Clinicopathological Study of Primary Superficial Leiomyosarcomas[†]

Chairat Burusapat MD*, Kantang Satayasoontorn MD**, Scott D Nelson MD***

[†] Poster presented at the International Surgical Week 2011, Yokohama, Japan, August 28 through September 1, 2011
 * Division of Plastic and Reconstructive Surgery, Department of Surgery, Phramongkutklao Hospital, Bangkok, Thailand
 ** Department of Anatomical Pathology, Army Institute of Pathology, Bangkok, Thailand
 *** Department of Pathology, David Geffen School of Medicine at UCLA, Los Angeles, California, US

Background: Primary superficial leiomyosarcomas (PSL) are rare malignant lesions that are subdivided into cutaneous and subcutaneous tumors. Primary cutaneous and subcutaneous leiomyosarcomas differ not only as to primary site of origins, but also to differences in prognosis. Guidelines for management and follow-up are not clearly defined in the literature. **Material and Method:** Retrospective review was conducted from the patient's chart between January 2000 and December 2009. Histopathology, immunohistochemistry, and clinical and surgical records were reviewed.

Results: The authors found five cases of PSL and divided them into two cases of cutaneous leiomyosarcomas and three cases of subcutaneous leiomyosarcomas. Overall, mean age of the patients was 42.4 years, male: female ratio was 4:1. Clinical presentations were painless mass. Wide excisions were performed in three cases with 2 cm margins. No local recurrence was found in the period of follow-up (6 months to 3 years). One case presented with bony metastasis five years after operation. **Conclusion:** PSL are rare tumors. Surgical resection remains the main option for curative treatment. Wide excision with at least 2 cm peripheral margins and a depth that includes subcutaneous tissue and fascia are recommended. The natural history of these tumors is not clearly defined. All patients should be followed-up for a period of at least five years after treatments. The authors hoped that further study of these tumors would result in better treatments and follow-up guidelines to be a benefit to such patients in the future.

Keywords: Primary superficial leiomyosarcomas, Clinicopathology

J Med Assoc Thai 2013; 96 (3): 294-301 Full text. e-Journal: http://jmat.mat.or.th

Leiomyosarcomas are aggressive soft tissue sarcomas. Primary superficial leiomyosarcomas (PSL) are rare malignant lesions and constitute 2 to 3% of all leiomyosarcomas⁽¹⁾. PSL are divided into cutaneous and subcutaneous tumors by their primary site of origins.

Primary cutaneous leiomyosarcomas are derived from the erector muscles associated with the hair and sweat glands in the dermis. Primary subcutaneous leiomyosarcomas are derived from the muscular coats of blood vessels.

Primary cutaneous and subcutaneous leiomyosarcomas differ not only as to primary site of origins, but also to differences in prognosis. However, most of the current literatures usually combine the cutaneous and subcutaneous leiomyosarcomas in their

Correspondence to:

E-mail: pataranat@hotmail.com

reports. Guidelines for management are not clearly defined in the literature. Cutaneous leiomyosarcomas had high rates of local recurrence ranging from 14% to 50%. Subcutaneous leiomyosarcomas had a higher recurrence rate than cutaneous leiomyosarcomas, ranging from 50% to 70% and distant metastases have been known to occur⁽¹⁻⁵⁾.

In the authors review of cases between January 2000 and December 2009, the authors found five cases of PSL. The authors found two cases of cutaneous leiomyosarcomas and three cases of subcutaneous leiomyosarcomas in all clinicopathologic presentations.

Material and Method

This was a retrospective review of patients in our hospital with primary cutaneous and subcutaneous leiomyosarcomas. Between January 2000 and December 2009, five patients were diagnosed with PSL. Histopathology and immunohistochemistry were reviewed in all patients.

Burusapat C, Division of Plastic and Reconstructive Surgery, Department of Surgery, Phramongkutklao Hospital, Bangkok 10400, Thailand. Phone: 081-371-3392

Clinical and surgical records were reviewed for age at diagnosis, sex, presenting symptoms, location, preoperative imaging findings, recurrence, metastasis, and treatment.

