



피부 평활 근육종의 표재성 장골 회선 천공지 유리 피판술을 이용한 재건 증례

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Treatment of Cutaneous Leiomyosarcoma on the Cheek with Superficial Circumflex Iliac Artery Perforator Fasciocutaneous Free Flap: A Case Report

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Cutaneous leiomyosarcoma (CLM) is a rare soft tissue sarcoma derived from the dermis or subcutaneous portion of the skin. Recently, a young male patient presented with a very rare case of CLM. This patient was 33 years old and a smoker with no specific medical or trauma history. During the clinical examination, a 1.6×0.8×1.7 cm sized, oval shape, hard, fixed, and non-tender mass on the right cheek was noted. He underwent a simple excision at first and pathology reported CLM of grade 1. We performed a muscle depth wide excision again with a 1 cm margin for the superior medial side and a 2 cm margin for the inferior lateral side. Lesion coverage was ensured by a superficial circumflex iliac artery perforator fasciocutaneous free flap after confirming the free margin sections.

Key Words: Leiomyosarcoma, Micro surgical free flaps

Cutaneous leiomyosarcoma (CLM) represents about 3% of the superficial soft tissue sarcomas¹. It can develop almost in any part of the body sometimes arising from a previous scar². Most cases present as a slow growing solitary nodule with a reddish coloration³. Lesions are most commonly found on the lower extremities and mistreatment is relatively common due to the rarity of this type

of tumor. Tumor location and its extension to adjacent structures can limit the range of excision and determine the prognosis of patient. In this report, we present a rare case of CLM on the cheek of a young male patient and share our treatment experience.

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CASE REPORT

A 33-year-old male presented to Wonju Severance Christian's Hospital's dermatology department for a painless 1.6×0.8×1.7 cm solitary nodule on the right perinasal area, which increased in size during one month according to the patient (September 5th, 2018). He had no significant medical or trauma history and had his face examined by ultrasonography (October 1st, 2018). A well-defined, heterogenous, and hypoechoic nodule with internal vascularity was confirmed during the test (Fig. 1). The dermatology department considered it as a benign inflammatory nodule and referred him to us for surgical excision. We operated by a simple excision and sent the clinical specimen to the pathology department (October 31st, 2018). In the pathologic report, we noticed that the

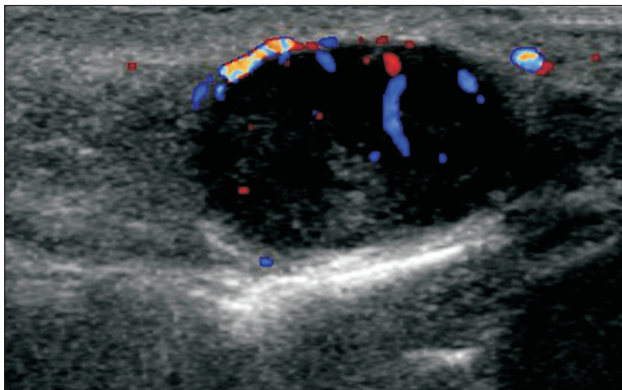


Fig. 1. Ultrasonography.

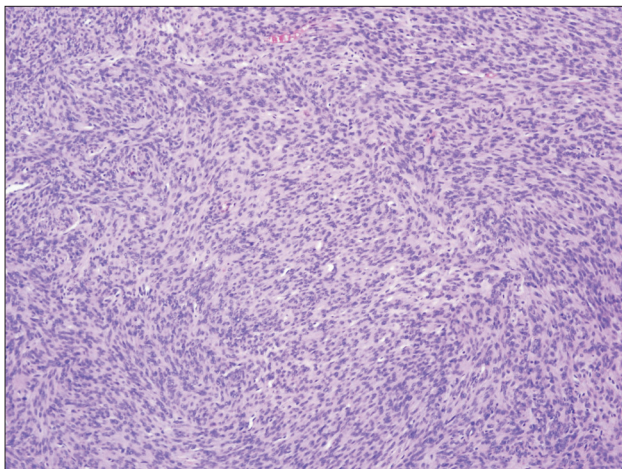


Fig. 2. Pathology (H&E, ×400).

mass was a CLM of grade 1 without necrosis (Fig. 2). Immunohistochemical staining revealed positive actin and desmin. After that, we referred the patient to the oncology department for thorough staging examinations. Neck computed tomography (CT; November 11th, 2018) revealed no grossly significantly enlarged bilateral cervical lymph nodes. Also, torso positron emission tomography-CT scan (November 19th, 2018) showed no definite hyper-metabolic lesions suggesting that a residual or metastatic malignancy was present. We decided to operate with a wide excision again and chose the superficial



Fig. 3. Preoperation.



