Zosteriform Venous Malformations with an Atypical Presentation

Shinji Noda[1,2], Yoshihide Asano[2]*, Rumi Sugie[1], Kyoko Mizuno[1] and Makoto Adachi[1]
*Department of Dermatology, [1]Kanto Rosai Hospital, Kawasaki, Kanagawa, [2]Faculty of Medicine, University of Tokyo, 7-3-1 Hongo, Bunkyo-ku, Tokyo, 113-8655, Japan. *E-mail: yasano-tky@umin.ac.jp*
Accepted October 18, 2010.

Venous malformations, previously called cavernous haemangiomas, are categorized as slow-flow vascular malformations. A few cases of venous malformations with a dermatomal distribution have been described previously (1–4). We call this clinical entity "zosteriform venous malformations", while the terms "zosteriform cavernous haemangiomas" and "unilateral dermatomal cavernous haemangiomatosis" were previously used to describe the lesions. We report here a case of venous malformations with a unilateral C6 dermatomal distribution and summarize the available literature regarding venous malformations with this characteristic distribution pattern.

CASE REPORT

A 58-year-old Japanese man presented with a 5-year history of two gradually-enlarging, bluish nodular lesions involving the right upper extremity. The patient was otherwise healthy. No pain or pruritus was noted. The lesions were sessile, spherical, smooth-surfaced, and were located on the lateral flexor surface of the right forearm and the radial aspect of the wrist joint (Fig. 1). They measured 35 mm and 25 mm in diameter, respectively. They were distributed along the C6 dermatome. No tenderness was noted, but palpable pulsations and surrounding dilated veins were found on the wrist lesion. Neither hyperhidrosis over the lesions nor hypertrophy of the right arm was detected. The patient's family history was negative for similar lesions. Laboratory examinations were unremarkable. No visceral masses were detected in imaging studies, including computed tomography, esophagogastroduodenoscopy, and colonoscopy.

Magnetic resonance imaging (MRI) revealed two intramuscular masses, both measuring approximately 30 mm in diameter, in addition to two peripheral ones corresponding to the skin lesions (Fig. 2). All four lesions were oriented along the long axis of the extremity, strictly following radial neurovascular distribution: two intramuscular lesions followed the deep branch of the radial nerve that runs along the radius, while two superficial lesions followed the superficial branch of the radial nerve. These masses had signal intensities similar to that of skeletal muscle on T1-weighted images, and higher signal intensities in comparison with skeletal muscle on T2-weighted images. Inhomogeneous enhancement was observed on gadolinium-enhanced T1- and T2-weighted MRI. Needle puncture of the wrist tumour caused spurts of bright-red blood. Histological examination of an excisional biopsy specimen from the forearm tumour demonstrated multiple irregular dilated blood vessels with congested erythrocytes in the dermis and subcutaneous tissue (Fig. 3). A single layer of flat mature endothelial cells lined each vessel. A diagnosis of venous malformations was made.

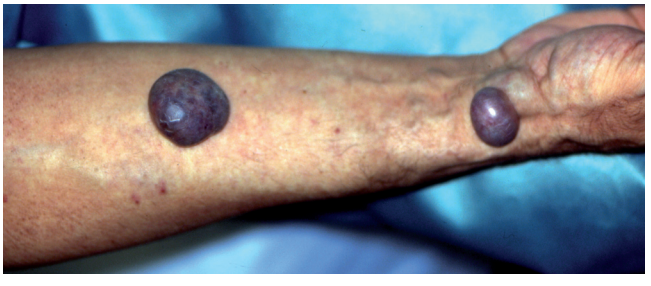

Fig. 1. Pedunculated, spherical, smoothly surfaced, bluish tumours on the lateral flexor surface of the right forearm and the radial aspect of the wrist joint, distributed along the C6 dermatome.