Results

In the 10 years period of review, the authors found five cases of PSL and divided them into two cases of cutaneous leiomyosarcomas and three cases of subcutaneous leiomyosarcomas.

Overall, mean age of the patients was 42.4 years (2-84 years). Patients with cutaneous leiomyosarcomas ranged in age from 2 to 39 years (mean 20.5 years) and patients with subcutaneous leiomyosarcoma ranged in age from 35 to 84 years (mean 57 years). Four of five patients were male. However, all three of subcutaneous leiomyosarcomas were male (Table 1, 2). Clinical presentation in four cases was that of a painless nodular mass, with the remaining case presented additionally with intermittent bleeding. There were incorrect clinical diagnoses in three cases (clinical suspicious of inclusion cyst in case 1, hemangioma in case 2 and lipoma in case 4). Wide excision with a 2 cm margin was performed in three cases, and local excision was performed in two cases. There was no local recurrence in the period of follow-up (6 months-3 years). One case with close

surgical margins was lost of follow-up and presented with bony metastasis five years after the excision (case 1). Two cases of subcutaneous leiomyosarcomas with close surgical margins received post operative radiotherapy. One case of cutaneous leiomyosarcoma received post-operative radiotherapy at the metastatic site (spine).

Cases report

Case 1

A 39-year-old Thai male presented with one-year of a slow growing nodular mass that involved his right cheek. Physical examination showed 2 cm smooth edge, cystic mass at the right cheek. Excisional biopsy showed a cutaneous leiomyosarcoma with close excision margins to tumor (Fig. 1). The patient was lost of follow-up after the excision. Five years later, the patient presented with back pain and paraplegia of the lower extremities. CT myelography revealed spinal cord compression at T11. Bone biopsy of spine showed metastatic leiomyosarcoma (Fig. 2). Decompress laminectomy of T10-T12 with partial posterior vertebrectomy of T11 with debulking of tumors was performed. Post-operative radiotherapy at T-L spine was given. The patient refused adjuvant chemotherapy and was again lost in follow-up after completion of radiotherapy.

	Case No. 1	Case No. 2	Case No. 3	Case No. 4	Case No. 5
Age/sex	39/M	2/F	52/M	35/M	84/M
Clinical	Painless cystic mass 1 year	Intermittent bleeding on painless cystic mass 2 years	Painless nodular mass 6 months	Painless nodular mass 2 months	Painless nodular mass 1 year
Location	Right cheek	Right back	Left thigh	Left hip	Left neck
Size	2 cm	5 cm	7 cm	5 cm	3 cm
Treatment	Excision	Excision	Wide excision 2 cm margins	 Excision Wide excision cm margins with superficial groin lymphadenectomy 	Wide excision 2 cm margins
Adjuvant	No	No	Radiotherapy	No	Radiotherapy
Follow-up	Lost follow-up	1 year	6 months	2 years	3 years
Predisposing factor	No	No	No	No	Previous radiation 15 years
Recurrence	No	No	No	No	No
Metastasis	Bone metastasis 5 years after surgery	No	No	No	No

Table 1. Clinical data, treatment and follow-up

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	Case No. 1	Case No. 2	Case No. 3	Case No. 4	Case No. 5
Growth pattern	Nodular	Nodular	Nodular	Nodular	Nodular
Cellularity	High	High	High	High	High
Pleomorphism	Mild	Mild	Moderate	Moderate	Marked
Giant cells	+	-	+	++	+++
Mitosis/10 HPF	18	2	6	12	15
Necrosis	20%	-	-	10%	10%
Ulceration	-	-	-	-	-
Sbct extension	+	+	Sbct LMS	Sbct LMS	Sbct LMS
FNCLCC Grd	2	1	2	2	3
IHC: SMA	+	+	+	+	+
IHC: Desmin	N/A	-	N/A	+	+
IHC: MSA (HHF35)	N/A	+	N/A	N/A	N/A
IHC: Vimentin	+	N/A	+	N/A	N/A

