Intradermal Nodular Fasciitis: A Case Report
Po-Hsuan Lu   Yang-Chih Lin   Yu-Hung Wu

Nodular fasciitis is a reactive myofibroblastic proliferative lesion frequently misdiagnosed as a sarcoma. Most lesions are located in the subcutaneous tissue of the upper extremities. Rarely, intravascular, intradermal, and cranial variants have been described. Intradermal nodular fasciitis, first reported by Goodlad and Fletcher in 1990, may be confused with fibrohistiocytic tumors because of the unexpected location.

A 32-year-old male had a rapidly growing tender nodule in the occipital region of the scalp for one month. Histopathology revealed an ill-defined dermal nodule composed of interwining fascicles of spindle cells in a loose myxoid stroma with focal hemorrhage. The diagnosis was intradermal nodular fasciitis. Because of the possibility of cranial fasciitis, skull films were obtained to evaluate for possible bony involvement, but no lesion was identified. Although the deep surgical margin contained tumor spindle cells, MRI showed only a linear low intensity signal suggestive of scar. We report a case of intradermal nodular fasciitis and discuss the clinical, pathologic and radiographic findings. (Dermatol Sinica 23: 212-216, 2005)

Key words: Intradermal nodular fasciitis, Cranial fasciitis
INTRODUCTION
Nodular fasciitis is a benign, pseudosarcomatous lesion that often presents in the forearm of young adults. Nodular fasciitis usually occurs in the subcutis, less often in muscle, and rarely in the dermis. Since the first report by Goodlad and Fletcher in 1990, only 11 intradermal nodular fasciitis have been described. We report such an intradermal lesion on the scalp and review the literature.

CASE REPORT
A 32-year-old male noted a rapidly growing tender nodule in on the posterior occipital scalp for one month (Fig. 1). He denied any previous trauma. Despite oral antibiotics and nonsteroidal anti-inflammatory agents, the nodule continued to enlarge and remained painful. There was a 2 x 2 cm, erythematous, firm, immovable, tender nodule with a central slit-like opening in the posterior occipital scalp. The physical examination was otherwise normal. The initial clinical diagnosis was an inflamed epidermal cyst, and local excision was performed.

Histopathology revealed an ill-defined dermal nodule composed of intertwining fascicles of spindle cells in a loose, myxoid, vascular stroma presenting a tissue culture-like pattern. (Fig. 2 A & 2B). The spindle cells with ovoid, vesicular nuclei arranged haphazardly. The differential diagnosis included dermatofibroma, spindle cell carcinoma, dermatofibrosarcoma protuberans, and angiosarcoma. The spindle cells were arranged haphazardly. The vascular component included well-formed capillaries with extravasation of red blood cells. (Fig. 2C) The spindle cells with ovoid, vesicular nuclei arranged haphazardly. Although mitoses can be frequently found occasionally, no atypical mitoses or nuclear pleomorphism was seen. Multinucleated giant cells were frequently present. (Fig. 2D) Immunohistochemically, the spindle cells were negative for cytokeratin, S-100 and CD34 (Fig. 3A), which excluded the possibility of spindle cell squamous cell carcinoma, epithelioid sarcoma, melanoma, angiosarcoma and DFSP. They stained diffusely for smooth muscle actin (SMA) in a fine granular pattern suggestive of myofibroblastic proliferation (Fig. 3B). A few multinucleated giant cells expressing CD68 were present (Fig. 3C). A diagnosis of intradermal nodular fasciitis was made. Because of the lesion’s location was in the

Fig. 1
A 2 X 2 cm, tender, erythematous nodular in the occipital scalp.

Fig. 2
(A) An ill-defined nodular infiltration of spindle cells in the dermis. (H & E, 20X)
(B) Spindle cells are haphazardly arranged in a loose myxoid and vascular stroma. (H & E, 100X)
(C) Slit-like space with extravasation of erythrocytes in a myxoid feathery stroma (H & E, 200X)
(D) Multinucleated giant cells and scattered mitoses are present in a loosely structured stroma with extravasated erythrocytes (H & E, 400X)
occipital scalp, we were also concerned about the possibility of cranial fasciitis. However, skull films were showed negative for any abnormalities.

There were no symptoms or palpable residual lesion after excision. However, because of the extension presence of the tumor spindle cells into the surgical margins, as well as the rare intradermal, occipital location of the tumor, further excision was performed. The pathology of the specimen was indistinguishable from the initial tissue other than being more well-defined. The deep surgical margin was still involved by the tumor spindle cells. Therefore, to evaluate the extent of soft tissue involvement, an MRI was performed with and without contrast enhancement with T1WI, FLAIR, T2WI as well as DWI and ADC mapping on axial sections and T1WI on sagittal and coronal sections (Fig. 4A & 4B). There was no evidence of a tumor or bony extension. A linear low intensity signal seen in the left occipital subcutaneous area was consistent with a postoperative scar. There was no evidence of recurrence. Four months after the second excision, there was no evidence of recurrence.