Fig. 4. Intra operation: wide excision.

circumflex iliac artery perforator fasciocutaneous free flap for coverage (Fig. 3). During operation (December 14th, 2018), a muscle depth wide excision was performed with a 1 cm margin for the superior medial side and a 2 cm margin for the inferior lateral side (Fig. 4). A facial artery in the nasolabial fold was prepared as the recipient vessel and the superficial circumflex iliac artery perforator free flap was elevated on the right inguinal area (Fig. 5). We used Doppler ultrasound to locate the perforators during the flap design. Elevation was started from the inferior margin and trimmed as the depth and width of the patient's defect. Superficial branch of superficial circumflex iliac artery was found and the diameter seemed proper for micro-anastomosis. We also dissected concomitant vein of superficial circumflex iliac artery elaborately during the flap elevation. After margins were confirmed as tumor free in the frozen section test, micro-

anastomosis was performed with a 10-0 nylon on the zygoma level with end to side manner (concomitant vein micro-anastomosis was also done with the same level and manner as the artery) (Fig. 6). The patient moved to the recovery room after the surgery without major abnormality, and we confirmed that the Doppler was well traced. Color and temperature of the flap were also appropriate. The flap behaved well after surgery and there was no palpable mass on the postoperative area with no evidence of recurrence, so far (Fig. 7).

DISCUSSION

CLM can occur at any age with a peak point between the sixth to seventh decade⁴. CLM is a relatively rare soft tissue malignancy that not only occurs in the lower limbs but can also occur on the face or torso⁵. CLM usually originates from the arrector pili muscle of hair follicles and is known to be associated with a lesser distant metastasis and have a better prognosis compared to the subcutaneous type or other forms of leiomyosarcomas who have deeper origins. CLM is usually 5-60 mm in diameter and can often be found as a singular form of a reddish color. The architectural pattern can be divided into the nodular growing pattern and the diffuse growing pattern⁶. The biologic pathway of this malignancy is yet being researched, although radiation therapy, sunlight



Fig. 5. Intra operation: flap elevation.

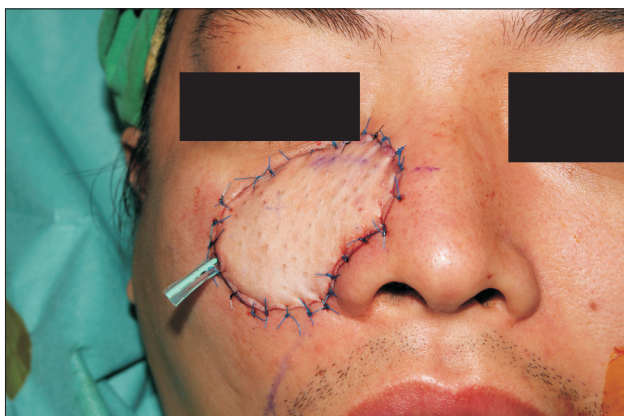


Fig. 6. Immediate postoperation.



Fig. 7. Ninety days after surgery.

