A Case of Zosteriform Pilar Leiomyoma

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Cutaneous pilar leiomyomas are benign, often painful, smooth muscle tumors that usually present as red-brown nodules. They are usually multiple and commonly found on the extensor aspects of the arm, trunk, and face. Multiple lesions may be grouped or widespread. They are rarely distributed in a linear fashion or seem to follow a dermatome. We report a 54-year-old woman with multiple pilar cutaneous leiomyomas distributed in a zosteriform pattern on her right trunk. (Ann Dermatol 15(3) 128~131, 2003).

Key Words : Zosteriform, Leiomyoma

Cutaneous leiomyoma is a rare benign tumor that originates from smooth muscles derived from arrector pili muscle, media of blood vessels and dartos muscle of the scrotum, vulva or nipples[1]. It is classified into 3 types according to the site of origin, namely pilar leiomyoma, angioleiomyoma, and genital leiomyoma. Pilar leiomyomas usually occur in the second to third decades of life with an equal incidence in men and women. They can be solitary, but are multiple in 80% of patients[1-3]. Although multiple leiomyomas are often grouped, unilateral zosteriform pilar leiomyoma is rare. We report multiple zosteriform leiomyomas on right trunk of a 54-year-old woman.

CASE REPORT

A 54-year-old woman presented with reddish skin-colored papules. She had noted these lesions 7 years ago, and they gradually extended to right flank and chest. She complained of episodes of tolerable pain in the involved region in response to touch. She had had a hysterectomy for uterine leiomyoma about 20 years ago. On physical examination, there were multiple dark red to skin-colored tender dermal nodules distributed in a zosteriform pattern on right chest, flank, and back (Fig.1). Histological examination of a lesion showed a well-demarcated, non-encapsulated tumor within the dermis (Fig.2A), which is composed of interlacing spindle-shaped cells with a blunt-ended nucleus (Fig.2B). The bundles of tumor showed vacuolization, especially in cross sections, as a result of a perinuclear clear zone (Fig.2C). Masson's trichrome stain showed that these tumor cells stained red and collagen blue (Fig.3). These findings confirmed the diagnosis of leiomyoma. After the diagnosis was made, no specific treatment was done because she did not want any treatment.

DISCUSSION

Pilar leiomyomas are firm, painful, and intradermal nodules, most commonly seen in early adult life with an equal incidence in male and female[2]. They can occur in sites where smooth muscle exists, but are most commonly found on the extensor aspects of the limbs[4]. They range in size from a few millimeters to several centimeters and the color varies from pink to yellow or brown[2,3]. The lesions are often sensitive to touch, cold and emotional stress. Pain usually occurs spontaneously and may be paroxysmal in nature with stabbing, burning or

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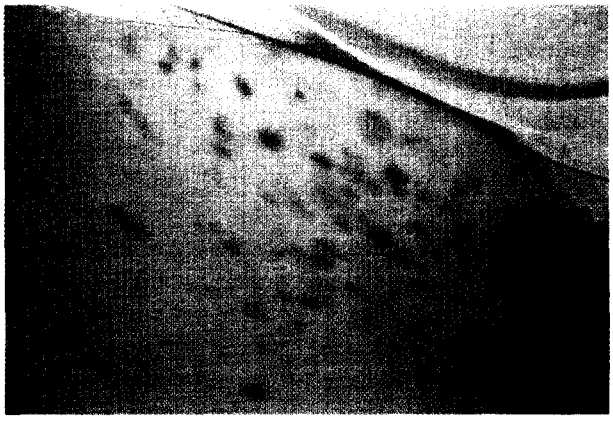

Fig. 1. Multiple red-brown nodules in a zosteriform pattern on right trunk

Fig. 2. (A) The tumor is well-demarcated and composed of bundles of spindle-shaped cells (H&E, ×40). (B) The spindle-shaped cells contain bluntly edged nuclei (H&E, ×200). (C) The spindle-shaped cells show vacuolization in cross section, as a result of a perinuclear clear zone (H&E, ×200).

