Two Cases of Solitary Adult Myofibroma

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Solitary myofibroma is a recently described benign neoplasm of the skin or superficial soft tissue and it represents the adult counterpart of infantile myofibromatosis. This new clinicopathological entity is being recognized increasingly. We report two cases of solitary adult myofibroma. A skin-colored, dome-shaped hard subcutaneous nodule was found on the neck of a 62-year-old female and a tender, bean-sized, movable hard subcutaneous deep-seated nodule was found on the trunk of a 31-year-old female.
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Key Words: Myofibroma, Adult, Solitary

INTRODUCTION

In 1954 Stout first described infantile myofibromatosis under the designation of "congenital generalized fibromatosis." This benign condition was characterized by the presence of solitary or multiple fibrous nodules in the mesenchymal tissue of newborns[1]. In 1981, Chung and Enzinger reviewed 61 cases of this disorder and proposed the term "infantile myofibromatosis" due to the age of onset and histologic features of the tumors[2]. Benign myofibromas normally present as dermal, subcutaneous, or rarely deep muscle lesions. The vast majority of myofibromas present in children less than 2 years of age. However, similar lesions have been diagnosed in adults. The terms myofibroma and myofibromatosis have been used to describe the solitary and multicentric forms, respectively. The solitary forms are, at least, twice more common than the multiple forms. The occurrence of such lesions in adults has only recently been recognized.

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CASE REPORT

Case 1.

A 62-year-old female presented with a solitary subcutaneous nodule on her neck. The mass was first noticed 1 year prior to presentation and had been growing slowly. Examination revealed a tender, 1.0 × 1.0 cm in size, slightly movable, normal skin colored, dome-shaped hard subcutaneous nodule on the neck (Fig. 1). The nodule was clinically diagnosed as an epidermal cyst, pilomatricoma and myofibroma, and it was surgically excised. Examination of hematoxylin and eosin-stained sections revealed that the tumor was circumscribed, but not encapsulated. It was composed largely of spindle cells with oval vesicular nuclei and eosinophilic cytoplasms. Some areas demonstrated more rounded cells. Mitotic figures were scarce. In areas the biphasic nature was evident with smaller rounded cells surrounded by elongated spindle cells (Fig. 2A, B). There were alternating areas of fibrosis that stained blue with Masson's trichrome and hemangiopericytoma-like perivascular spindle cell areas that stained red (Fig. 3).

Immunohistochemically, the spindle cells were positive for vimentin and smooth muscle actin, but were negative for desmin and S-100 protein, compatible with myoepithelial differentiation (Fig. 4). The diagnosis of myofibroma was made. No evidence of recurrence has been found after 4 months of follow-up.

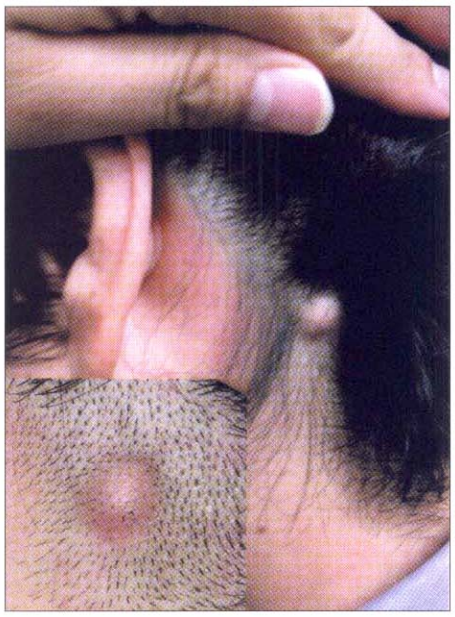

Fig. 1. A tender, 1.0 × 1.0 cm in size, slightly movable, normal skin-colored, dome-shaped hard subcutaneous nodule on the neck.

Case 2.