Table 2. Histopathological and Immunohistochemistry of primary superficial leiomyosarcoma

Sbct = subcutaneous; FNCLCC = Fédération Nationale des Centres de Lutte Contre le Cancer (The National Federation of French Cancer Centres); HPF = high-power-fields; IHC = immunohistochemical study; SMA = smooth muscle actin; MSA = muscle specific actin; HHF35 = muscle actin antibody type HHF35

Case 2

A 2-year-old Thai girl presented initially with a small cystic mass on her back since two months after birth. The mass grew slowly and was painless but with some itching and intermittent contact bleeding. The initial diagnostic impression was hemangioma and conservative treatment was recommended to her parents. Subsequently, new small nodular lesions arose adjacent to the first mass. Physical examination showed 5 cm. smooth, cystic mass and a 2 cm nodular mass on her back. Ultrasonography showed a large mixed solid and cystic mass in the subcutaneous tissue of the lower back, with mass effect on the underlying muscle and hypervascularity on color Doppler examination. Excision was performed and pathological examination



Fig. 1 Cutaneous leiomyosarcoma, case 1.
(A) Low-power view (original magnification x40) shows an intradermal tumor composed of interlacing bundles of elongated spindle-shaped cells.
(B) Low power view (original magnification x40). The tumor cells express smooth muscle actin.

showed primary cutaneous leiomyosarcoma with all surgical margins was free from tumor (Fig. 3). Patient has was followed clinically every three months subsequent to excision. The one-year follow-up showed no recurrence or metastasis of the tumor.

Case 3

A 52-year-old Thai male presented with six months of large slowly growth, painless nodular



Fig. 2 Metastasis of cutaneous leiomyosarcoma to spine. Section shows trabecular bone of vertebral body. The spaces in between are permeated by intersecting, sharply marginated groups of spindle cells. These spindle cells show immunoreactivity with SMA.

mass on his left leg. He had other medical problems including Diabetes Mellitus, hypertension, and dyslipidemia but they were under control by medicine. Physical examination showed 7 cm round, rubbery, nodular mass on left medial thigh. MRI showed a 3x3.5x7 cm mass at the left upper thigh without underlying muscular invasion. Core needle biopsy revealed malignant spindle cell neoplasm with positive for SMA and MSA and negative for S-100, CD34, and desmin. Wide excision with 2 cm margins around the tumor was performed and the fascia of underlying muscle was removed. Pathological report showed subcutaneous leiomyosarcoma with moderate pleomorphism (Fig. 4). Although, all surgical margins were free from tumor, post-operative radiotherapy also was given. The patient was followed at three months and six months and there was no recurrence or metastasis of tumors.

Case 4

A 35-year-old Thai male presented with a history of a large, rapidly growing, nodular soft tissue mass on the left hip for two months. Excisional biopsy was performed for a diagnosis of lipoma but pathological examination revealed a subcutaneous leiomyosarcoma, pleomorphic type, with all margins involved with tumor. The tumor stained positively for SMA and desmin and negatively for S-100. The patient was referred to the surgical Department. Physical examination revealed a 5 cm long surgical scar on left hip and 1 cm lymph node in the left groin. Abdominal CT revealed matted lymph nodes in the left groin. Wide excision with 2 cm margins around tumor and left inguinal lymphadenectomy were performed. Pathological examination revealed a subcutaneous leiomyosarcoma, pleomorphic type (Fig. 5). All of twelve inguinal lymph nodes showed lymphoid hyperplasia. No adjuvant chemoradiotherapy was given. The patient was examined clinically every three months and after two years of follow-up, there was no recurrence or metastasis of tumor.