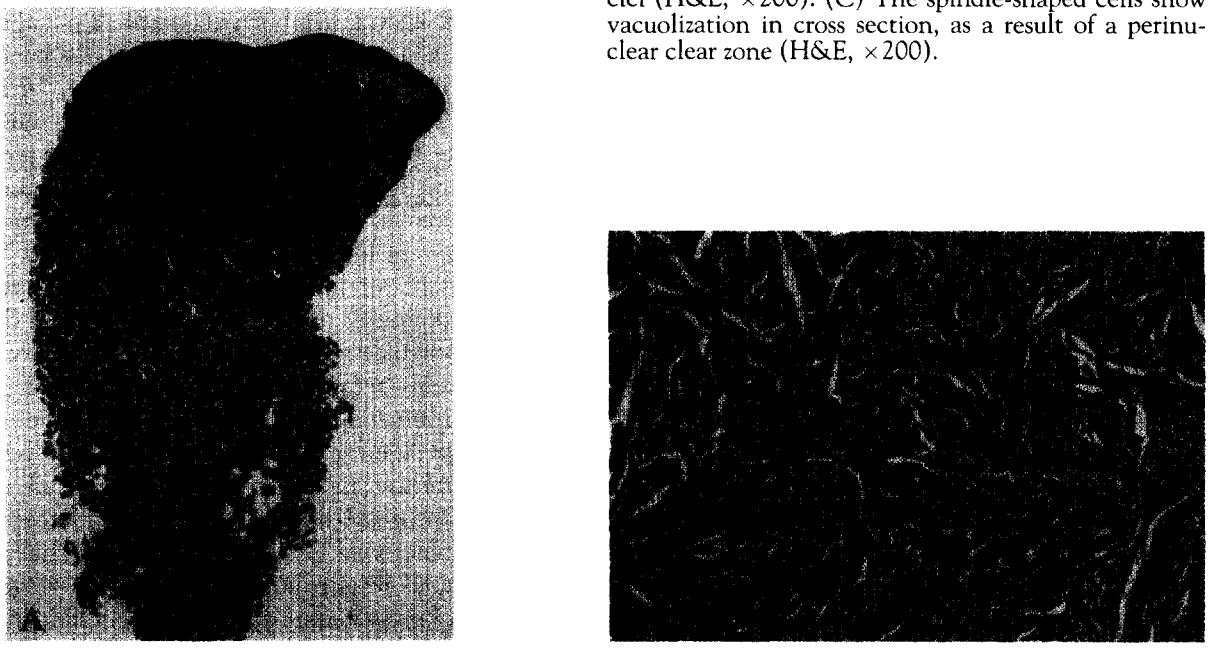

Fig. 3. (A) Masson's trichrome stain differentiates between smooth muscle fibers and collagen bundles (Masson's trichrome stain, ×40). (B) The red colored smooth muscle fibers are intermingled with small amount of green colored collagen bundles in the dermis (Masson's trichrome stain, ×100).

pinching qualities. The cause of pain has been debated. Some authors postulated that it may be due to pressure on the cutaneous nerve[5]. Some described increased numbers of nerve fibers in these tumors and believed compression of them may be the cause of pain[6-8]. In our case, however, we don't find any evidence of increased numbers of nerve fibers. Some believed that muscular spasm is the cause of pain and demonstrated visible contraction of the lesions in some cases[2].

Pilar leiomyomas can be solitary, but they are usually multiple[3]. In cases of multiple pilar leiomyomas, unilateral zosteriform pilar leiomyoma is rare. The zosteriform pattern of pilar leiomyoma is difficult to explain. It may be due to an association with the cutaneous nerves underneath, as suggested earlier[5]. Some authors proposed spontaneous somatic recombination giving rise to a pattern mimicking zosteriform distribution[4].