**DISCUSSION**

Classically, nodular fasciitis is a benign, soft tissue lesion usually occurring in young adults with an equal sex distribution. Most frequently the upper extremities are involved, followed by the lower extremities and the trunk. The lesion is usually subcutaneous, fascial, or intramuscular and rarely occurs in the dermis. Postoperative or post-traumatic spindle cell nodules have also been described as the dermal analog of nodular fasciitis. Eleven cases of intradermal nodular fasciitis have been previously reported in the English literature (Table 1). The reported age of onset has ranged from 2 to 59, with a median of 25.3 years. There is no apparent sexual preponderance. The lesions usually present over weeks to months as rapidly growing nodules for months to weeks. Three of the 12 patients, including ours, had tender nodules sensation. Several patients have had a history of trauma, although ours did not.
not. The treatment in all cases was simple excision, and recurrences have been rare. In fact, based on a large series of cases of studies in subcutaneous nodular fasciitis, it has been suggested that recurrence may exclude this diagnosis. In our patient, the tumor was still seen on the specimen from after the second procedure—excision. We consider it to be the result of incomplete excision rather than recurrence.

Pathohistologically, dermal spindled cell tumors including dermatofibroma, spindle cell squamous cell carcinoma, spindled cell melanoma, atypical fibroxanthoma, dermatofibrosarcoma protuberans, and angiosarcoma, should be considered in the differential diagnosis. Although all these tumors may have a fascicular arrangement of spindled cell, most of them have unique immunohistochemical staining profiles. Despite of the unusual dermal location, the histology of the lesion in our patient was typical of nodular fasciitis, characterized by the haphazardly arranged spindle cells with vesicular nuclei showed distinctive alternating cellular and “feathery” myxoid zones, with vascular components. The bland-looking cytologic features, absence of nuclear atypia, hyperchromasia, and the immunohistochemical stains results helps to establish the diagnosis of nodular fasciitis despite of the unusual dermal location. The differential diagnosis of dermatofibroma, a dermal lesion, was excluded because of the absence of storiform pattern and entrapped collagen. The diagnosis of intradermal nodular fasciitis was therefore based on the histology and immunohistochemical stains and was supported by no recurrence after second excision.

The etiology of nodular fasciitis remains unknown. It appears to be a reactive myofibroblastic proliferative lesion, suggesting a traumatic cause. However, only a small number of patients have a definite history of trauma. Of the 12 cases of intradermal nodular fasciitis, seven have occurred on the head. This may be due to the more direct attachment of the facial musculature to relatively thin skin via the superficial musculoaponeurotic system. In contrast, only 10% to 20% of nodular fasciitis in subcutaneous, muscular or fascial location involve the head and neck. They usually appear in the subcutis of the face, so that a scalp lesion as in our patient is very rare. Because his tender nodule in the occipital scalp, we considered the possibility of cranial fasciitis. Although that entity occurs almost exclusively in children under the age of 6 years, it has been reported in at least one adult. Cranial fasciitis in adults may be silent since infancy. Cranial fasciitis of childhood is an unusual variant of nodular fasciitis that occurs in infants and children. It is a rapidly growing mass in the subcutaneous tissue of the scalp that extends into the underlying cranium. Radiographically, the lesion usually appears as a single lytic defect in the calvarium with an associated soft tissue mass. In one report of cranial fasciitis in adults, the tumor produces cortical saucerization on CT scan and exhibits intermediate intensity on T1 and T2-weighted MR images. In the other case of adult cranial fasciitis, imaging studies reveal a left frontal cystic lesion with variable signal intensities disrupting of the inner surface of the skull. Because there is no evidence of infiltration of the cranial bone on skull films, or MRI or histology in our patient, we exclude the possibility of cranial fasciitis.

The appearance of nodular fasciitis on MRI is nonspecific and varies according to the histology of the lesion. Myxoid or cellular lesions are hyper-intense to skeletal muscle on T1-weighted and hyper-intense to fat on T2-weighted MR images. Nodular fasciitis with a fibrous histology has low signal intensities on all sequences. In fact, most soft tissue masses involving the head and neck have a nonspecific imaging appearance. Because intradermal nodular fasciitis is a benign lesion, when it occurs on the scalp we suggest that MRI or other imaging techniques be used to look for were helpful for determining local bony destruction or intracranial extension. If none is seen, more extensive surgery and its attendant anxiety can be avoided.
REFERENCES