytes (Fig. 3C), and SMA was strong in fibroblastic spindle cells (Fig. 3D). As a result of clinical and pathologic findings, the diagnosis was interpreted as DF.

DISCUSSION

DF is a common benign dermal fibrohistiocytic tumor that presents as a firm, red-brown nodule. There has been controversy about the pathogenesis of DF. Whether DF is a true neoplasm or reactive hyperplasia induced by mechanical stimuli is still unclear². Prolonged sun exposure, traumatic injury and chronic infection have been suggested as possible causative agents. DF occurs anywhere on the body surface, most commonly on the extremities, and usually on the lower legs. Involvement of the oral mucosa is quite rare. There have been a few published reports of DF that involved the oral and maxillofacial regions³⁻⁷. Gray et al.⁶ reviewed the literature on oral and perioral DF and reported that the buccal mucosa/vestibular region was the most commonly affected site. Also, cases of DF/BFH of the tongue, gingiva, mandible, maxilla, lower and upper lip have been described. Yamada et al.⁷ reported a case of BFH of the upper lip in an infant. Five cases of DF of the lip have

been reported in the literature (Table 1)^{3,5-7}.

Histologically, the epidermis demonstrates hyperplasia, hyperpigmentation of the basal layer, and elongation of the rete ridges ("dirty fingernail" sign), which are separated by a clear (Grenz) zone from the tumor in the dermis. The tumor itself is composed of fibroblast-like spindle cells, histiocytes, and blood vessels in varying portions.

DF has a variable immunohistochemical profile. Early lesions are highly reactive for macrophage markers such as CD68⁸. Most cells in early phase DFs have been found to react with factor XIIIa, a marker of normal dermal dendrocytes. This reactivity is mostly seen at the periphery of the tumor, continuously diminishes with aging of the lesion, and is completely absent in atrophic variants⁹. Other variably expressed markers include smooth muscle actin, CD56, and NSE. Labeling for SMA is most prominently seen in myofibroblastic DF¹⁰. Variable reactivity is seen with vascular markers such as factor VIII or CD31. Exceptional cases with characteristic histologic features of DF show diffuse reactivity for CD34¹¹; other rare cases express S-100 protein. Mentzel et al.¹² reported that tumor cells of facial DF showed immunoreactivity for factor XIIIa in 13 out of 17 cases and a focal immunopositivity for CD68 in 6 out of 10 cases; Mentzel et al.¹² also showed that spindle-shaped tumor cells in 16 out of 19 neoplasms stained at least focally positive for SMA. The more cellular forms of DF tend to express SMA,