Case 5

An 84-year-old Thai male presented with a slowly growing, painless, nodular mass on the left neck for one year. Physical examination showed a 3 cm round, firm nodular mass on left neck. CT neck showed an inhomogeneous enhanced well-defined mass occupied left levator scapula muscle from level C2-C4 diameter 3 cm. The patient had a history of lymphoepithelial carcinoma of the left buccal



Fig. 3 Cutaneous leiomyosarcoma, case 2.
(A) Low-power view (original magnification x40) shows the tumor located in dermal layer.
(B) High-power view (x400). The tumor cells shows immunoreactivity with muscle-specific actin (HHF35).



Fig. 4 Subcutaneous leiomyosarcoma, case 3.
(A) Low-power (original magnification x40) view shows the tumor located in subcutaneous tissue.
(B) Subcutaneous leiomyosarcoma (x200). case 3, Tumor shows intersecting fascicles of atypical elongated cells with abundant eosinophilic cytoplasm.





mucosa 15 years ago, and underwent wide excision with post-operative radiotherapy at the left buccal and left neck regions. Head and neck examination showed no abnormality of the oral cavity and pharynx. Fine needle aspiration of the mass was non-diagnostic. Core needle biopsy revealed a pleomorphic leiomyosarcoma. A bone scan showed no evidence of metastasis. Wide excision with 2 cm margins was performed.



Fig. 6 Subcutaneous leiomyosarcoma, case 5.
(A) Subcutaneous leiomyosarcoma (x200) shows desmin staining.
(B) Subcutaneous leiomyosarcoma (x200), smooth muscle actin (SMA) staining.

Pathological examination revealed a subcutaneous, pleomorophic leiomyosarcoma, and with close surgical margins focally (Fig. 6). Post-operative radiotherapy was performed.

The patient was followed clinically with examinations every three months for one year, and then every six months. Three years follow-up showed no recurrence or metastasis of tumors.

Histopathology (Table 2)

Both cases of cutaneous leiomyosarcoma showed a neoplastic spindle cell proliferation predominantly occupying the reticular dermis. The interface between the tumors and reticular dermis was infiltrative and permeative. Extension into deeper subcutaneous tissue was evident in both cases. The diagnosis of leiomyosarcoma was established based on cellularity, increased mitotic activity, and focal areas of necrosis (case 1 only) and confirmed by immunohistochemical studies. Both cases were highly cellular and locally infiltrative neoplasms composed of interlacing bundles of spindle-shaped cells with eosinophilic cytoplasm and blunt-ended, cigar-shaped, often vesicular nuclei. Pleomorphism was mild in both cases. Case 1 contained a few scattered multinucleated giant cells, which were not present in case 2. A high mitotic rate (18/10 HPF) and several foci of necrosis (20% of total area) were present in case 1.

The mitotic rate of case 2 was 2/10 HPF, which was low but met the criteria for a diagnosis of cutaneous leiomyosarcoma. There was no necrosis present in case 2. Immunohistochemical studies confirmed the diagnosis of leiomyosarcoma in both cases. Tumor cells in both cases showed diffuse strong immunoreactivity for smooth muscle actin (SMA). In addition, tumor cells in case 2 also showed positivity for muscle specific actin (MSA, HHF-35).

Histology of the three cases of subcutaneous leiomyosarcoma displayed irregular lesions located within subcutaneous tissue, infiltrating into adjacent striated muscle bundles. Two out of three cases were composed of perpendicularly arranged fascicles of moderately pleomorphic spindle cells with eosinophilic fibrillary cytoplasm. Scattered multinucleated giant cells were also present in both cases. The remaining case of subcutaneous leiomyosarcoma consisted of markedly hyperchromatic and pleomorphic cells admixed with numerous bizarre multinucleated giant cells in background of chronic inflammatory cell infiltration. Areas of typical interlacing bundles of elongated spindle-shaped cells with eosinophilic cytoplasm and blunt-ended nuclei were also present. All three of the subcutaneous leiomyosarcoma cases contained increased mitosis with rates of 6 to 15 per 10 high-power-fields. Necrotic areas were present in case 4 and 5 but not in case 3. Upon immunohistochemical staining, all of these subcutaneous tumors expressed diffuse smooth muscle actin (alpha-SMA), while a small number of tumor cells of case 4 and case 5 were also focally positive for desmin. The combined histopathologic appearance and immunohistochemical profile of these tumors was compatible with subcutaneous leiomyosarcoma.