A 31-year-old female presented with a mild painful subcutaneous nodule on the trunk. This was of insidious onset and progressive in nature, and there was no past history of trauma. Physical examination revealed a tender, large bean-sized, slightly movable hard subcutaneous deep-seated nodule on the trunk (Fig. 5). The clinical impression included lipoma, angiolipoma, myositis ossificans and nodular fasciitis, and it was surgically removed by incisional biopsy. Histopathologic examination revealed a benign spindle cell tumor with marked vascularity and no evidence of mitotic activity. The perivascular spindle cell proliferation was more pronounced at the periphery of the tumor (Fig. 6A). The central portion of the mass was largely fibrotic, with residual vascularity in some fibrotic areas (Fig. 6B). The spindle cell areas of the tumor showed immunoreactivity to vimentin and smooth muscle actin, but were negative for desmin and S-100 protein (Fig. 7), compatible with myoepithelial differentiation. The diagnosis of myofibroma was made.

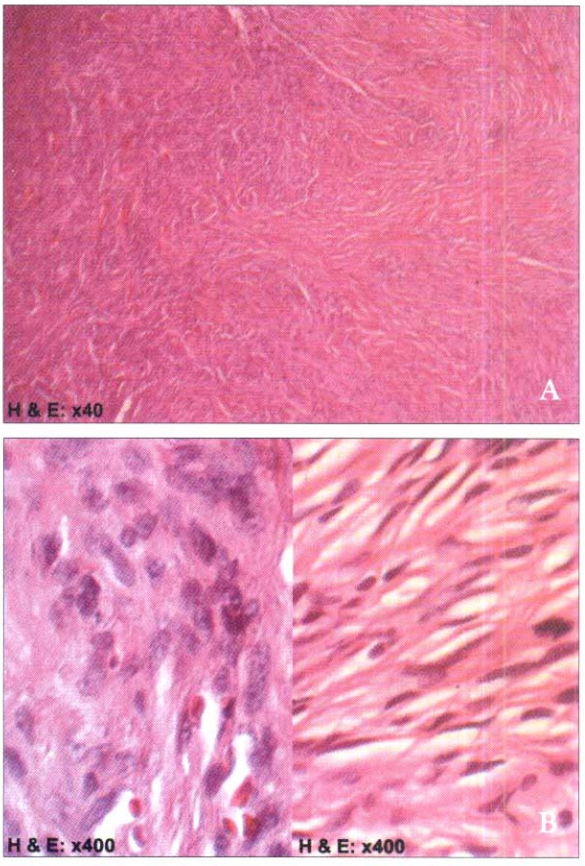

Fig. 2. A biphasic nature of the myofibroma with elongated spindle cells in the right half of the field and more rounded cells with eosinophilic cytoplasms in the left half of the field (A and B, H & E: A, × 40, B, × 400).

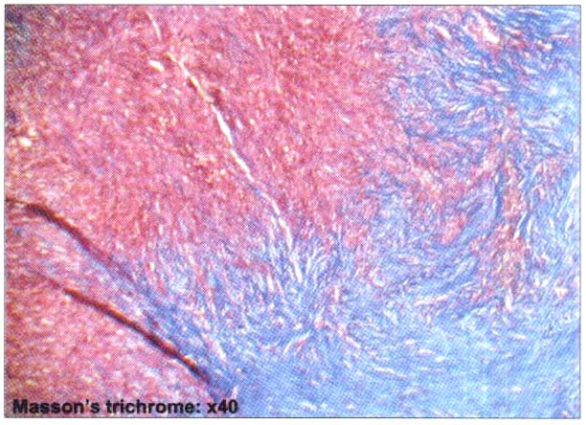

Fig. 3. Alternating area of fibrosis that stain blue with Masson's trichrome and hemangiopericytoma-like perivascular spindle cell areas that stain red (Masson's trichrome: × 40).

