

A Case of Superficial Angiomyxoma

Kozo YAMAMOTO^{*1}, Akio KONDO^{*1}, Kenichi IWASHITA^{*1}, Yoshinori UMEZAWA^{*1},
Yukinori OHTA^{*1}, Takashi MATSUYAMA^{*1}, Akira OZAWA^{*1}, Kohta ICHIKAWA^{*2},
Muneo MIYASAKA^{*2}, Chie INOMOTO^{*3} and Masanori YASUDA^{*3}

^{*1} Department of Dermatology, (Chief: Prof. A. OZAWA)

^{*2} Department of Plastic and Reconstructive Surgery, (Chief: Prof. R. TANINO)

^{*3} Department of Pathology, Tokai University School of Medicine (Chief: Prof. Y. UEYAMA)

(Received January 24, 2006; Accepted March 14, 2006)

We report a case of a 19-year-old man who showed a brownish, elastic-soft, multilocular, pedunculated, solid tumor (10 × 6 × 6 cm), with scale crust and erosion, on the medial side of the right thigh. The histopathology of a specimen removed completely revealed a tumor that was located between the middle layer of the dermis and the subcutaneous fatty tissue, was filled with a mucoid material, and was surrounded by a fibrous tissue. The tumor consisted of small thin-walled blood vessels and spindle-shaped or stellate tumor cells without cytological atypia in addition to the mucoid material. Immunohistochemistry revealed that the tumor cells express the vimentin stain. Based on these clinical and histologic findings, we diagnosed the skin condition as superficial angiomyxoma.

Key words: superficial angiomyxoma, carney's complex

INTRODUCTION

In 1988, Allen *et al.* reported cutaneous tumors that were histologically similar to cutaneous myxomas of Carney's complex but had no evidence of the complex as superficial angiomyxomas in 28 patients [1]. Superficial angiomyxoma is a rare benign neoplasm characterized by a conglomerate of multiple, moderately to sparsely cellular angiomyxoid nodules with scattered small blood vessels. Therefore we report one rare case of superficial angiomyxoma.

CASE REPORT

A 19-year-old Japanese man found an asymptomatic mass on the medial side of the right thigh in 2002. The mass enlarged slowly, and he did not seek medical advice until normal daily friction caused erosion on the mass surface. He was referred to our hospital by a local dermatologist on April 9, 2004. The initial physical examination showed a brownish, elastic-soft, multilocular, pedunculated, solid tumor (10 × 6 × 6 cm), with scale crust and erosion, hanging from the skin surface on the medial side of the right thigh (Fig. 1). The tumor was removed completely. The transected tumor contained a yellow gelatinous substance separated by fibrous septa (Fig. 2). Pathological studies of hematoxylin-eosin-stained tumor sections at lower magnification revealed the normal epidermis and a tumor that was located between the middle layer of the dermis and the subcutaneous fatty tissue, was filled with a mucoid material, and was surrounded by a fibrous tissue (Fig. 3a). At higher magnification, the tumor consisted of small thin-walled blood vessels and spindle-shaped or stellate tumor cells without cytological atypia in addition to the mucoid material (Fig. 3b). The tumor cells were nega-



Fig. 1 A brownish, elastic-soft, multilocular, pedunculated, solid tumor (10 × 6 × 6 cm), with scale crust and erosion, on the medial side of the right thigh.



Fig. 2 The transected tumor contained a yellow gelatinous substance separated by fibrous septa.

