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CASE REPORT

Desmoplastic Fibroblastoma of the Scalp Accompanied by Severe Pain; Unusual Location and Symptom

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Desmoplastic fibroblastoma is a rare fibrous tumor that usually presents as a painless, slow-growing mass in the subcutaneous tissues and skeletal muscles. It has a wide anatomic distribution, with the most common involvement being the arm and shoulder. Here, we report a case of a tiny painful desmoplastic fibroblastoma arising on the scalp. According to a microscopic examination, this tumor was composed of spindle-shaped fibroblasts in the dense collagenous stroma. On immunohistochemical staining, tumor cells were positive for vimentin and negative for smooth muscle actin, CD34, and S100. Our case is unique in that desmoplastic fibroblastoma developed on the scalp and there was presence of pain despite its small size. **(Ann Dermatol 30(6) 712~715, 2018)**

-Keywords-
Collagenous fibroma, Desmoplastic fibroblastoma, Pain, Scalp

INTRODUCTION

Desmoplastic fibroblastoma, also known as collagenous fibroma, is a benign fibrous neoplasm composed of spindle to stellate-shaped fibroblasts sparsely distributed in the rich collagenous matrix. Since Evans' first description in 1995[1], approximately 100 cases of desmoplastic fibroblastoma have been reported in the English literature. It has conventionally been presented as a well-circumscribed, solitary, non-tender, and slow-growing mass in the subcutaneous tissue or intramuscular region[2,3], commonly affecting individuals between the ages of 50 and 70 years[4]. Desmoplastic fibroblastoma most commonly appears in the arm and shoulder, but it has also been reported to appear on the neck, back, hip, and extremities[5]. The current treatment of choice is complete surgical excision, and there have not been any reports of recurrence or metastases thus far[3]. Herein, we report an extraordinary case of desmoplastic fibroblastoma that presented as a tiny painful mass on the scalp with clinical, pathological, and immnohistochemical findings.

CASE REPORT

An 82-year-old man presented a tiny painful tender mass on the left posterior scalp over the course of 6 months. On physical examination, the mass was very small; it was not palpable. At the beginning, the lesion seemed to cause pain only upon touching; but with progression, patient complained of pain even in resting state. Pain was intermittent and aggravated with cold air exposure. Previously at another hospital, he was treated using triamcinolone acetonide and hyaluronidase intralesional injection, but with no effect. Ultrasonography showed a 0.4×0.2×0.4

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cm-sized hypoechoic nodule on aponeurosis with no compressibility and no vascularity (Fig. 1). However, the lesion was too small to be characterized on an ultrasonography. The patient did not want surgical removal.

After 10 weeks, the patient revisited our clinic with aggravated pain in the lesion. The pain was so severe that he was unable to sleep for the past three days. There was no apparent change on palpation. An emergency operation was performed under the impression of nerve origin tumor (Fig. 2). We received the patient's consent form about publishing all photographic materials. Macroscopically, the tumor was oval in shape and firm, with a size of $0.5 \times 0.3 \times 0.3$ cm. Microscopically, the tumor was composed of spindle-shaped cells embedded in the dense collagenous stroma, with moderately increased cellularity and nuclear pleomorphism (Fig. 3). No mitosis or necrosis was observed. Immunohistochemically, spindle-shaped cells were positive for vimentin, but negative for α-SMA, CD34, and S100. The degree of proliferation measured with Ki-67 showed only a mild expression (Fig. 4). Based on the pathological findings, the tumor was finally diagnosed as a desmoplastic fibroblastoma. Pain disappeared after surgery, and there were no signs of recurrence during the three-month follow-up period.