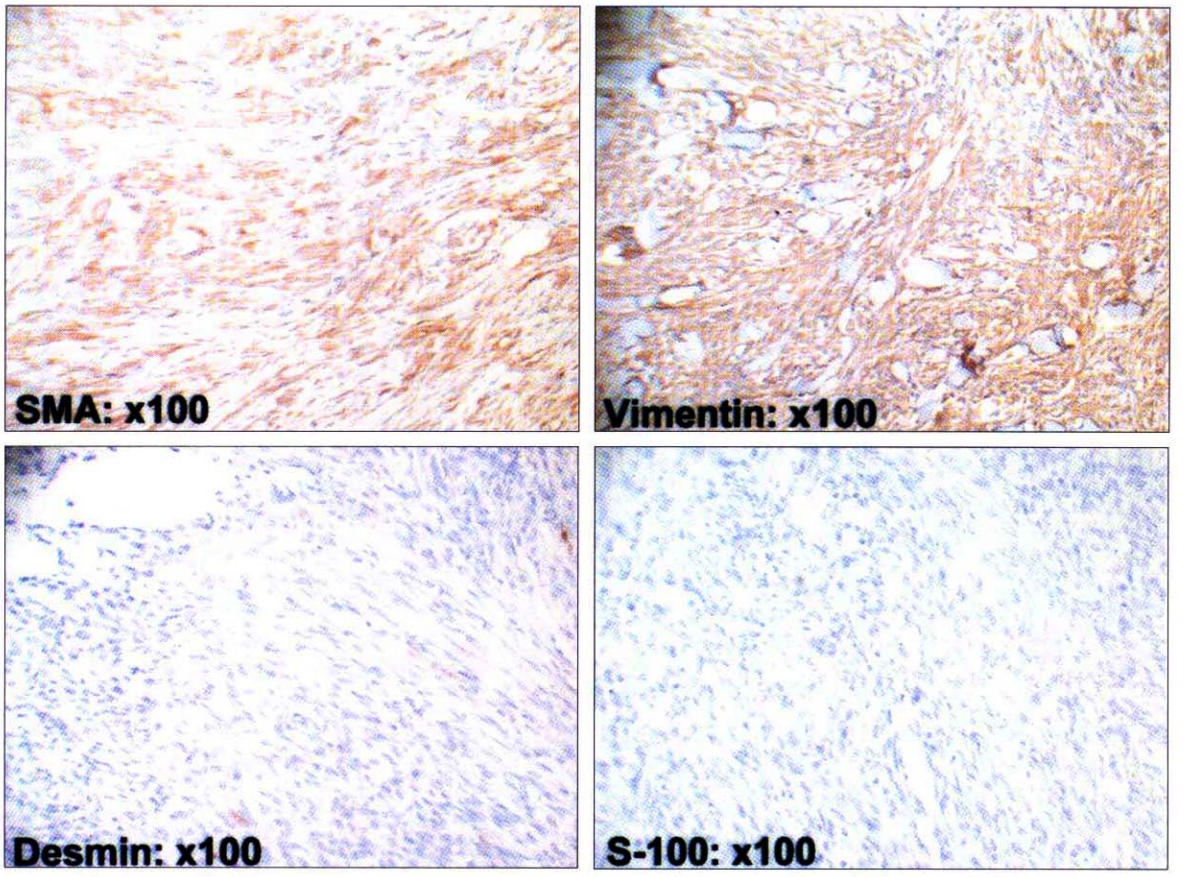

Fig. 4. Spindle shaped cells show immuno-reactivity to smooth muscle actin and vimentin but are negative to desmin and S-100 (original magnifications: × 100).

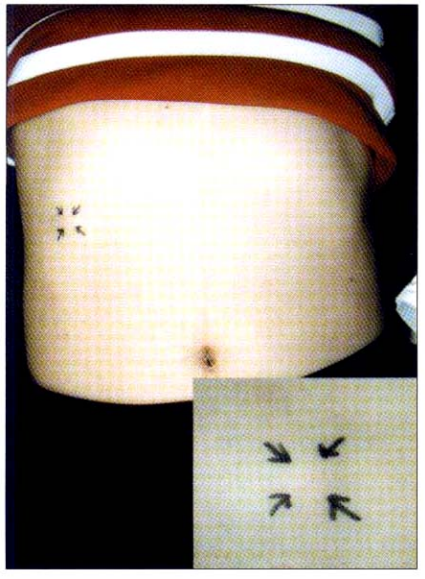

Fig. 5. A tender, large bean-sized, slightly movable hard subcutaneous deep-seated nodule on the trunk.