Discussion

PSL are rare malignant tumors and can be subdivided into cutaneous leiomyosarcoma and subcutaneous leiomyosarcoma, based on the primary site of origin. Most reports had usually combined cutaneous and subcutaneous leiomyosarcomas together, although there were histopathologic and prognostic differences⁽⁶⁾. PSL have been reported in patients aged between 10 and 93 years of age, with an increase in incidence after middle age, and with the peak occurrence being between 50 and 70 years of ages^(2,3,7). One of our patients was 2-years-old, which is the youngest patient that has ever been reported. In the presented cases, the age of occurrence of cutaneous leiomyosarcoma was younger than that of subcutaneous leiomyosarcoma, with a mean age 20.5 years, compared with 57 years for subcutaneous leiomyosarcoma.

There were differences in sex ratio of PSL in previous reports. Auroy S et al reported that liomyosarcoma could occur at any age without predominant sex-ratio⁽⁸⁾. Kaddu et al showed a slight female preponderance, but reports on larger patient populations by Dahl et al and Fields et al showed male predominance with the ratio was $2-3:1^{(2,3,7)}$. This corresponds with the authors'findings of a male predominance with a ratio of 4:1. However, all of our cases of subcutaneous leiomyosarcoma occurred in males. Fields and Helwig found that the tumors occurred most frequently in the extremities and showed a predilection for the hair-bearing extensor surface regions associated with greater density of both hair follicles and erector pili muscles⁽³⁾. Lin et al reported that leiomyosarcoma of the face are very rare⁽⁴⁾. However, there are many reports of these tumors in head and neck regions^(1,3,7,9-12) as well as other unusual locations including the scrotum⁽¹⁾. In the present study, the authors contend that both cutaneous and subcutaneous leiomyosarcoma can occur anywhere in the body.

PSL usually occurs as a solitary nodule ranging in size from 0.4 to 6 cm. The lesions may be smooth, ulcerated, or hemorrhagic⁽¹⁰⁾. Limaiem F et al reported all four of their patients presented with a painless nodule, with ulceration present in half of them and with one case of erythematous skin nodule⁽¹⁾. Wargon reported cases of cutaneous leiomyosarcoma and showed that the suspected clinical diagnosis did not correlate with the final actual correct diagnosis⁽¹³⁾.

Most of the presented patients complained of a slowly growing, painless nodule (4 of 5) or were asymptomatic. One patient presented with intermittent contact bleeding and incorrect clinical diagnoses was hemangioma. The clinical appearance of PSL was non-specific led to misdiagnoses including squamous cell carcinoma, basal cell carcinoma, malignant melanoma, and other benign lesions including inclusion cyst, hemangioma, and lipoma. PSL cannot be diagnosed clinically because their gross appearance are non-specific⁽¹⁰⁾.

The diagnosis of PSL is based on cellularity, mitotic activity, focal areas of necrosis and is confirmed by immunohistochemical studies. In small biopsies, PSL may be misinterpreted as a benign smooth muscle proliferation such as leiomyoma⁽¹⁾. Predisposing factors for PSL are inconclusive. While one of our patients with subcutaneous leiomyosarcoma had received radiotherapy 15 years previously, there was no evidence of an association between previous radiotherapy and PSL, especially cutaneous leiomyosarcoma. Guidelines for management and follow-up are not clearly defined. Surgical resection remains the main option for curative treatment. Controversy remains as to what qualifies as an adequate surgical margin. Some reports recommended wide excision with 3 to 5 cm lateral margins and a depth that includes subcutaneous tissue and fascia^(1,4,14). Other reports, as with the present cases, proposed that a 2 cm margins resulted in no recurrence or metastasis in the follow-up period^(5,15). Limaiem F et al reported recurrence of cutaneous leiomyosarcoma 12 months after surgery in a patient whose tumor demonstrated high mitotic rate (17/10HPF) and extension into subcutaneous tissue⁽¹⁾.