DISCUSSION

Although there have been reports of the clinicopathological features of desmoplastic fibroblastoma since 1995, it may be an unfamiliar tumor to many dermatologists due to its rarity[3,4]. According to Miettinen and Fetsch[5], desmoplastic fibroblastoma has been observed in the arms (24%), shoulders (19%), posterior area of the neck or upper back (14%), ankles and feet (14%), legs (14%), hands (8%), as well as the abdominal wall and hip (6%). To the best of our knowledge, this is the first report of desmoplastic fibroblastoma presented on the scalp, aside from some other unusual locations on the head, such as oral cavities, parotid gland, forehead, and orbital rims[6-9]. Furthermore, our case is unique in that severe pain and tenderness were accompanied with the tumor, which is highly unusual. In most previous cases, patients displayed no subjective symptoms, including pain or tenderness.

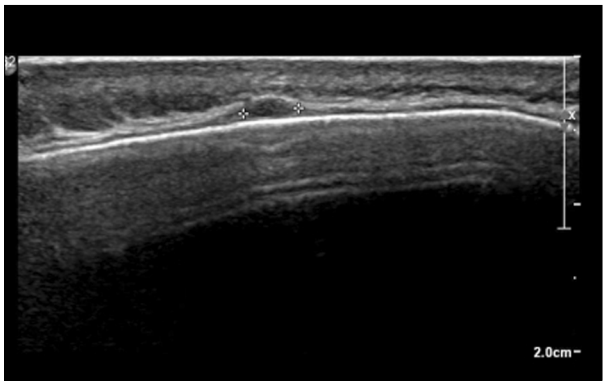

Fig. 1. Ultrasonography of the scalp. The tumor is shown on the aponeurosis as a hypoechoic mass.

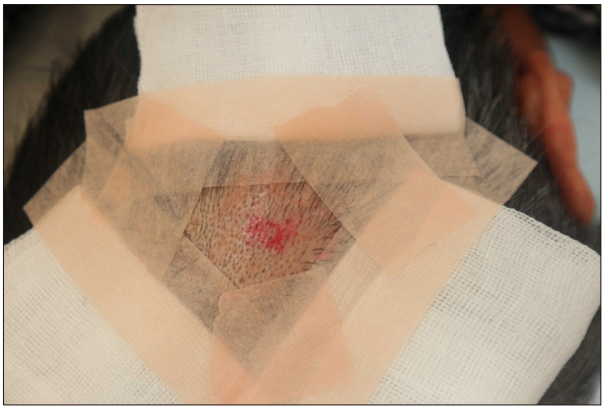

Fig. 2. The operation field of the tumor. Suspected tumor contour is indicated with red circle.

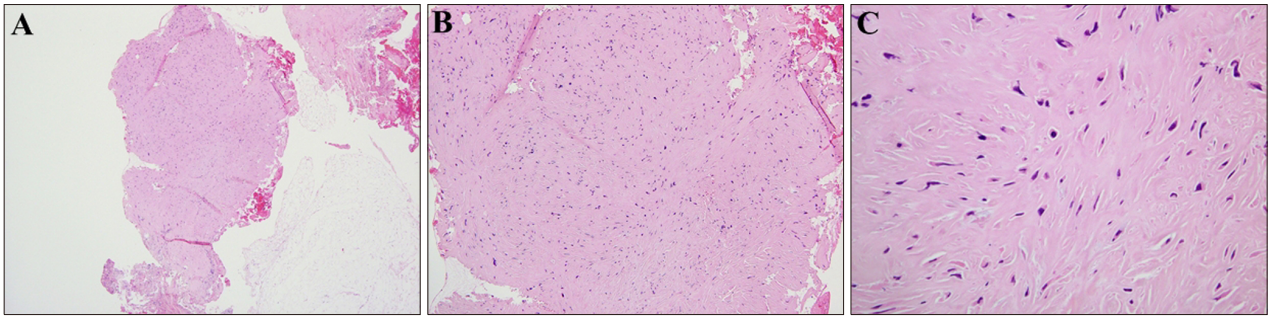

Fig. 3. (A) Spindle cell neoplasm without encapsulation (H&E, ×40). (B) The tumor is composed of spindle-shaped cells surrounded by dense fibrous stoma (H&E, ×100). (C) No mitosis and necrosis are found. Note that cellularity and nuclear pleomorphism are moderately increased (H&E, ×400).

