Multiple, neonatal, self-healing, cutaneous glomuvenous malformations

Sir,

Glomuvenous malformations, formerly known as glomangiomas, arise from the neuromyoarterial elements of the glomus body and are histologically characterized by the presence of modified smooth muscle cells (glomus cells).[1-3] These benign tumors are identified by the presence of large and irregular vascular spaces covered by a thin coat of endothelial cells, further surrounded by a layer of glomus cells with a prominent central nucleus.[2,4]

A 4-month-old girl was brought to our clinic with hard, purplish macular lesions on the back and on the upper and lower extremities, all of which had appeared progressively since birth [Figures 1 and 2]. None of the patient's relatives had similar lesions. A biopsy was taken from one of the macules. Histological examination revealed irregular, dilated vascular channels surrounded by cuboidal cells with a central nucleus and eosinophilic cytoplasm (identified as glomus cells) [Figure 3]. A diagnosis of multiple cutaneous glomangiomas was made. It was decided to keep the patient under observation. At the next consultation, 3 months later, physical examination revealed that only two glomangiomas remained. The parents asserted that the child had been subjected to no procedure nor had she undergone any treatment. One year later, a complete skin examination revealed all the lesions to have disappeared [Figure 4].

Glomuvenous malformations appear at an early age as red-bluish papules or nodules; they are generally painless although some may be painful when palpated.[2] They can be classified into three variants: single glomus tumors (90% of all cases), multiple glomus tumors and jugular glomus tumors.[2,4] Jugular glomus tumors are spherical conglomerations of cells and small blood vessels; they are therefore known as glomera.[5] An extremely rare congenital variant is also known that appears as papules which coalesce to form purplish plaques; these have been shown to grow with development.[2,4]

Multiple glomuvenous malformations account for only 10% of all glomus tumors.[2] When they do appear, a family background is common (60% of cases); an autosomal dominant inheritance pattern with incomplete penetration has been described (70% at 5 years old; 100% at 30 years old).[1,4] Glomuvenous malformations are associated with a mutation in the glomulin gene located on the 1p21-22 chromosome, the locus of which has been named VMGLOM.[1,6] Congenital lesions are even rarer, with fewer than 20 cases described.[4] This is associated with an early postzygotic

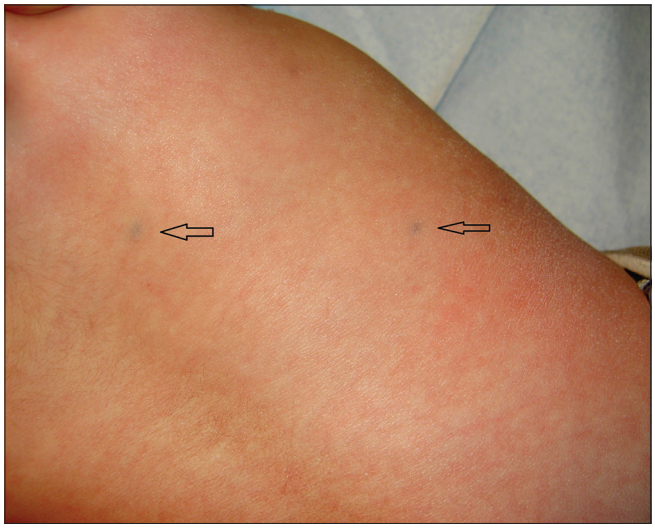

Figure 1: Two glomangiomas on the back (arrows)

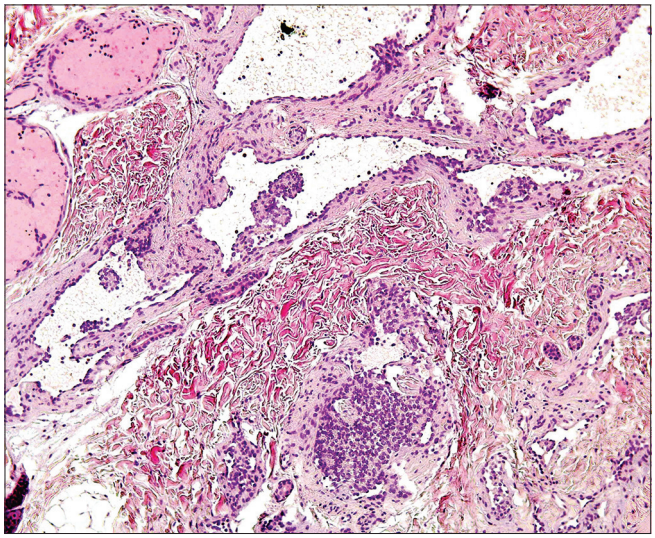

Figure 3: Dilated vascular channels surrounded by glomus cells (H and E, ×400)

