

# Myxoid Myofibromatosis-type Perivascular Myoma Showing Prominent Verocay Body-like Formation

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Differential diagnoses showing Verocay body-like formation include schwannoma, palisading myofibroblastoma, palisading cutaneous fibrous histiocytoma, dermatofibroma with myofibroblastic differentiation, leiomyoma, palisaded encapsulated neuroma, and neuroma. A 60-year-old Korean man presented with a 10-year-history of an asymptomatic nodule on the right forearm. Histopathological examination revealed well-circumscribed multi-micronodules with prominent Verocay body-like formation consisted of spindle cells and extensive infiltrate of mucinous materials. The spindle cells were negative for S-100 protein and desmin, but positive for vimentin and  $\alpha$ -smooth muscle actin by immunohistochemical staining. The micronodules were lined by CD34 and factor VIII positive endothelial cells. The mucinous materials were stained with alcian blue at pH 2.5, but not at pH 0.5. We diagnose it as myxoid myofibromatosis-type perivascular myoma showing Verocay body-like formation.

(Ann Dermatol 12(4) 295~298, 2000).

*Key Words* : Verocay body-like, Perivascular myoma

Adult myofibromatosis was characterized by biphasic pattern that consisted of fascicles of spindle cells with abundant eosinophilic cytoplasm that resembled smooth muscle, in addition to a population of more primitive spindled cells associated with a hemangiopericytoma-like vascular pattern<sup>1</sup>. Recently, adult myofibromatosis, glomangiopericytoma, and myopericytoma were designated as perivascular myomas<sup>2</sup>. We report a case of myxoid myofibromatosis-type perivascular myoma with prominent Verocay body-like formation.

## CASE REPORT

A 60-year-old man presented with an asymptomatic rubbery firm subcutaneous 1 × 1 cm nodule on the right forearm that had slowly grown over 10 years (Fig. 1). He was otherwise healthy. The lesions excised and microscopic examination demonstrated a well-circumscribed multinodular lesion from upper dermis to subcutis (Fig. 2). The lesion consisted of numerous spindle cells. The features of palisaded nuclei surrounding micronodules were prominent (Fig. 3). Extensive infiltrate of bluish materials was also revealed. The spindle cells were positive for vimentin (Fig. 4A) and  $\alpha$ -smooth muscle actin (Fig. 4B), but negative for S-100 protein and desmin. The micronodules were lined by CD34 and factor VIII positive endothelial cells (Fig. 4C). The bluish materials were stained with alcian blue at pH 2.5 (Fig. 4D), but not at pH 0.5.

Received May 23, 2000.

Accepted for publication September 7, 2000.

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Fig. 1. Subcutaneous 1 × 1 cm nodule on the right forearm.

Fig. 2. Well-circumscribed multinodular lesion from upper dermis to subcutis (H & E, × 12.5).

## DISCUSSION

Infantile myofibromatosis was first described by Chung and Enzinger<sup>3</sup> in 1981. They also noted that infantile myofibromatosis occasionally occurs in adults. Adult counterpart of infantile myofibromatosis is characterized by solitary lesions mainly involving the dermis and showing benign biological behavior<sup>4</sup>. The lesions have been named cutaneous adult myofibroma or solitary myofibroma in adults<sup>5-7</sup>. The characteristic histology of infantile myofibromatosis is a distinctive biphasic pattern.

Fig. 3. Palisaded nuclei surrounding micronodules and spindle cells (H & E, × 200).

The peripheral areas of the tumor tend to be composed of bundles and sweeping fascicles of plump spindled cells with bland nuclei and abundant eosinophilic cytoplasm that bear a close resemblance to smooth muscle. The second component tends to be located centrally and is characterized by more cellular areas of primitive small, round to spindled cells with scant cytoplasm. The primitive cellular component is often associated with a hemangiopericytoma-like pattern of branching vessels<sup>2</sup>. The main histologic differences in adult myofibroma from infantile form have been described to be smaller and less conspicuous hemangiopericytoma-like pattern. The characteristic zonation pattern is less marked in the usual adult form of the disease. Frequent reversal of the pattern of zonation has been noted in adult forms<sup>2,6</sup>. In our case, micronodules are composed of centrally less cellular areas and peripherally more cellular components. The cells have cigar-like or wavy spindled nuclei without typical two cell patterns. Although there were no typical Verocay bodies, tumor cells were partly arranged in palisaded pattern with extensive infiltrate of mucin, which led to the tentative diagnosis of neural tumors, such as schwannoma. However, immunohistochemical stain for S-100 protein was negative. Prominent palisaded nuclei, so called Verocay body or Verocay body-like, have

**Fig. 4.** (A) The spindle cells were positive for vimentin ( $\times 200$ ), (B) the spindle cells were positive for  $\alpha$ -smooth muscle actin ( $\times 200$ ) (C) the micronodules were lined by CD34 positive endothelial cells ( $\times 200$ ) (D) the bluish materials were stained with alcian blue at pH 2.5 ( $\times 200$ ).

been described in palisading myofibroblastoma, palisading cutaneous fibrous histiocytoma, dermatofibroma with myofibroblastic differentiation, and leiomyoma, as well as neural tumors, such as schwannoma, palisaded encapsulated neuroma, neurofibroma, and neuroma<sup>8,9</sup>. Therefore, Verocay bodies may not be a clue to the diagnosis of neural tumors.

Requena et al<sup>7</sup> identified four patterns of adult myofibroma according to the stage of the lesions: vascular type in early stage, nodular or cellular type and multinodular or biphasic type in fully developed lesions, and leiomyoma-like or fascicular type in late stage. And they suggested that cutaneous adult myofibroma may be derived from myopericytes and proposed the term of myopericytoma. Recently, the concept of perivascular myoid tumors arising from a pluripotent periendothelial

cell capable of differentiating along smooth muscle, pericytic, and glomus cell line was introduced<sup>2</sup>. Because the histopathological features of adult myofibroma, hemangiopericytoma, glomus tumor, and myopericytoma show morphologic overlap, the term of perivascular myoma may be appropriate<sup>2,10</sup>. Our case may be included in myofibromatosis-type perivascular myoma.

Our case have two interesting findings. The one is the histopathological feature showing distinctive Verocay body-like pattern, which may lead to the diagnosis of neural tumors. Neural tumors have been often differentiated from adult myofibromas<sup>1,3,5-7,11</sup>. The other is the finding of extensive infiltrate of mucin.

We describe a unique case of myxoid myofibromatosis-type perivascular myoma showing prominent Verocay body-like formation.

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