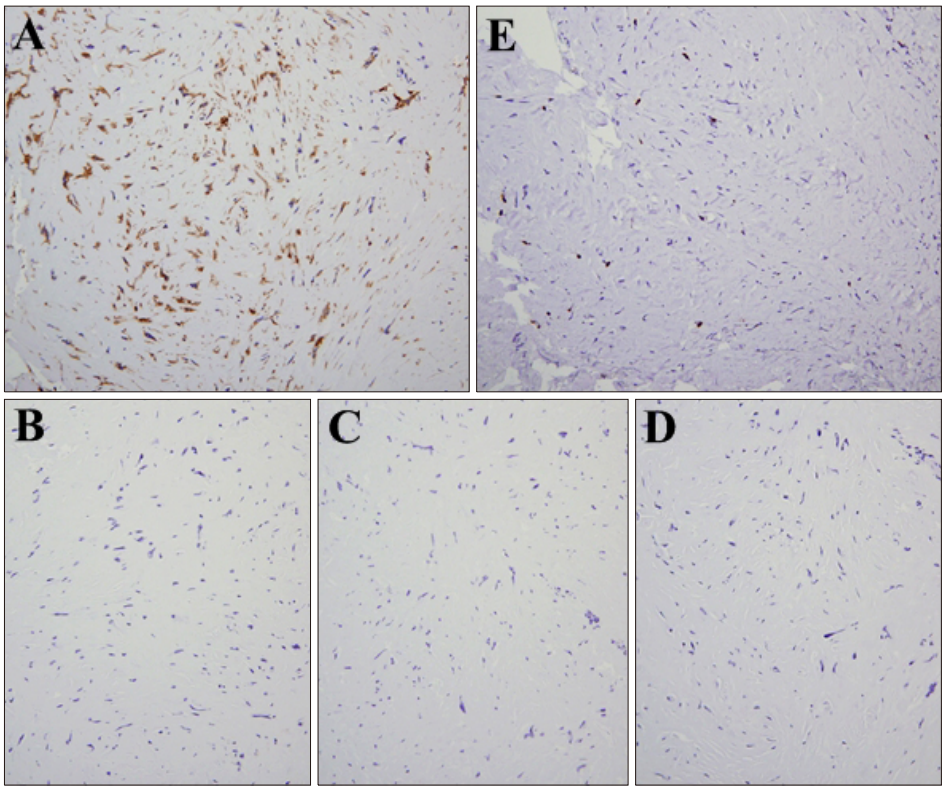

Fig. 4. Immunohistochemical staining of desmoplastic fibroblastoma shows that tumor cells are positive for (A) vimentin (immunoperoxidase staining, ×200), but negative for (B) α-SMA (immunoperoxidase staining, ×200), (C) CD34 (immunoperoxidase staining, ×200), and (D) S100 (immunoperoxidase staining, ×200). (E) Proliferation index estimated by Ki-67 is mildly expressed (immunoperoxidase staining, ×200).

Additionally, with respect to size, our case was unusually smaller than the typical desmoplastic fibroblastoma observed previously to have a maximum diameter of approximately 4 cm[1,5]. Neurological symptoms have not been reported as a feature of desmoplastic fibroblastoma, despite frequent nerve entrapment of the tumor[5]. Only in one previous report, desmoplastic fibroblastoma on the right neck resulted in sensorimotor deficit of the right arm and shoulder[3]. In that case, however, the neurological symptom was likely due to the compression of the right upper cervical plexus and the right spinal accessory nerve by the tumor. In our case, the tumor was too small to invade or affect any adjacent nerves, and according to our microscopic findings, there was no evidence of nerve invasion.