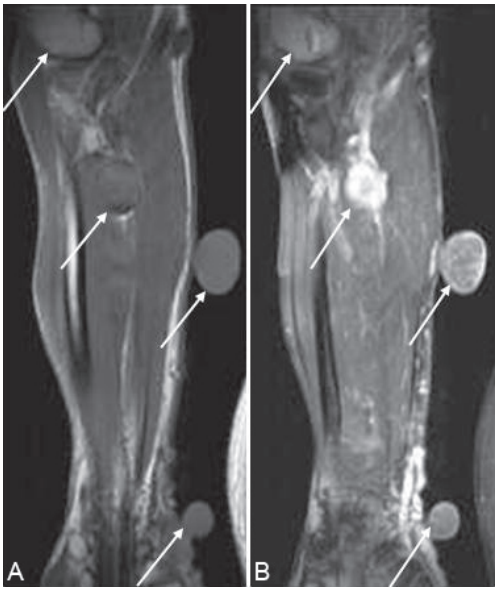

Fig. 2. (A) Coronal T1-weighted magnetic resonance imaging shows two intramuscular masses and two peripheral ones compatible with the skin lesions (*arrows*). These lesions have signal intensity similar to that of skeletal muscle. All four lesions strictly follow a radial neurovascular distribution. (B) On coronal T2-weighted magnetic resonance imaging, the lesions are generally hyperintense to skeletal muscle while signal intensity is varied (*arrows*).

DISCUSSION

Zosteriform venous malformations, first reported in 1977 by Steinway & Fretzin. (1), have a distinct distribution pattern limited to unilateral single or multiple adjacent dermatomes, further characterized by the absence of visceral lesions or enchondromas and a negative family history. Blue rubber bleb naevus syndrome is a rare disorder characterized by multiple venous malformations of the skin and gastrointestinal tract (5). Maffuci's syndrome is also a rare sporadic disorder consisting of enchondromas and vascular anomalies including venous malformations (5). Autosomal dominantly inherited cutaneomucosal venous malformations, caused by mutations in angiopoie-

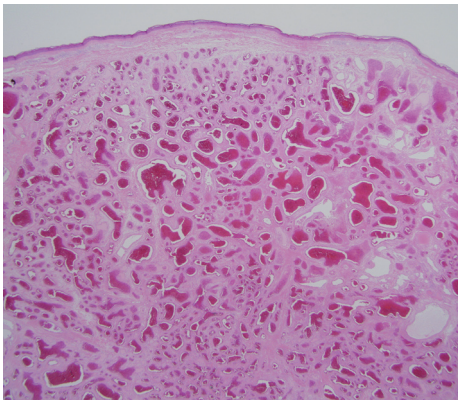

Fig. 3. Multiple irregular dilated blood vessels with congested erythrocytes in the dermis and subcutaneous tissue. Haematoxylin-eosin × 12.5.

tin receptor TIE2/TEK, have also been reported (6). These syndromes were excluded in our patient because visceral lesions and enchondromas were absent, and no family members had similar lesions. Our case is compatible with venous malformations, although they usually manifest as blue or purple ill-defined grouped nodules, whereas our patient had two discrete solitary tumours. Histologically, vascular channels were relatively small and less irregular than typical cases (7, 8). In addition, palpable pulsations and spurts of bright-red blood on needle puncture indicate the presence of an associated arteriovenous fistula in the wrist tumour. These three features have been described in rare cases of venous malformations (3, 7, 8).

Although there are no established treatments for this disease, possible therapeutic options include compression garments, aspirin, sclerotherapy, and surgical removal. Due to the widespread nature of the lesions, complete removal is difficult, especially for a zosteriform type. Thus, local recurrence can occur at a high rate. Therefore, simple observation, or conservative treatment with compression garments or aspirin are recommended, unless severe symptoms are present.

