Acquired Zosteriform Naevus Flammeus: A Cutaneous Marker of Central Nervous System Angiomatosis

Sir,

Naevus flammeus refers to a congenital vascular malformation, which is usually unilateral and commonly involves the forehead, face and neck, although other sites can be involved too. Most of the lesions are small, located on the glabellar region (salmon patch) and tend to disappear during infancy or childhood. Lesions located on the nucha, known colloquially as "stork bite" also tend to fade with age. On the contrary, large facial naevus flammeus, also named port-wine stain, persists during adulthood, and in some cases purple, nodular, warty, or cobblestone-like elevations may develop on them. On the face, naevus flammeus often follows the distribution of cutaneous nerves. Naevus flammeus may also be a manifestation of a series of syndromes in which internal organs, besides the skin, are involved. Cobb syndrome is characterized by a naevus flammeus or another vascular lesion in a dermatomal distribution on the trunk or limb associated to angioma in the spinal canal at the same segmental level as that of the cutaneous angioma (1).

CASE REPORT

A 29-year-old male patient was first seen in our Department in March 1994, with a 10-year history of pink macules on the left side of the chest and back. The patient had a history of a cerebral arteriovenous malformation involving the left middle cerebral artery at the temporo-occipital area, which caused intracerebral haemorrhage. It had been excised when the patient was 18 years old, and a bone cyst in the distal segment of the right humerus had been excised when he was 20 years old. Since excision of the cerebral arteriovenous malformation, the patient had been treated with anticonvulsant drugs until he was 25 years old, when complete discontinuation of all medication was achieved with no neurologic manifestations or sequelae.

Physical examination revealed an area of erythema distributed unilaterally and in dermatomal fashion, involving the left chest from the pectoral area to the ipsilateral scapular region (Fig. 1). A close-up view demonstrated that the lesion consisted of numerous grouped, fine, threadlike superficial telangiectases. The pink colour, however, faded only slightly upon pressure.

A cutaneous biopsy was performed. Histopathologic examination demonstrated that the main changes consisted of increased, discrete, jagged or widely dilated blood vessels, most of them situated in the upper part of the reticular dermis, but some of them positioned throughout the entire reticular dermis. These blood vessels were thin-walled and endothelium-lined.

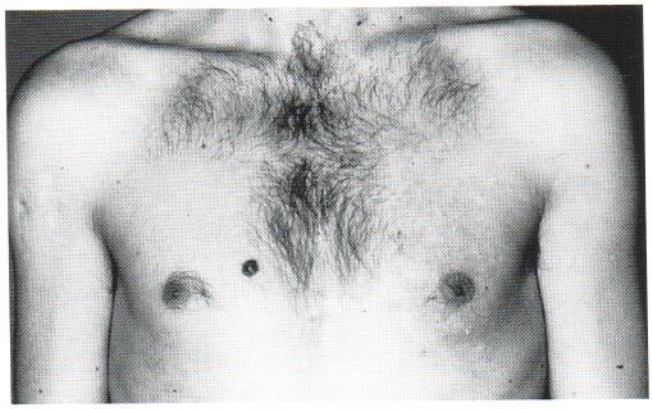

Fig. 1. Zosteriform naevus flammeus involving the left side of chest.

Brain magnetic resonance findings only showed residual lesions of the previously excised arteriovenous malformation, but this exploration failed to demonstrate any abnormality in the spinal canal.

DISCUSSION

According to Kissel & Dureux, in their comprehensive review of the Cobb syndrome (2), two criteria are necessary and sufficient to establish a diagnosis of cutaneo-meningospinal angiomatosis or Cobb syndrome, namely, (1) a vascular skin naevus, and (2) an angioma in the spinal canal. The size and appearance of the cutaneous lesion may vary, but its segmental level must correspond, within a segment or two, to that of the spinal angioma. The spinal angioma likewise may vary in appearance, nature, and extent but it must be confirmed anatomically, surgically, or, if need be, radiologically.

Our patient did not fulfill the criteria of the Cobb syndrome, since involvement of the central nervous system by vascular malformation was not located at the same segmental level as that of cutaneous naevus flammeus. Nevertheless, in our patient, zosteriform naevus flammeus involving the left side of the chest was associated to an arteriovenous malformation on the left middle cerebral artery. So, we think that zosteriform naevus flammeus may be a cutaneous marker of cerebral angiomatosis. The presence of the bone cyst in the right humerus seems to be coincidental, since no other features of Maffuci's syndrome were present.

Wyburn-Mason (3) reported 4 patients with venous haemangiomas involving the spinal cord at different levels, whereas cutaneous angiomas (described as "vascular skin naevus" "telangiectatic naevus" "port-wine marks" and "vascular naevus") were located at the sole, occipital region, arm, and nape of the neck, and in all these patients skin lesions were unrelated segmentally to the spinal angioma. Hurt (4) described a patient with intradural arteriovenous haemangioma at level Th4 and Th5 and several cutaneous haemangiomas in the lumbar region and lower limbs. Furthermore, in other reported cases (3, 5) the skin haemangioma accompanying an intraspinal angioma was so diffuse, being situated not only on the dermatome corresponding to the cord lesion but on other dermatomes as well, that metameric correspondence could not be proved. Such cases do not satisfy criteria of Cobb syndrome. In any event, in all reported cases in which cutaneous vascular abnormality was distributed in metameric fashion, an associated cerebral or spinal vascular malformation was present as well. The cutaneous vascular abnormality may show a different clinical morphology and it could be preceded, appear simultaneously, or develop later than neurologic angiomatosis. We think that acquired zosteriform naevus flammeus can be a cutaneous marker of central nervous system angiomatosis, and that in all patients with a cutaneous haemangioma showing metameric distribution, involvement of the central nervous system by vascular malformations should be suitably ruled out.

REFERENCES

1. Jessen RT, Thompson S, Smith EB. Cobb syndrome. Arch Dermatol 1977; 113: 1587–1590.
2. Kissel P, Dureux JB. Cobb syndrome. Cutaneo-meningospinal angiomatosis. In: Bruyn GW, Vinken PJ, eds: Handbook of clinical neurology. New York: North Holland Publishing Co, 1972: 429–445.
3. Wyburn-Mason R. The vascular abnormalities and tumours of the spinal cord and its membranes, London: H. Kimton, 1943.
4. Hurt H. Les anéurysmes artério-veineux de la moëlle épinère. Considérations anatomo-cliniques et thérapeutiques. A propos de 11 cas étudiés par artériographie. Thèse Med. R. Foulon, Paris, 1964: 104.
5. Gilbert L. Angioma venosum racemosum with angiomatous lesions on the skin and omentum. BMJ 1952; 1: 468–470.

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