Intravascular fasciitis of the scalp: A case report

Case report

A healthy 24-year-old woman presented with an asymptomatic and rapidly growing swelling on the scalp that had been present for 3 months. Clinical examination revealed a 3 cm × 2.5 cm, movable, firm, and deep-seated subcutaneous nodule on her occipital scalp (Figure 1). No irritation or erosion was noted. There was no history of previous trauma, insect bites, or nodule drainage. A schwannoma was suspected clinically and surgical excision performed.

Histopathological examination revealed a well-circumscribed, sausage-shaped, subcutaneous neoplasm that was entirely localized within the lumen of a segmental dilated vein (Figure 2A). The lesion was largely composed of interlacing fascicles of predominantly pleomorphic, plump spindle cells in a loose stroma (Figure 2B). Bland vesicular nuclei, extravasations of erythrocytes, and scattered lymphocyte infiltration were evident under high magnification (Figure 2C). Immunohistochemical staining results showed that most of the spindle cells were positive for smooth muscle actin and negative for CD 31, evidence of myofibroblastic differentiation (Figure 3). Immunohistochemical staining for CD31 demonstrated positively stained vascular endothelial cells outlining the intravascular lesion (Figure 3). A diagnosis of intravascular fasciitis was made based on the clinical and pathological findings. The patient was asymptomatic with no evidence of tumor recurrence after 6 months.

Discussion

Nodular fasciitis, also known as nodular pseudosarcomatous fasciitis, is a common benign reactive proliferation of the soft tissue. Nodular fasciitis is composed of a high cellularity of myoblasts and fibroblasts, and it is sometimes mistaken for sarcoma because of its rapid clinical growth pattern and histological pleomorphism. Unusual clinical and pathological variants of nodular fasciitis such as intradermal, cranial, intravascular, intra-articular, and proliferative fasciitis have been reported.

Intravascular fasciitis, first described in 1981 by Patchefsky and Enzinger, is particularly rare. Since 1981, fewer than 30 cases of similar lesions have been reported, most of which were located in the limbs, trunk, or oral submucosa. Presentation on the scalp, as in our case, is extraordinary and increases the clinical and histopathological diagnostic difficulty. Clinically, the lesion usually presents as a solitary, well demarcated, rapidly growing, and asymptomatic nodule in the subcutaneous tissue or fascia at any age, especially in adults younger than 30 years. Males and females are equally affected. The lesion size varies (0.5–5 cm) and has a mean diameter of 1.5 cm.

Histologically, intravascular fasciitis demonstrates the same features as nodular fasciitis in the presentation of reactive plump myofibroblast proliferation in a storiform stroma; however, only intravascular fasciitis presents the distinct characteristics of intraluminal, intramural, or extravascular vessel involvement. The involved vessels range from small superficial veins to large subcutaneous arteries. Concurrence of extravascular nodular fasciitis and intravascular fasciitis has been reported, which suggests lesion multi-centricity or direct extension of the extravascular fasciitis to the adjacent vascular structure. The lesion grows primarily along the vascular lumen with a serpentine or multi-lobular pattern, and possibly involves vessel wall intima, media, or adventitia. Such feathery myofibroblastic violation of the vascular endothelial network in this benign condition may be inappropriately mistaken as malignant vascular invasion. As in ordinary nodular fasciitis, its components may be extravasations of erythrocyte, lymphocyte infiltration and the presence of osteoclast-like giant cells. Mitosis was observed throughout the entire lesion.

Controversy exists regarding the pathogenesis of intravascular fasciitis. Pre-existing trauma, venous thrombosis formation, and viral infection-related myofibroblastic proliferation have all been implicated in the etiology of intravascular fasciitis. Amarjit et al. reported intravascular fasciitis in a 16-week pregnant female, suggesting that the estrogen-related hormonal changes associated with pregnancy may be a contributing factor. There was no history of those events in our case. The reporting of similar cases will better define the epidemiology and pathogenesis of these extremely rare tumors.