Cutaneous leiomyosarcoma with high rates of local recurrence ranging from 14% to 50% have been reported. Subcutaneous leiomyosarcoma has a higher recurrence rate than cutaneous leiomyosarcoma, ranging from 50% to 70%⁽¹⁻⁵⁾. Distant metastases via hematogenous spreading have been known to occur in subcutaneous leiomyosarcomas and this may be from the origin of tumor derived from muscular coats of blood vessels. This shows that while recommendations for treatment of cutaneous and subcutaneous leiomyosarcomas are usually the same, accurately classifying and discriminating between these two lesions has prognostic significance.

Guidelines for surgical treatment of PSL have been given, grouping cutaneous and subcutaneous leiomyosarcomas together despite recognizing them as entities with different prognoses^(2,7,11). It has been claimed that the rate of recurrence is decreased with wider excision, but there are no data substantiating these claims^(3,11). The reports of high local recurrence rates may represent high rates of incomplete surgical excision. Although, metastases have been rarely described in patients with cutaneous leiomyosarcoma^(2,6,7,16), the authors had one patient with metastasis to spine five years after excision. Massi D et al reported one patient with dermal leiomyosarcoma with minimal extension into fat who developed a distant metastasis 15 years after diagnosis⁽¹⁷⁾. None of our patients had recurrence in the period of follow-up. None of our patients have shown lymph node metastasis. Oliver et al proposed that the risk of local recurrence was related to the adequacy of excision, whereas metastatic spread was related to depth, histological grade of the tumor, and the DNA content⁽¹⁸⁾. This correlates with our findings in that in our patients metastasis occurred in a cutaneous leiomyosarcoma with a high mitotic rate (18/10 HPF) and several areas of necrosis (20% of total neoplastic area). Auroy et al reported a study of 32 patients with primary cutaneous leiomyosarcomas and showed that five patients with leiomyosarcomas involving the subcutis developed local recurrences,

and two of them died of the disease⁽⁸⁾. They suggested that the main prognostic factors were tumor size, distal location, depth of tumor invasion and pathological grade. Jensen et al found that poor prognostic factors were tumor size more than 5 cm, deep location with fascia involvement, and high-grade tumor⁽¹⁹⁾. Dahl et al showed that metastases were seen particularly in patients with subcutaneous and multiple leiomyosarcomas⁽²⁾. Their study suggested that size and mitotic activity of tumor appear to have some prognostic value. Fourteen in 47 patients died with metastases, especially in the lungs. The authors recommend that main prognostic factors are type of tumor (cutaneous or subcutaneous leiomyosarcomas), tumor size, depth of tumor invasion, and tumor grade (mitotic rate and area of necrosis). Other treatments attempted include radiotherapy^(2,20) and Mohs micrographic surgery^(14,21). Humphreys reported the overall cure rate of leiomyosarcoma treated by Mohs micrographic surgery was 87%. Fish et al suggested that Mohs micrographic surgery was proven to be very useful in treating these difficult neoplasms⁽¹⁴⁾. Lin et al suggested that wide excision and postoperative radiotherapy be performed in cases with deep tumor invasion, high-grade tumor, and large tumor size⁽⁴⁾. However, the role of adjuvant therapy and Mohs micrographic surgery in PSL remains controversial and no sufficient statistical evidence exists regarding its efficacy. The authors recommend at least five years follow-up after treatment in all patients.