exposure, traumatic injury, chemicals, and small pox can be associated with this type of malignancy. It is debatable, but male gender may also be associated with this condition. CLM diagnosis depends on the histopathological examination. A differential diagnosis with pathologic confirmation is essential because CLM's clinical manifestation mimics pyogenic granuloma, dermatofibroma, dermatofibrosarcoma, basal cell carcinoma, squamous cell carcinoma, and especially spindle cell carcinoma. In the case of spindle cell melanoma or spindle cell squamous cell carcinoma, a histologic differential diagnosis is also difficult. According to pathologists, CLM has a multitude of elongated smooth muscle cells which could be well or poorly differentiated, grouped in an irregular pattern. The tumor's boundary is indistinct and atypical cells lie in the periphery surrounding the dermal collagen bundles. Nuclear atypia and high cellularity are common; therefore, a mitotic rate of one or more per 5 high power field is regarded as the bottom line criterion for CLM diagnosis⁷. Immunohistochemical findings can aid in establishing the appropriate diagnostic. Desmin and smooth muscle actin are frequently positive in the histological examination. Both of them are considered as distinctive markers of smooth muscle differentiation. Vimentin is also commonly encountered in the immunophenotypical investigation⁸. The local recurrence rate of CLM is estimated as 30% to 50% in previous studies⁹. The metastasis rate of CLM is still debatable but approximately 5% of patients with CLM suffer complications in the long-term follow-up. Compared to the subcutaneous leiomyosarcoma where metastasis occurrence is 30 to 60%, CLM presents a much lower rate. A hematogenous pattern of metastasis is common but lymph node metastases are rarely reported. The lung is the most frequently affected organ with a metastasis occurrence found at 1 to 3 years after the first diagnosis. Prognostic factors include the size of the tumor, mitotic rate, necrosis, and vascularity¹⁰. The survival rate of CLM patients with a smaller tumor than 2 cm was of 95%. However, for patients with a larger tumor than 5 cm, the survival rate drops to 30%. The classical choice of therapy, a wide excision with a safety margin of 3 to 5 cm is still controversial⁴. In the case of

a recurrence a re-excision is recommended, but this is problematic when the tumor is located on the face. It is almost impossible to ensure a good safety margin due to the nearby structure like the nose in our case. The usage of radiotherapy is limited due to its property of causing CLM. In the case of our patient, we performed a surgical excision with a 1 cm margin for the superior medial side and a 2 cm margin for the inferior lateral side including the fascia and muscle. The safety margins were applied differently because we wanted to minimize damage to the major structures in the face. Superior medial side was concerned more about the structures related to eye and nose.

Superficial circumflex iliac artery perforator fasciocutaneous free flap has been applied not only for limb and penile defects but also for head and neck defects due to its thin and pliable quantity. Based on the superficial circumflex iliac artery which arises from the femoral artery (2.5 cm inferior to the inguinal ligament), this flap can be harvested from in a concealed inguinal area. The superficial circumflex iliac artery could be presented as a superficial and a deep branch. One of them are frequently absent, but there is usually a dominant perforator through the sartorius muscle, more chance to be the deep branch. Perforator diameter has mean value of 0.85 mm and physician's elaborate microdissection ensures pedicle with an average length of 7 cm. When the concomitant vein of superficial circumflex iliac artery is too small, cutaneous vein (1.5 to 2 mm in diameter) in the fat layer can be used as a venous drainage. Direct closure of the donor site is usually possible if the flap width is lesser than 10 cm.

Reports on the application of the superficial circumflex iliac artery perforator fasciocutaneous free flap to facial defect were relatively few considering its many advantages. Compared with full thickness skin graft, this flap can be more flexible in size selection and contain adipose tissue. However, superficial circumflex iliac artery perforator fasciocutaneous free flap has some disadvantages too. Superficial circumflex iliac artery perforator fasciocutaneous free flap is a hair-bearing flap, so it may cause cosmetic disadvantages even after survival. Color or texture mismatch also can happen too. These are why the

most proper treatment should be selected after considering the aspect of defect after tumor excision and the goals of reconstruction.

CONFLICTS OF INTEREST

The authors have nothing to disclose.

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피부 평활 근육종은 표재성 드문 질환이나 얼굴 등 신체의 어느 부분에서도 발생할 수 있다. 치료에는 적절한 광범위 절제술이 중요하며, 절제 후 유리 피판술 등을 통한 적절한 재건이 필수적이다. 환자는 33세 남자환자이며 본 사례 보고에서 특이 과거력 및 외상 경력이 없는 흡연자 환자의 얼굴에 피부 평활 근육종이 발견됐다. 우리는 내측 1 cm, 하 측면의 경우 2 cm의 여백을 두고 근육 깊이의 광범위 절제술을 시행했다. 수술 부위는 표재성 장골 회선 천공지 유리 피판술을 시행하여 절제 부위를 성공적으로 재건하였다.

색인단어: 평활근육종, 미세 천공지 피판

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