In the present case, two asymptomatic intramuscular masses incidentally detected by MRI were also considered to be venous malformations because they showed a characteristic signal intensity pattern (8, 9). To our knowledge, this is the first case of zosteriform venous malformations with clearly identified intramuscular lesions reported. MRI is certainly a great tool to evaluate the exact distribution of the lesions in venous malformations (10). Another important observation in the present case was that cutaneous and intramuscular lesions strictly follow the superficial branch and the deep branch of the radial nerve, respectively, indicating that the lesions are distributed along dermatome, but not Blaschko's lines, because Blaschko's lines are not applicable to intramuscular lesions. The cause of this characteristic neurovascular distribution has not yet been determined. However, an increasing body of evidence has allowed us to propose a plausible hypothesis for the development of the characteristic distribution of this disease. Borisov et al. (11) initially demonstrated that selective denervation induces loss of capillaries in skeletal muscle. Bates et al. (12) showed that overexpression of the soluble form of neuropilin-1 causes abnormally formed blood vessels as nerve defasciculation and a severe disruption in the peripheral nerve pattern. Indeed, vascular and nervous systems use similar signals and support each during development of the neurovascular system (13). Thus, we can speculate that ectopic overgrowth of venous blood vessels due to dysregulation of signalling in a particular nervous segment may be involved in the development of venous malformations along dermatomes.

Four further cases with zosteriform venous malformations have been reported thus far (1–4). In three cases lesions were located on several adjacent dermatomes. Although rapid enlargement in puberty and changes in size associated with the menstrual cycle were observed in one case (2), hormonal effects were not noted in the other cases. Although venous malformations are congenital anomalies caused by inborn error of vascular morphogenesis (14, 15), they are not always evident at birth (14). Rapid enlargement can occur in association with trauma, haemorrhage, partial resection, or hormonal changes. The late onset of venous malformations in our case is unusual. Vascular fragility associated with ageing and unnoticed minor traumas may influence the growth of the lesions.

REFERENCES

1. Steinway D, Fretzin D. Acquired zosteriform cavernous hemangiomas: brief clinical observation. Arch Dermatol 1977; 113: 848–849.
2. Wilkin J. Unilateral dermatomal cavernous hemangiomatosis. Dermatologica 1980; 161: 347–354.
3. Watabe H, Kashima M, Baba T, Mizoguchi M. A case of unilateral dermatomal cavernous haemangiomatosis. Br J Dermatol 2000; 143: 888–891.
4. Kosaka H, Matsumoto S, Ueda T. [A case of unilateral dermatomal cavernous hemangiomatosis]. Jpn J Dermatol 2005; 115: 2292–2294 (in Japanese).
5. Garzon M, Huang J, Enjolras O, Frieden I. Vascular malformations. Part II: associated syndromes. J Am Acad Dermatol 2007; 56: 541–564.
6. Wouters V, Limaye N, Uebelhoer M, Irrthum A, Boon L, Mulliken J, et al. Hereditary cutaneomucosal venous malformations are caused by TIE2 mutations with widely variable hyper-phosphorylating effects. Eur J Hum Genet 2010; 18: 414–420.
7. Allen P, Enzinger F. Hemangioma of skeletal muscle. An analysis of 89 cases. Cancer 1972; 29: 8–22.
8. Hein K, Mulliken J, Kozakewich H, Upton J, Burrows P. Venous malformations of skeletal muscle. Plast Reconstr Surg 2002; 110: 1625–1635.
9. Burrows P, Laor T, Paltiel H, Robertson R. Diagnostic imaging in the evaluation of vascular birthmarks. Dermatol Clin 1998; 16: 455–488.
10. Vilanova J, Barceló J, Villalón M. MR and MR angiography characterization of soft tissue vascular malformations. Curr Probl Diagn Radiol 2004; 33: 161–170.

11. Borisov A, Huang S, Carlson B. Remodeling of the vascular bed and progressive loss of capillaries in denervated skeletal muscle. Anat Rec 2000; 258: 292–304.
12. Bates D, Taylor G, Minichiello J, Farlie P, Cichowitz A, Watson N, et al. Neurovascular congruence results from a shared patterning mechanism that utilizes Semaphorin3A and Neuropilin-1. Dev Biol 2003; 255: 77–98.
13. Carmeliet P. Blood vessels and nerves: common signals, pathways and diseases. Nat Rev Genet 2003; 4: 710–720.
14. Garzon M, Huang J, Enjolras O, Frieden I. Vascular malformations: part I. J Am Acad Dermatol 2007; 56: 353–370.
15. Dobson M, Hartley R, Ashleigh R, Watson Y, Hawnaur J. MR angiography and MR imaging of symptomatic vascular malformations. Clin Radiol 1997; 52: 595–602.