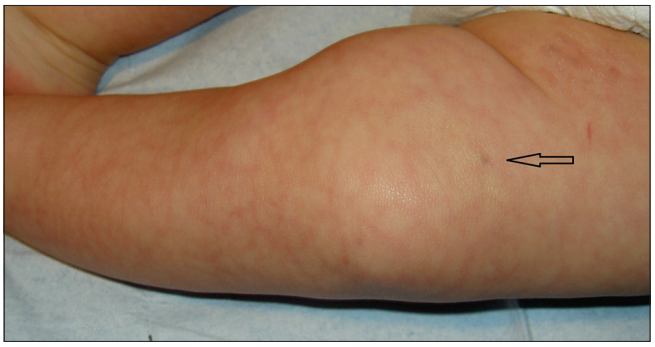

Figure 2: Blue lesion on the thigh (arrow)

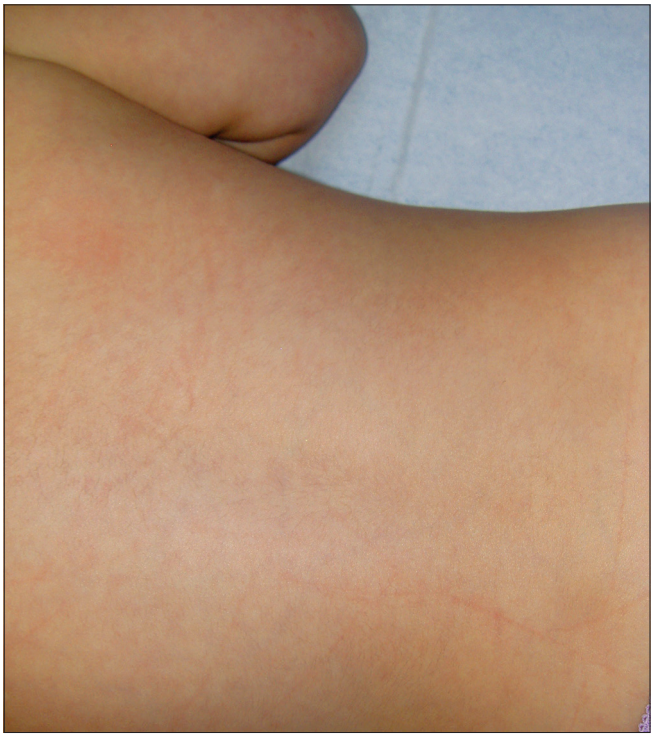

Figure 4: The back of the patient showing resolution of glomangiomas, one year later

mutation (type 2 mosaic) originating from a loss of heterozygosity affecting a larger anatomical region.[4] Its association with type 1 neurofibromatosis has recently been discovered.[3] Differential diagnosis should include blue rubber bleb nevus syndrome; this presents with similar tumors (although with no glomus cells) and intestinal involvement.[2]

The present case represents an uncommon manifestation of glomangiomas since the patient had neither the plaques typical of the congenital type nor any associated family history. In addition, her tumors disappeared spontaneously. We were unable to find any similar previous reports. Indeed, one case of localized, partial regression of a plaque lesion which underwent several years of gradual involution has been described.[7] Segmental patterns of glomus tumors with unilateral, patchy presentations have also been reported in the literature. In those patients, new lesions appeared in their lifetime with no disappearance of the existing lesions.[8] Although rare, spontaneous remission has been reported in segmental genetic skin diseases such as epidermolysis bullosa or Kindler syndrome, each time involving changes in DNA correcting the germ line mutations.[9,10] Although the present patient did not have segmental disease, reverting mutations may have played a role in inducing the spontaneous resolution.

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Conflicts of interest
There are no conflicts of interest.

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