DISCUSSION

Infantile myofibromatosis was first described in 1954 by Stout as congenital generalized fibromatosis[1] and since then numerous examples of this entity have been reported in the medical literature. Even though it has been variously described as generalized hamartosis, multiple congenital mesenchymal hamartoma, multiple vascular leiomyoma, or metastatic congenital fibrosarcoma in the literature, the term infantile myofibromatosis has been preferred. This is due to the microscopic resemblance to smooth muscle tissue, the electron microscopic findings and the need to clearly distinguish this process from the locally more aggressive desmoid type of infantile fibromatosis. As implied by the name, infantile myofibromatosis is often diagnosed in newborns and infants, with approximately 90% of cases presenting in the first 2 years of life.

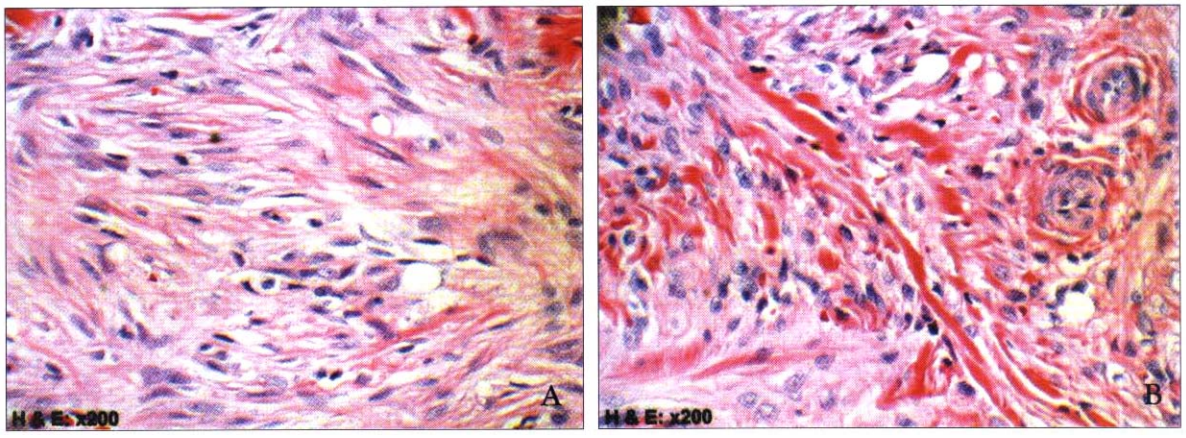

Fig. 6. A. The perivascular spindle cell proliferation is more pronounced at the periphery of the tumor (H&E: × 200). B. The central portion of the mass is largely fibrotic, with residual vascularity in some fibrotic areas (H&E: × 200).

