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SHORT COMMUNICATION

Scalp Angiosarcoma Presented as Skin-coloured Papules

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Angiosarcomas of the skin are uncommon but highly malignant tumours that mainly occur in the head and neck regions of elderly patients. The usual presentation of angiosarcoma is as bruise-like patches or nodules, but variable clinical features are demonstrated (1). We describe a case of angiosarcoma on the scalp that presented as skin-coloured papules.

CASE REPORT

A 77-year-old Korean man visited our clinic after noticing two papules on his scalp that had persisted for several months. His medical history revealed diabetes, hypertension, and a history of colon polyp. On examination, one papule of 0.7 × 0.7 cm and another of 0.5 × 0.5 cm were observed on the parietal scalp. Both papules were dome-shaped, smooth-surfaced, skin-coloured, and non-tender (Fig. 1). A skin biopsy was performed under the presumption of benign appendageal tumours. The histological examination, however, revealed a dermis infiltrated by irregularly anastomosing vascular channels lined by moderately differentiated enlarged endothelial cells permeating between collagen bundles (Fig. 2A). Atypical and hyperchromatic endothelial cells were frequently encountered (Fig. 2B). Additionally, dense lymphocytic infiltration was observed between vascular components. The immunohistochemical investigation documented that the tumour cells were positive for CD31 and CD34, and about 30% of them were positive for Ki-67. They were found to be negative for von Willebrand factor, smooth muscle actin, HMB-45, and HHV-8. Vascular endothelial growth factor (VEGF) and VEGF receptor

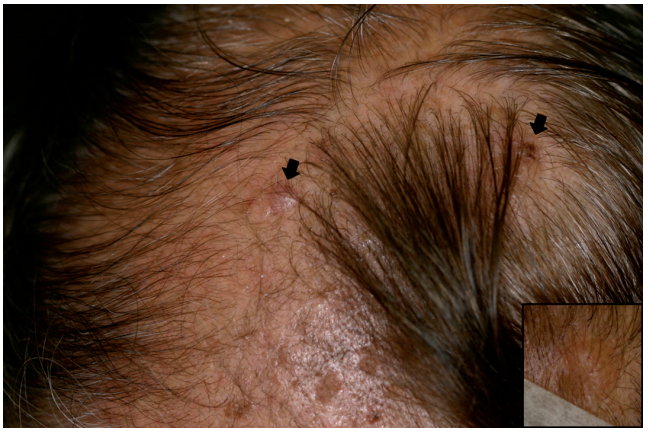

Fig. 1. Two skin-coloured, dome-shaped, non-tender papules were observed on the parietal scalp. The crust of the papule on the right side formed as a result of biopsy. (Inlet: close-up view.)

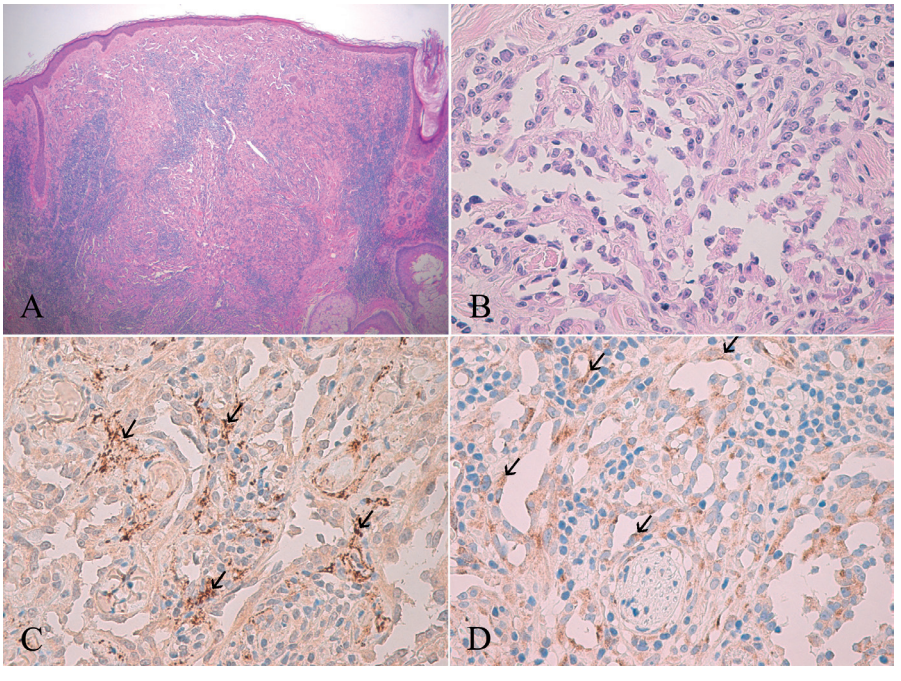

Fig. 2. The dermis was infiltrated by irregularly anastomosing vascular channels lined by moderately differentiated enlarged endothelial cells permeating between collagen bundles. Dense lymphocytic infiltration was observed between vascular components. (A: H&E, × 40) Hyperchromatic or enlarged atypical endothelial cells were poorly lining the vascular channels. (B: H&E, × 400) The expression of vascular endothelial growth factor (VEGF) was detected mostly in the interstitium (*arrows*) and VEGF receptor was moderately expressed in the endothelial cells (*arrow*). (C: VEGF, × 400; D: VEGFR, × 400).

