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Primary Cutaneous Leiomyosarcoma in a Healthy Child

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Dear Editor:

Cutaneous leiomyosarcoma is a rare soft tissue sarcoma of smooth muscle derivation. Although leiomyosarcomas account for approximately 24% of all soft tissue sarcoma, cutaneous leiomyosarcoma accounts for less than 5% of all adult soft tissue sarcomas[1]. Cutaneous leiomyosarcoma can be classified into dermal and subcutaneous forms. These two forms differ in clinical features and, more significantly, in predicting metastatic potential rather than mitotic activity. Cutaneous leiomyosarcoma most commonly occurs in male aged 50～69 years, and rarely affects children[2]. Although these lesions can occur anywhere on the body, they exhibit a predilection for the trunk or lower extremities and rarely arise on the head.

A 13-year-old boy presented with a solitary pink scalp nodule of several months' duration. Physical examination revealed a 1.5-cm-sized, dome-shaped, solitary pink-colored scalp nodule without ulceration or bleeding (Fig. 1). He had no underlying disease. He also had no history of trauma or radiation. Excisional biopsy revealed a poorly demarcated dermal neoplasm comprising variably oriented fascicles of spindle-shaped or fusiform cells that extended between collagen bundles, with nuclear pleomorphism and mitosis (Fig. 2A, B). Immunohistochemistry revealed diffusely positive staining for smooth muscle actin (SMA) and vimentin, but negative staining for desmin, carcinoembryonic antigen (CEA), CD34, epithelial membrane antigen, cytokeratin (CK), S-100 and human melanoma black-45 (Fig. 2C, D). The above findings led to a diagnosis of cutaneous leiomyosarcoma. There were no abnormal findings on imaging studies, including positron emission tomography-computed tomography. We did wide excision including frozen biopsy with a 1 cm resection margin, considering location, size, etc. Two years after surgery, He remains under observation with no evidence of recurrence.

As noted, cutaneous leiomyosarcomas are uncommon malignant soft tissue tumors that tend to affect older male. These lesions may correlate with radiation dermatitis, lupus vulgaris, angioleiomyoma, and trauma. The dermal form occurs in the dermis and mainly appears as a single, tender, pink or red nodule on the head or neck. The local recurrence rate of the dermal form is 30%, and distant metastasis is extremely rare. By contrast, the subcutaneous form develops in the subcutis and usually presents as subcutaneous nodules without epidermal changes on the lower extremities. The local recurrence rate is 50%～70%, and the distant metastasis rate is 30%; the lung is the most

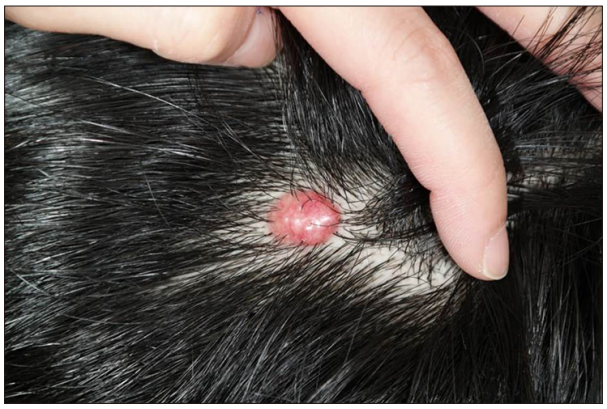

Fig. 1. Macroscopic image of a 1.5-cm-sized, dome-shaped solitary pink scalp lesion without ulceration or bleeding.

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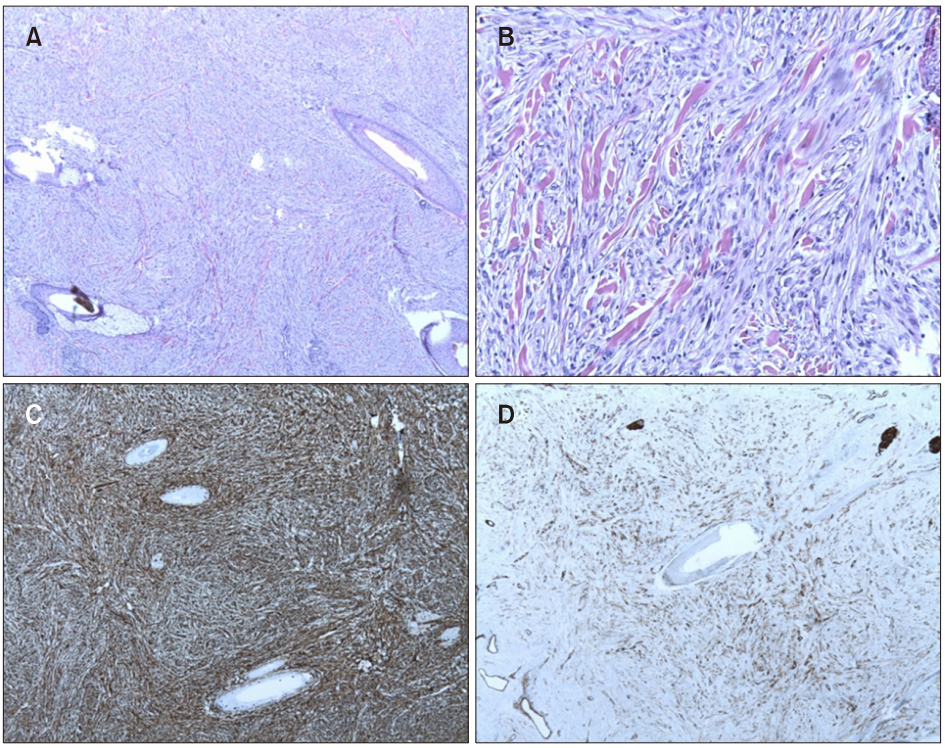

Fig. 2. (A) A poorly demarcated dermal neoplasm composed of variably oriented fascicles of spindle-shaped or fusiform cells (H&E, 40× magnification). (B) Fascicle extension between collagen bundles with nuclear pleomorphism and mitosis (H&E, 200×). Positive immunohistochemistry staining for (C) vimentin and (D) smooth muscle actin (SMA) (H&E, 40×).

common metastatic location[3].

Histological analysis may not distinguish cutaneous leiomyosarcoma from other cutaneous spindle cell tumors. The differential histologic diagnosis includes atypical fibroxanthoma, dermatofibrosarcoma protuberans, dermatofibroma, angiosarcoma, desmoplastic melanoma, and spindle cell squamous cell carcinoma. In most reported cases, cutaneous leiomyosarcomas express vimentin and SMA; however, positive desmin staining is reported in approximately 60% of cases. Sparsely positive CK and S-100 staining may be observed[4].

For both forms, wide surgical resection with a 3 to 5-cm margin is the recommended treatment. Important prognostic factors include the tumor origin, as mentioned earlier[5].

In summary, this is the first reported case of a primary cutaneous leiomyosarcoma on the scalp of an otherwise healthy 13-year-old male Korean child.

We received the patient's consent form about publishing all photographic materials.

CONFLICTS OF INTEREST

The authors have nothing to disclose.

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