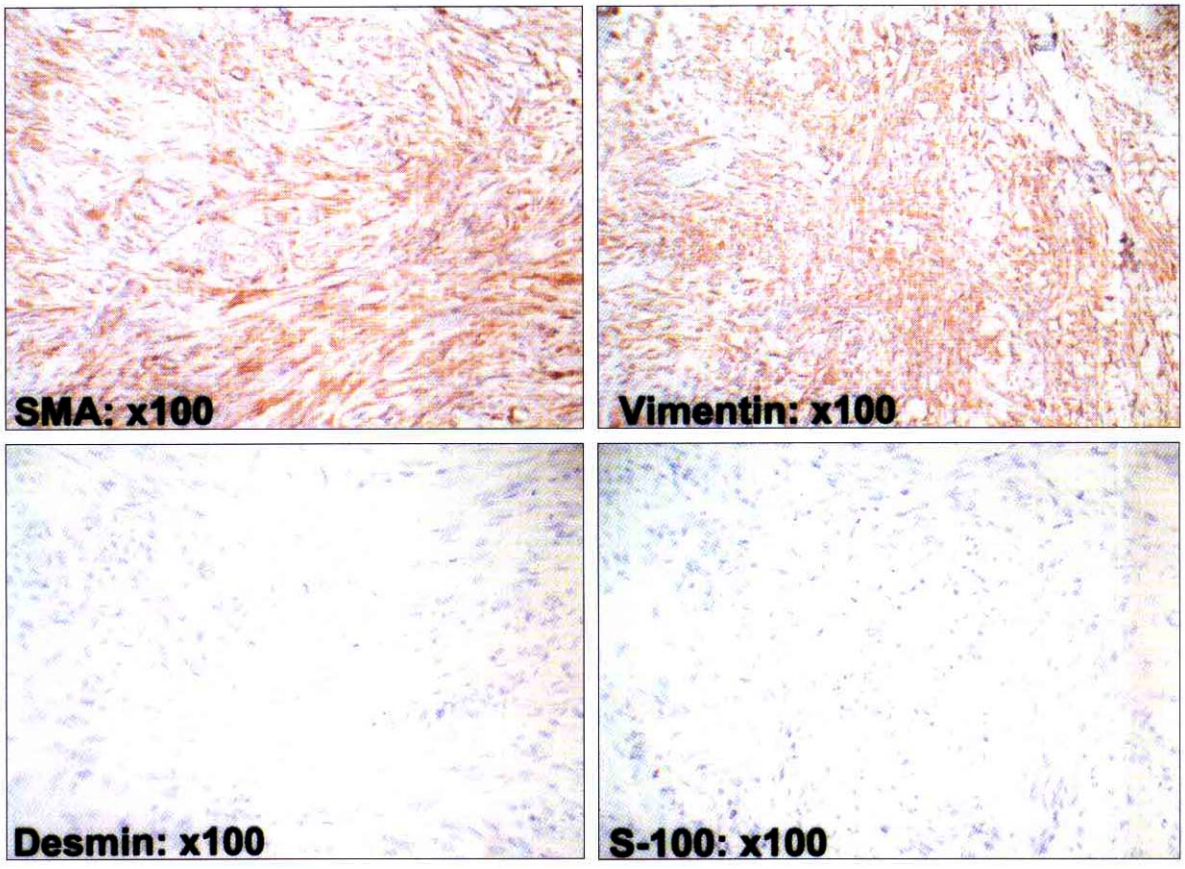

Fig. 7. The spindle cell areas of the tumor show immunoreactivity to vimentin and smooth muscle actin, but are negative for desmin and S-100 protein (original magnifications: × 100).

However, several cases in adults have been reported[3].

In 1981 Chung and Enzinger reviewed 61 cases of the disorder, and distinguished two distinct forms:

a multicentric form and a solitary form. Of the 61 cases, 16 were multicentric and 45 were solitary. The multicentric form occurs almost without exception in neonates and infants, and this form can be further divided into somatic and somatovisceral types. If the condition is entirely somatic, it means a good prognosis, often with eventual regression of lesions or no recurrence of lesions that are surgically excised. However, visceral involvement of the lungs, heart, and/or gastrointestinal tract with multicentric lesions is an ominous sign[1,2]. The data reported by Chung and Enzinger showed that the solitary form of this disorder is more frequent and has a slight male predominance, while the multicentric form is slightly more predominant in females. The solitary form is also predominantly a disease of the first year of life, but does occur sporadically in adolescence and young adulthood[4,5]. In cases without visceral involvement, many lesions regress spontaneously or do not recur after surgery and the prognosis is excellent[1,2].

Clinically, the solitary myofibroma almost invariably presents as a painless, firm, occasionally bluish, cutaneous or subcutaneous nodule. It is found most commonly in the region of the head and neck[6]. Histopathologically, the lesion has a distinctive appearance, well recognized in children but much less so in adults. Classically, it manifests a biphasic pattern or a zoning arrangement of two cell types[7]. The central cells are smaller with large pale staining nuclei, polygonal or round and often associated with dilated irregular blood vessels which gives a hemangiopericytomatous appearance. At the periphery of the lesion, spindle-shaped cells predominate. These cells have eosinophilic cytoplasms, arranged in short bundles and fascicles resembling leiomyoma. These cells demonstrate features of both myofibroblasts and fibroblasts[3,6]. Myofibroblastic differentiation of the tumor cells is supported by their immunophenotype. The spindle cells are desmin negative but smooth muscle actin positive.