Histologically, desmoplastic fibroblastoma is a hypocellular tumor with spindle to stellate-shaped fibroblasts in the dense fibroumyxoid stroma[10]. With respect to the immunohistochemistry of desmoplastic fibroblastoma, our results were consistent with previous reports: Positivity for vimentin, variable expressions of α-SMA, and negativity for CD34, desmin, and S100[11].

Differential diagnosis for desmoplastic fibroblastoma includes fibromatosis, neurofibroma and low grade fibromyxoid sarcoma. Fibromatosis is more cellular and has more prominent vascular structures than desmoplastic fibroblastoma[2]. Neurofibroma may have similar fibromyxoid stroma but cells are strongly positive for S100[12]. A low grade fibromyxoid sarcoma is distinguishable from desmoplastic fibroblastoma by increased cellularity and characteristic swirling, whorled growth pattern[13].

Conclusively, we reported a unique case of desmoplastic fibroblastoma on the scalp presenting with unusual tenderness and pain. Complete excision was performed, resulting in complete improvement of pain and tenderness.

CONFLICTS OF INTEREST

The authors have nothing to disclose.

REFERENCES

1. Evans HL. Desmoplastic fibroblastoma. A report of seven cases. Am J Surg Pathol 1995;19:1077-1081.
2. Jang JG, Jung HH, Suh KS, Kim ST. Desmoplastic fibroblastoma (collagenous fibroma). Am J Dermatopathol 1999;21:256-258.
3. Fong F, Odell E, Simo R. Collagenous fibroma (desmoplastic fibroblastoma) of the neck presenting with neurological symptoms. Head Neck Pathol 2009;3:47-50.
4. Park JM, Koh JK, Lee MG. Unusual case of desmoplastic fibroblastoma (collagenous fibroma) with ulceration on toe: mimicking diabetic foot ulcer. J Dermatol 2012;39:420-422.

5. Miettinen M, Fetsch JF. Collagenous fibroma (desmoplastic fibroblastoma): a clinicopathologic analysis of 63 cases of a distinctive soft tissue lesion with stellate-shaped fibroblasts. Hum Pathol 1998;29:676-682.
6. Stacy RC, Jakobiec FA, Fay A. Collagenous fibroma (desmoplastic fibroblastoma) of the orbital rim. Ophthal Plast Reconstr Surg 2013;29:e101-104.
7. Weisberg NK, DiCaudo DJ, Meland NB. Collagenous fibroma (desmoplastic fibroblastoma). J Am Acad Dermatol 1999; 41:292-294.
8. de Sousa SF, Caldeira PC, Grossmann Sde M, de Aguiar MC, Mesquita RA. Desmoplastic fibroblastoma (collagenous fibroma): a case identified in the buccal mucosa. Head Neck Pathol 2011;5:175-179.
9. Nagaraja V, Coleman HG, Morgan GJ. Desmoplastic fibroblastoma presenting as a parotid tumour: a case report and review of the literature. Head Neck Pathol 2013;7:285-290.
10. Kim JH, Oh DY, Kim SW, Lee JH, Ahn ST, Rhie JW. Desmoplastic fibroblastoma of the finger tip in an adult. Arch Plast Surg 2012;39:84-86.
11. Nielsen GP, O'Connell JX, Dickersin GR, Rosenberg AE. Collagenous fibroma (desmoplastic fibroblastoma): a report of seven cases. Mod Pathol 1996;9:781-785.
12. Back SJ, Kim DH, Kim N, Lee Y, Seo YJ, Park JK, et al. A case of isolated plexiform neurofibroma in a patient with myasthenia gravis. Ann Dermatol 2009;21:53-55.
13. Watanabe H, Ishida Y, Nagashima K, Makino T, Norisugi O, Shimizu T. Desmoplastic fibroblastoma (collagenous fibroma). J Dermatol 2008;35:93-97.