The clinical diagnosis of intravascular fasciitis is difficult, and surgical excision is usually required to obtain a definitive answer. The major pathological differential diagnosis of intravascular fasciitis includes intravascular fibroangiomatous proliferations: Mason's tumor, angioleiomyoma, intravascular pyogenic granuloma, and intravascular myopericytoma. Masson's tumor, also known as intravascular papillary endothelial hyperplasia, is characterized by unusual endothelial proliferation in an organizing thrombus usually on the extremities. The histopathological features of Masson's tumor usually present with a mass of anastomosing vessel channels and a varying degree of intraluminal papillary projection. Angioleiomyoma, a relatively common neoplasm, usually presents as painful subcutaneous nodules on the lower legs of middle-aged women. It originates from the tunica media of a vessel wall and is composed of monomorphic mature smooth muscle cells that are distributed and intersected between numerous vascular channels. Intravascular pyogenic granuloma is characteristic of asymptomatic polypoid masses of angiomatous tissue, with typical lobular growth of the capillaries lined by flattened or rounded endothelial cells. It usually involves veins of the neck and upper extremities, and has a slight female predominance.
myopericytoma, a recently identified vascular tumor, is characterized by myoid-like spindle cells arranged in a concentric pattern around the blood vessels. In addition, immunohistochemical staining supports the notion that the proliferative spindle cells in intravascular fasciitis are of myofibroblastic differentiation by positive smooth muscle actin staining and negative CD31 expression, whereas CD31 markers are often observed in angiomyofibroblastic neoplasms.

Due to rich cellularity and vascular invasion, intravascular fasciitis should also be differentiated from vascular leiomyosarcoma. Vascular leiomyosarcoma, a very rare malignant neoplasm, differentiates from the tunica media of the vessels. Compared with intravascular fasciitis, intravascular leiomyosarcoma is usually located in the large veins, e.g., the inferior vena cava or the external iliac vein, and presents with an ill-defined infiltrative pattern, significant cytological pleomorphism, and marked mitotic activity histologically.

Cranial fasciitis should be considered in the differential diagnosis in our case, which presented lesions with nodular fasciitis-like histological features on the scalp. However, cranial fasciitis is a benign fibroblastic proliferation of the scalp that occurs almost exclusively in children younger than 6 years.

Treatment is surgical excision and the general prognosis is good without a tendency for recurrence or hematogenous spread. Only rare cases of local recurrence following surgical excision have been reported.

In conclusion, it is important to be aware of intravascular fasciitis, as it could be misdiagnosed as an intravascular neoplasm or sarcoma. Although intravascular fasciitis is quite rare, it should be included in the differential diagnosis for rapidly growing subcutaneous scalp nodules in patients whose age and clinical presentation suggest this possibility. To avoid a misdiagnosis of malignancy, careful attention should be paid to be the bland histological

Figure 1 On physical examination, the lesion is a 3 × 2.5-cm, movable, firm and deep-seated subcutaneous nodule on her occipital scalp. The overlying epidermis is intact without irritation or erosion.

Figure 2 (A) At scanning magnification, there is an intravascular sausage-shaped, well-demarcated polypoid nodule (hematoxylin–eosin staining [H&E], original magnification, ×40). (B) The nodule is composed of pleomorphic spindle cells with plump vesicular nuclei in a dense fascicular growth pattern (H&E, original magnification, ×100). (C) On high magnification, bland-appearing nuclei, extravasated erythrocytes, and scattered lymphocytes are evident (H&E, original magnification, ×400).
features. Proper diagnosis is crucial to prevent unnecessary aggressive investigation and wide excision.

Yi-Hsin Hsiao, Cheng-Sheng Chiu*
Department of Dermatology, Chang Gung Memorial Hospital, Taiwan
*Corresponding author. 199, Tun-Hwa North Road, Taipei 105, Taiwan. Tel.: +886 2 27135211 x3397; fax: +886 2 27191623. E-mail address: a9239@adm.cgmh.org.tw (C.-S. Chiu)

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Received: Jun 20, 2011
Revised: Oct 4, 2011
Accepted: Dec 12, 2011