(VEGFR) stains were moderately positive (Fig 2C, D). Lymphocytes were positive for CD3, CD4, CD5, CD8, CD45RO and CD20 but negative for CD56 and granzyme, indicating mixed infiltration of T and B lymphocytes. Brain magnetic resonance imaging, computed tomography (CT) of the abdomen and whole-body positron emission tomography were negative for metastasis. Mildly enlarged lymph nodes were noted in the left paratracheal and hilar area on the chest CT, but lymph node biopsies showed no evidence of malignancy. The patient was treated with wide local excision and skin grafts. After one year of follow-up, two new papules, which were also skin-coloured and similar to the initial lesions, developed on the scalp beside the surgical scars. Re-biopsy revealed the multifocal appearance of angiosarcomas and the patient is now under radiation therapy.

DISCUSSION

Angiosarcoma of the face and scalp in the elderly usually presents as bruise-like patches or violaceous nodules. On progression, the lesions may ulcerate and bleed. Rare clinical presentations include rosacea-like eruption, angiooedema, and scarring alopecia (1). Based on this case, skin-coloured papules should be added to the clinical features of cutaneous angiosarcoma. When skin-coloured papules or nodules arise on the head and neck region, appendageal tumours should be differentiated. A variety of appendageal tumours, such as syringoma, trichoepithelioma, and trichoblastoma, etc., are included in the differential diagnosis because of indistinctive clinical findings, requiring histopathologic examination for diagnosis (2).

Our case was characterised by a prominent lymphocytic infiltrate whereas inflammatory infiltrate is generally sparse in angiosarcoma. On rare occasions, tumours prominently infiltrated by lymphocytes simulating cutaneous lymphoma or pseudolymphoma are reported (3). Dense lymphocytic infiltration and less differentiation of the tumour may account for the colour of the lesions. Maddox & Evans (4) previously reported that a moderate to marked lymphoid infiltrate in and around the tumour has significantly favourable prognostic value in angiosarcoma and showed that the tumours with the most intense inflammatory responses had longer patient survival, increased intervals to recurrence, and less frequent metastases. Our case showed a prominent lymphoid infiltrate and is possibly expected to have a better prognosis. Overexpression of VEGF was observed in 80% of angiosarcomas (5) and VEGFR expression was also reported in angiosarcoma. VEGFR expression was more frequent in better differentiated tumours and may play a role in predicting a particular patient's clinical course (6). Consistent with the previous reports, VEGF and VEGFR were moderately expressed on vascular channels in this case. This may suggest the possible use of anti-angiogenic treatment targeting VEGF, such as bevacizumab, if required along this patient's clinical course.

REFERENCES

1. Mentzel T, Kutzner H, Wollina U. Cutaneous angiosarcoma of the face: clinicopathologic and immunohistochemical study of a case resembling rosacea clinically. J Am Acad Dermatol 1998; 38: 837–840.
2. Srivastava D, Taylor RS. Appendage tumors and hamartomas of the skin. In: Goldsmith LA, editors. Fitzpatrick's dermatology in general medicine. New York: McGraw-Hill Medical 2012; p. 1337–1362.
3. Requena L, Santonja C, Stutz N, Kaddu S, Weenig RH, Kutzner H, et al. Pseudolymphomatous cutaneous angiosarcoma: a rare variant of cutaneous angiosarcoma readily mistaken for cutaneous lymphoma. Am J Dermatopathol 2007; 29: 342–350.
4. Maddox JC, Evans HL. Angiosarcoma of skin and soft tissue: a study of forty-four cases. Cancer 1981; 48: 1907–1921.
5. Zietz C, Rossle M, Haas C, Sendelhofert A, Hirschmann A, Sturzl M, et al. MDM-2 oncoprotein overexpression, p53 gene mutation, and VEGF up-regulation in angiosarcomas. Am J Pathol 1998; 153: 1425–1433.
6. Gennaro M, Valeri B, Casalini P, Carcangiu ML, Gronchi A, Conti AR, et al. Angiosarcoma of the breast and vascular endothelial growth factor receptor. Tumori 2010; 96: 930–935.