

A Case of Multiple Agminated Spitz Nevi Showing Desmoplastic Changes

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We report a case of multiple agminated Spitz nevi in a 16-year-old boy who presented with multiple nodules on the right side of his face. His past history revealed that there had been erythematous and brown colored papules on the same site at the age of four. At that time one lesion was excised and histopathological findings showed fairly well circumscribed spindle and epithelioid cell nests consistent with a Spitz nevus. Twelve years later, he presented with multiple agminated brown to black colored nodules on the face. Microscopic evaluation revealed nevus cells diffusely distributed throughout the dermis showing no or little junctional activity. In addition, these cells were embedded in a strikingly desmoplastic stroma which consisted of acellular collagen fiber. Immunohistochemical staining for S-100 protein was positive. To our knowledge this is the first report of multiple agminated Spitz nevi in Korea.

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Key Words : Agminated, Desmoplastic, Spitz nevus

Spitz nevus is a benign melanocytic lesion that shares many melanocytic features with malignant melanoma. The solitary form, which is the most common, usually arises during early life and appears as a pink or brown dome shaped nodule on the head and neck¹. In rare instances, multiple tumors are encountered either agminated in one area or widely disseminated². Desmoplastic changes are noted in some Spitz nevi. We present a case of multiple agminated Spitz nevi showing desmoplastic changes.

CASE REPORT

A 16-year-old male visited our department for the evaluation of multiple brown to black colored nodules arising in a hyperpigmented area on the

right side of his face(Fig.1). His past history revealed that there had been erythematous and brown colored papules on the same site at the age of four(Fig.2). At that time one of the papules was excised. A microscopic review of the biopsy block revealed fairly well circumscribed spindle and epithelioid cell nests in the upper dermis(Fig.3) and an eosinophilic globoid body(Kamino body) in the epidermis(Fig.4). Immunohistochemical staining for S-100 protein was positive. Since that time, the number and size of the papules have been progressively increasing and enlarging in size on the right side of face only. When he was seen again at the age of sixteen, his general condition was relatively healthy and on physical examination, the cervical lymph nodes were not palpable and there were no other similar skin tumors on the rest of his body. Routine laboratory tests including a complete blood cell count, urinalysis, liver function test and chest X-ray were negative or within normal limits. A biopsy specimen was obtained from a black colored nodule. Microscopic evaluation revealed nevus cells diffusely distributed throughout the dermis. Nevus cells were embedded in a strikingly desmoplastic stroma which consisted of acel-

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Fig. 1. Brown to black colored multiple agminate nodules arising in an area of slight hyperpigmentation on the right side of the face.

Fig. 2. Multiple erythematous and brown colored papules on the right side of the face.

Fig. 3. Fairly well circumscribed spindle and epithelioid nevus cells in the dermis(H & E stain, $\times 200$).

Fig. 4. Eosinophilic globoid body(arrow) in the epidermis(H & E stain, $\times 200$).

lular collagen fiber and showed no or little junctional activity(Fig.5). Some nevus cells had intranuclear inclusion bodies. With increasing depth, these nevus cells became smaller and looked more like the cells of the common melanocytic nevus. Immunohistochemical staining revealed positivity to S-100 protein(Fig.6) and vimentin but negativity to cytokeratin suggesting that these cells originated from the melanocytic nevus.

Surgical excision of some nodules were done and a follow-up examination for 2 years revealed no evidence of recurrence.

COMMENT

Spitz nevus usually arises during the first two decades of life and appears as a pink or brown dome-shaped nodule on the head, neck, leg, or trunk¹⁻³. The solitary form is the most common but

Fig. 5. Nevus cells diffusely distributed throughout a desmoplastic dermis with no junctional activity (H & E stain, $\times 40$).

clinical variants include the multiple disseminated type often showing an eruptive development and very rarely, multiple Spitz nevi in an agminated distribution^{4,8}. The most common site for this agminated variant to arise is the face, predominantly the cheeks^{5,6}. To the best of our knowledge, in spite of several reported cases of Spitz nevi in Korea, we could not find any case of multiple agminated presentation.

Various etiological factors involved in the development of multiple agminated Spitz nevi have been postulated. These include sun exposure⁹, trauma, intralesional injection, radiation therapy, excessive innervation of the involved tissue and excision of a single Spitz nevus¹⁰. An association between multiple agminated Spitz nevi and inconspicuous melanocytic nevi within the affected area has also been observed¹⁰, which suggests that multiple agminated Spitz nevi are a type of melanocytic hamartoma. In our case, new lesions began to develop after excision of a single papule at the age of four. So we suspected that excision of a lesion might contribute to the development of new lesion.

In some Spitz nevi, desmoplastic changes are noted. Unlike ordinary Spitz nevus, desmoplastic Spitz nevi rarely show junctional activity, nest for-

Fig. 6. Immunohistochemically positive with S-100 stain (S-100 stain, $\times 400$).

mation or pigmentation³ and occur predominantly in adults. Generally, desmoplasia was related to involutional fibrosis and tumor regression but it could be a response to trauma, and reactive stromal induction by tumor cells rather than by a regressive phenomenon¹¹. For example, Spitz nevi have been known to recur following excision¹²⁻¹⁴. The recurrent lesion is commonly a deeply pigmented nodule and shows atypical clinical features especially appearing as a thickened, keloid-like lesions. Histological particularities include no or little junctional activity, extension to the deep reticular dermis or occasionally into subcutaneous fat, and dermal fibroplasia¹⁴. All these histological features were present in our case. Thus we thought that desmoplasia observed in our case might be induced by trauma such as excisional biopsy.

Desmoplastic Spitz nevi may be confused with a variety of fibrohistiocytic lesions and desmoplastic malignant melanoma¹¹. Desmoplastic melanoma almost always show an associated lentiginous melanoma, whereas any junctional activity is uncommon in desmoplastic nevus. The fibroblasts in desmoplastic nevus show no evidence of atypia, but in desmoplastic melanoma the reactive fibroblasts may be almost indistinguishable from the spindle-shaped melanoma cells. Finally, necrosis of tumor cells and collagen can be seen in desmoplastic melanoma but is not a feature of desmoplastic nevus¹¹. Recently, eosinophilic globules, at the dermoepidermal junction in Spitz nevi, have been described by several authors¹⁵⁻¹⁶. These globules were first described by Kamino in 1979 who re-

ported the presence of these eosinophilic bodies within the epidermis in 60% of 293 cases of Spitz nevi and in only 2% of 293 cases of malignant melanoma¹⁵. These globules were named "Kamino bodies" and awareness of this morphological marker has aided in the ability to distinguish between Spitz nevi and malignant melanoma. In our case, multiple agminated Spitz nevi developed following a previous surgical excision of a lesion on the face. They simulated melanoma clinically, because of heavily pigmented larger nodules as opposed to ordinary Spitz nevi and a long duration elapsed since diagnosis. However, the biopsy specimen showed marked desmoplasia with compressed nevus cells, no cellular atypia, and eosinophilic globules in the epidermis.

To our knowledge, this variety of presentation has not been previously reported in Korea. Our case adds another case to the literature of multiple agminated Spitz nevi and documents a 12-year follow-up.

Herein, we confirm the benign nature of this entity and suggest that desmoplasia may be induced in the course of time following trauma such as surgical manipulation.

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