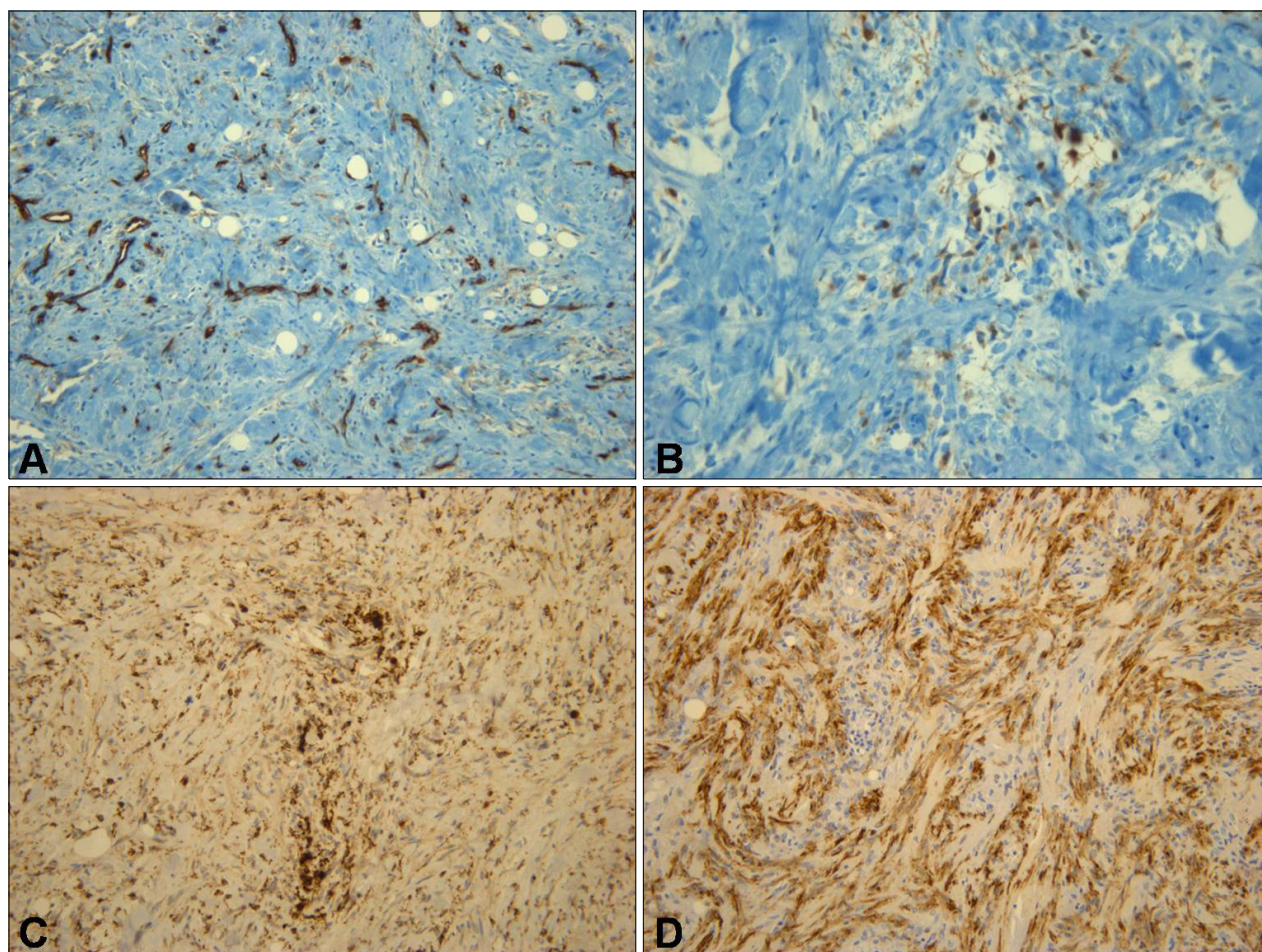


Fig. 3. (A) CD34 was negative in fibroblastic spindle cells and positive in vessels ($\times 200$). (B) Factor XIIIa was positive in only a few dendritic cells and negative in fibroblastic cells ($\times 200$). (C) CD68 was weak in spindle cells and strong in histiocytes ($\times 200$). (D) Smooth muscle actin (SMA) was strong in fibroblastic spindle cells ($\times 200$).

Table 1. Summary of 6 cases of benign fibrous histiocytoma of the lip reported in the literature

Patient No.	Age	Sex	History	Location	Size (mm)	Treatment	Date	Reference
1	52 yr	M	None	Lower lip	5×10	Excision	1975	Hillis and Beasley ³
2	22 yr	F	Trauma	Lower lip	20	Excision	1992	MacLeod and Jones ⁵
3	45 yr	M	None	Upper lip	20~30	Excision	1992	Gray et al. ⁶
4	45 yr	F	Unknown	Lower lip	Unknown	Excision	1992	Gray et al. ⁶
5	6 mo	M	None	Upper lip	15	Excision	2002	Yamada et al. ⁷
6	41 yr	F	None	Upper lip	10×10	Excision	2008	Present case

whereas the lesions with more epithelioid cells and giant cells tend to express histiocytic markers.

The main differential diagnoses of spindle cell neoplasms arising on the face, especially the oral cavity, include nodular fasciitis, low-grade myofibroblastic sarcoma, and neurofibroma. Nodular fasciitis may also present as spindle cells ordered in a storiform configuration, similar to that seen in DF, but the fascicles are separated by a myxoid stroma, mitoses are frequent and immunohisto-

chemical staining for S-100 antigen is reactive¹³. Low-grade myofibroblastic sarcoma is characterized morphologically by ill-defined fascicles of atypical, spindle-shaped myofibroblastic tumor cells that infiltrate preexisting structures in a diffuse pattern. Spindle-shaped tumor cells of DF do not stain positively for desmin, a marker that is frequently expressed in cases of low-grade myofibroblastic sarcoma¹⁴. Neurofibroma is composed of elongated spindle-shaped S-100 protein-positive tumor cells contain-

ing fusiform nuclei¹⁵. Cellular fibrous histiocytoma, the cellular variant of DF, must be especially distinguished from dermatofibrosarcoma protuberans (DFSP). DFSP is generally characterized by more uniform spindle cells and a more prominent storiform pattern than that seen in DF. CD34 staining is usually diffusely positive in DFSP and negative in most forms of DF. However, some DFs may express CD34, and some DFSP may express factor XIIIa¹⁶. Paradoxically, cellular atypia is often more pronounced in cellular fibrous histiocytoma than in DFSP, and factor XIIIa staining is often negative.

DFs arising on the face, including the oral cavity, are more often of the cellular type, extend into the subcutaneous fat or muscle, and frequently recur¹². Therefore, a wider initial excision is recommended, and local excision is the definitive treatment for DF of the head and neck region. Because oral DFs are rare and the long-term clinical behavior has yet to be elucidated, regular clinical follow-up is recommended.

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