Differential diagnosis includes nodular fasciitis, neurofibroma, benign fibrous histiocytoma, leiomyoma and leiomyosarcoma, dermatomyofibroma and hemangiopericytoma. The peripheral areas of myofibroma in adulthood can have the pooly circumscribed, non-fasciculated appearances of nodular fasciitis or a more cellular fascicular pattern reminiscent of leiomyoma. Both these tumors lack a biphasic pattern or hemangiopericytic areas, and nodular fasciitis in the dermis is extremely rare. Cutaneous smooth muscle tumors are almost invariably desmin positive and also lack a biphasic pattern; in addition leiomyosarcomas are generally more mitotic and more pleomorphic. The central areas of adult myofibroma closely resemble a hemangiopericytoma, from which it can be differentiated on the basis of the peripheral myoid areas and consistent reactivity to actin. Furthermore, hemangiopericytomas are very rarely so superficial in location. Most fibrous histiocytomas show at least focal lipidization, giant cells, and a storiform architecture and are therefore easily distinguished.

As myofibroblasts are considered to play an important part in wound healing it has been postulated that some form of trauma or injury may be responsible, at least in part, for their development[8]. Data presented by Lundgren et al.[9] support this theory as in their study some adult myofibromas stained for the endothelial antigen BNH9 and CD34. Indeed, they suggested that the entity might be a reactive vascular lesion with a primitive vascular progenitor cell being the cell of origin and differentiation into endothelial, pericytic and myofibroblastic cells can be observed in the lesions.

Provided that surgical excision of the lesions is complete, the prognosis of the solitary adult lesion appears to be excellent with few recurrences of the lesions.

In summary, we report two cases of solitary adult myofibroma which have distinct histological features, and immunohistochemical staining for smooth muscle actin and vimentin, compatible with previous reports in Korea[10-12].

REFERENCES

1. Stout AP: Juvenile fibromatosis. Cancer 1954;7: 953-978.
2. Chung EB, Enzinger FM: Infantile myofibromatosis. Cancer 1981;48:1807-1818.
3. Walsh RM, Leen EJ, Gleeson MJ: Solitary infantile and adult myofibromatosis of the nasal cavity: a report of two cases. Journal of Laryngology and Otology 1996;110:574-577.
4. Daimaru Y, Hashimoto H, Enjoji M: Myofibromatosis in adults (adult counterpart of infantile myofibromatosis). Am J Surg Pathol 1989;13:859-865.
5. Kozlowski K, Kan A, Marrsden FW: Case report

684. Infantile myofibromatosis. Skeletal Radiol 1991;20:397-399.
6. Beham A, Badve S, Suster S, Fletcher CDM: Solitary myofibroma in adults: clinical pathological analysis of a series. Histopathology 1993;22:335-341.
7. Lingen MW, Mostofi RS, Solt DB: Myofibromas of the oral cavity. Oral Surg Oral Med Oral Pathol Oral Radiol Endod 1995;80:297-302.
8. Darby I, Skalli O, Gabbiani G: Alpha-smooth muscle actin is transiently expressed by myofibroblasts during experimental wound healing. Lab Invest 1990;93:21-29.
9. Lundgren L, Kindblom LG, Meiskindblom JM: Adult myofibromatosis-reactive or neoplastic? Lab Invest 1995;72:A8.
10. Choi SW, Park JG, Kim TY, Kim HO, Kim CW, Kang SJ: A case of solitary myofibroma in adult. Korean J Dermatol 1997;35:1031-1035.
11. Lee JJ, Oh BM, Lee SJ, Kim DW, Chung SL, Kum YS: A case of solitary myofibroma in adult. Korean J Dermatol 2002;40:986-988.
12. Kim SJ, Park HS, Seo SL, Lee YS, Chon DK: A case of solitary adult type myofibroma in eyelid. Korean J Dermatol 2003;41:272-275.