Several conditions are associated with multiple

pilar leiomyomas. Uterine leiomyomas can be associated with pilar leiomyomas, known as Reed's syndrome[3,9,10]. The inheritance pattern is autosomal dominant with incomplete penetrance[10]. The typical family history for Reed's syndrome is several women with early hysterectomies. Women in these families may have multiple cutaneous leiomyomas. Approximately 50% have uterine leiomyomas, compared with about a 10% incidence in the general population of women in the same age group[2,9]. Women with multiple cutaneous leiomyomas need periodic gynecologic evaluation[7,9,10]. If uterine leiomyomas are present, the patient should be informed that a hysterectomy may be necessary at some point[7,10]. For some women, this information can help with family planning[11]. Leiomyosarcomas have been reported to arise within uterine leiomyoma, necessitating long-term observation[12]. Female family members also need gynecologic evaluation[9]. The zosteriform pattern, with a few discrete nodules, has been described in hereditary leiomyoma and in a patient with uterine leiomyoma. The involvement of internal organs, especially uterus, even with zosteriform leiomyoma was reported[11]. In that case, the patient's mother and a first cousin had had a hysterectomy for problems associated with uterine "fibrinoid". Our patient had multiple cutaneous leiomyomas in zosteriform pattern with a history of hysterectomy for uterine leiomyoma about 20 years ago, but she had no family history for early hysterectomy or uterine leiomyoma.

Besides familial occurrence of multiple cutaneous leiomyomas, the association of multiple cutaneous leiomyomas with visceral leiomyomatosis are well established. Visceral leiomyomas developing in the setting of immunodeficiencies (e.g. AIDS and organ transplantation) are associated with EBV (latent) infection[7].

Erythrocytosis has been reported in association with cutaneous leiomyoma in two patients[3,9,13]. Both patients had widespread leiomyomas. In one, erythrocytosis lessened after surgical excision of many of the lesions[13]. Erythropoietin-like activity has been demonstrated in tumor extracts[13,14]. In our patient, laboratory evaluations ,including complete blood count, were within normal limits or negative.

Histologically, cutaneous leiomyoma is an ill-defined dermal tumor composed of interlacing smooth muscle fibers with collagen bundles. The epidermis is typically uninvolved. The nuclei have a distinctive blunt ended shape[15]. Some authors demonstrated mitotic activity in cutaneous leiomyomas, but for the most part, mitotic activity was less than one per 10 HPF[8,16]. Smooth muscle cells can be visualized in Masson's trichrome, actin and desmin stain[15].

Treatment of cutaneous leiomyoma is difficult, and surgical excision is practicable only when lesions are few. Recurrences after excision are frequent. But, some author proved that mitotic activity does not adversely affect the prognosis[8]. Surgical treatment of multiple cutaneous leiomyomas is not feasible because of the large number of tumors. In these cases, pharmacologic treatment can offer pain relief. Successful use of several different oral and topical agents has been reported. Oral agents include α-blockers, calcium channel blockers, nitroglycerin, antidepressants, ethaverine, and analgesics[4,11,17,18]. Treatment with smooth muscle relaxants is effective in pain, and this represents that the cause of pain associated with cutaneous leiomyoma is more likely secondary to contraction of the muscle fibers making up the tumor[2]. Topical treatments include nitroglycerine paste, lidocaine, phentolamine, and hyoscine hydrobromide[16]. Electrocoagulation, carbon dioxide laser, cryotherapy, and radiotherapy are generally ineffective[19]. Gonadotropin-releasing hormone analogue therapy has been used successfully to treat uterine and some extrauterine smooth muscle tumors which demonstrate progesterone and/or estrogen receptor immunoreactivity[16]. But, it was reported that none of cutaneous leiomyomas demonstrated positive staining of progesterone and estrogen[20]. Our patient had minimal intermittent symptoms in response to touch and no intervention was offered.In Korea, 12 cases of multiple cutaneous leiomyoma have been reported. However, there have been no reported case of zosteriform distribution to our knowledge1[8,21-24]. In one case, there is room for doubt of unilateral zosteriform distribution, but the report contained no mention of this point. On the other hand, the patient of that case was female and had history of hysterectomy[22].

In summary, we report a patient with history of hysterectomy for uterine leiomyoma and unilateral zosteriform distribution of multiple pilar leiomyomas.

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