Conclusion

PSL are rare tumors. Cutaneous and subcutaneous leiomyosarcoma differ microscopically and in primary site of origins. They appear to have differences in regards to the clinicopathologic setting and in prognosis. Guidelines for treatment and follow-up are not clearly defined. Surgical resection remains the main option for curative treatment. Wide excision with at least 2 cm peripheral margins and a depth that includes subcutaneous tissue and fascia are recommended. Recurrence and metastasis may occur many years after resection and may result in a fatal outcome. One of our patients was 2-year-old female, which is the youngest patient that has ever been reported, and has very good prognosis even though delay in diagnosis compared to other age groups. The natural history of these tumors is not clearly defined. Both cutaneous and subcutaneous leiomyosarcoma may behave aggressively. All patients should be followed-up for a period of at least five years after

treatment. The authors hope that further study of these tumors will result in better treatments and follow-up guidelines to be benefit of such patients in the future.

Potential conflicts of interest None.

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การศึกษาทางพยาธิวิทยาและคลินิกของมะเร็งกล้ามเนื้อเรียบที่มีต้นกำเนิดในชั้นเนื้อเยื่อผิวหนัง

ชัยรัตน์ บุรุษพัฒน์, กัลตังต์ สัตตยสุนทร, Scott D Nelson

ภูมิหลัง: มะเร็งกล้ามเนื้อเรียบที่มีค้นกำเนิดในชั้นเนื้อเยื่อผิวหนังเป็นมะเร็งที่พบได้ค่อนข้างน้อย สามารถแบ่งได้เป็น cutaneous และ subcutaneous tumors โดยใช้จุดกำเนิดของเนื้องอกเป็นหลัก primary cutaneous และ subcutaneous leiomyosarcomas มีความแตกต่างทั้งในแง่ primary site of origins รวมถึง prognosis ด้วย นอกจากนี้พบว่า ยังไม่มี แนวทางในการรักษาและติดตามผลที่ชัดเจน

วัสดุและวิธีการ: เป็นการศึกษาย้อนหลังระหว่างเดือนมกราคม พ.ศ. 2544 ถึง ธันวาคม พ.ศ. 2553 โดยมีการรวบรวม histopathology และมีการย้อมทาง immunohistochemistry ของเนื้องอกรวมถึงอาการทางคลินิก และการผ่าตัด

ผลการสึกษา: พบว่ามีผู้ป่วยทั้งหมด 5 ราย โดยแบ่งเป็น cutaneous leiomyosarcomas 2 ราย และ subcutaneous leiomyosarcomas 3 ราย อายุเฉลี่ย 42.4 ปี อัตราส่วน เพศชาย:หญิง เท่ากับ 4:1 อาการทางคลินิกมักมาด้วยก้อนที่ไม่มีอาการ ผู้ป่วย 3 ราย ได้รับการผ่าตัด wide excision โดยห่างจากขอบเนื้องอก 2 ซม. และไม่พบการกลับเป็นซ้ำในช่วงเวลาที่ติดตามผล (6 เดือน-3 ปี) ผู้ป่วย 1 ราย มีการกระจายไปที่กระดูก 5 ปี หลังการผ่าตัด

สรุป: มะเร็งกล้ามเนื้อเรียบที่มีต้นกำเนิดในชั้นเนื้อเยื่อผิวหนังเป็นมะเร็งที่พบได้ค่อนข้างน้อย การผ่าตัดยังคงเป็นการรักษาหลัก โดยต้องผ่าตัดอย่างน้อย 2 ซม. ห่างจากขอบเนื้องอก ธรรมชาติของมะเร็งชนิดนี้ยังไม่ชัดเจน ผู้ป่วยควรได้รับการติดตามอย่างน้อย 5 ปี ภายหลังการรักษา ดังนั้นผู้นิพนธ์หวังว่าการศึกษามะเร็งชนิดนี้ต่อไปในอนาคตจะทำให้ผลการรักษาดีขึ้นและเกิดแนวทาง ในการรักษาที่เป็นประโยชน์ต่อผู้ป่วยประเภทนี้ต่อไป