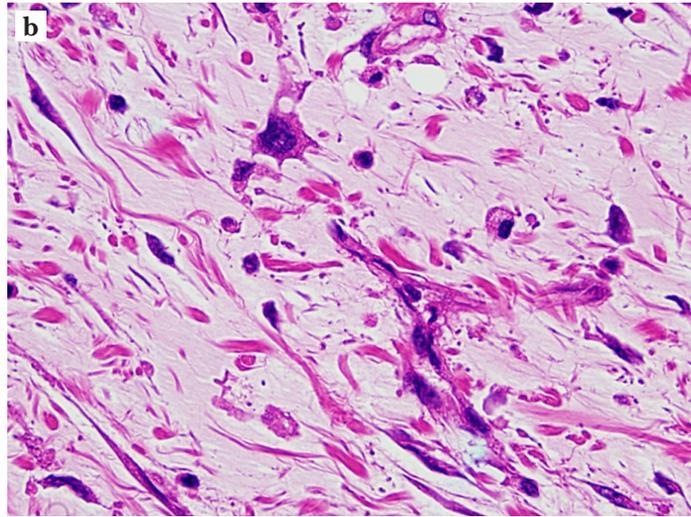
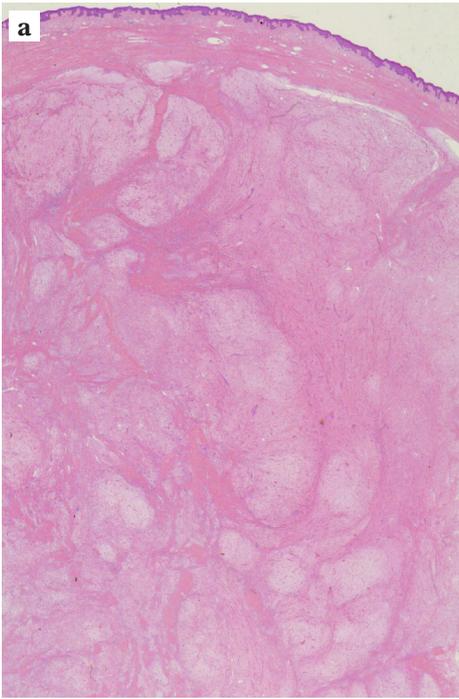


Fig. 3 **a:** The tumor, filled with a mucoid material, and surrounded by a fibrous tissue was located between the dermis and the subcutaneous fatty tissue. (Hematoxylin-eosin stain; original magnifications: $\times 40$) **b:** Small thin-walled blood vessels and spindle shaped or stellate tumor cells without cytological atypia in addition to the mucoid material. (Hematoxylin-eosin stain; original magnifications: $\times 100$)

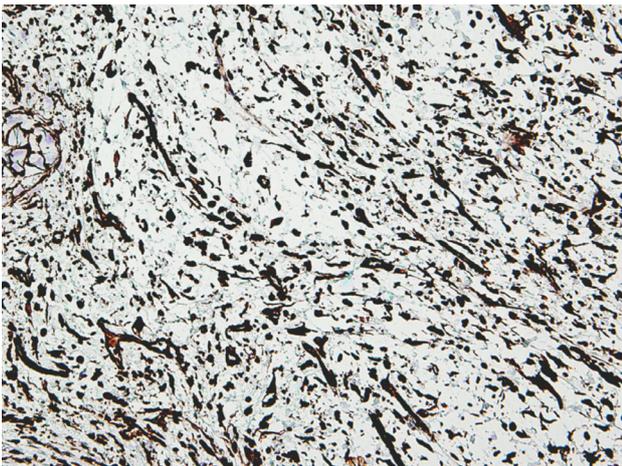


Fig. 4 Immunohistochemistry: almost all tumor cells showed expression of the vimentin stain. (Vimentin stain; original magnifications: $\times 100$)

tive for various immunohistochemical stains except the vimentin stain (Fig. 4). Based on these clinical and histologic findings, we diagnosed the skin condition as superficial angiomyxoma. The tumor has not recurred for 1.5 years since total removal.

DISCUSSION

Superficial angiomyxoma was first described as a cutaneous myxoma of Carney's complex by Carney *et al.* in 1986 [2]. Carney's complex is an autosomal dominant syndrome characterized by myxomas of the heart, skin, and breast; spotty pigmentation of the mucous membrane; and endocrine overactivity such as Cushing's syndrome and acromegaly [2].

In 1988, Allen *et al.* reported cutaneous tumors that were histologically similar to cutaneous myxomas of Carney's complex but had no evidence of the complex

as superficial angiomyxomas in 28 patients aged 4 to 78 years (mean, 39 years) [1]. Most of the tumors were soft and multilocular, measuring 0.5 to 9 cm, on the head, neck, trunk, or lower extremity. Microscopically, they were found between the dermis and the subcutaneous tissue and consisted of a mucoid material and small blood vessels.

In 1999, Calonje *et al.* analyzed and reported clinicopathologic and immunohistochemical features of superficial angiomyxoma as an independent disease entity in 39 patients [3]. The tumor was more common in men than women. The age of onset ranged from 0 to 82 years (median, 45.5 years; mean, 41.2 years). The tumor size ranged from 1 to 5 cm (mean, 2.3 cm). Immunohistochemically, tumor cells were positive for vimentin but negative for CD34, α -smooth muscle actin, HHF-35, S-100 protein, desmin, cytokeratin, and glial fibrillary acidic protein. However, Fetsch *et al.* described that superficial angiomyxoma could be positive for desmin and α -smooth muscle actin [4].

Our patient did not seem to have Carney's complex because his cutaneous tumor was not accompanied by non-cutaneous myxomas, pigmentation of the skin or mucous membrane, or endocrine disorders. We diagnosed the skin condition as superficial angiomyxoma based on clinical and histologic findings of the tumor. Pedunculated superficial angiomyxoma often hangs from the skin surface, as with our case, because the mucous tumor arising from the soft skin is easy to grow downward due to gravity [5]. In our case, the tumor was as large as $10 \times 6 \times 6$ cm. This growth can be explained by a delayed hospital visit resulting from the asymptomatic mass on the medial side of the thigh, which was difficult for the patient to see and would likely be embarrassing to reveal.

To our knowledge, no metastasis or malignancy of superficial angiomyxoma has been reported. However, long-term follow-up studies have revealed local recur-

rence of the tumor. The recurrence rate is 30% to 40%, associated with inadequate resection, according to Allen *et al.* and Calonje *et al.* [1, 3]. Our patient has had no recurrence for 1.5 years since total removal of the tumor.

In our case, the superficial angiomyxoma was negative for various immunohistochemical stains except the vimentin stain. A variety of immunohistochemical studies have been conducted to differentiate the tumor from other similar tumors or to explore the origin of tumor cells. Many of such studies have demonstrated that superficial angiomyxoma is positive only for vimentin, whereas others have produced different results [3, 4]. Since immunohistochemical features of superficial angiomyxoma have not been established, some researchers mention that immunohistochemical studies are not helpful for differential diagnosis of myxoid tumors [6]. More reports and further analyses will be needed to characterize superficial angiomyxoma clinically and immunohistochemically.

REFERENCES

- 1) Allen PW, Dymock RB, MacCormac LB: Superficial angiomyxomas with and without epithelial component. Report of 30 tumors in 28 patients. *Am J Surg Pathol* 12: 519-530, 1988.
- 2) Carney JA, Headington JT, Su WPD: Cutaneous myxomas. A major component of the complex of myxomas, spotty pigmentation, and endocrine overactivity. *Arch Dermatol* 122: 790-799, 1986.
- 3) Calonje E, Guerin D, MacCormac D, Fletcher CDM: Superficial Angiomyxoma. *Am J Surg Pathol* 23: 910-917, 1999.
- 4) Fetsch JF, Laskin WB, Tavassoli FA: Superficial Angiomyxoma (Cutaneous Myxoma). *Int J Gynecol Pathol* 16: 325-334, 1997.
- 5) Matsuda S, Oniki S, Kunisada M, Murata Y, Kumano K: Superficial Angiomyxoma. *Jpn J Dermatol* 115: 1333-1338, 2005. (In Japanese)
- 6) Nakayama H, Hiroi M, Kiyoku H, Naruse K, Enzan H: Superficial Angiomyxoma of the Right Inguinal Region. *Jpn J Clin Oncol* 27: 200-203, 1997.