Bluish nodules of the breast in an adolescent girl

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DESCRIPTION

A 15-year-old girl was referred due to skin lesions of the right breast, which had first been noticed shortly after birth and had enlarged slowly with the patient’s growth but more rapidly in the past few years. Family history disclosed that the patient’s father had similar lesions on the left shoulder. Physical examination revealed multiple bluish coalescent soft nodules on the right breast, which were slightly tender to palpation (figure 1). Histological examination demonstrated dilated irregular vascular channels surrounded by endothelial cells and one to several layers of uniform cuboidal cells with a pale or faintly eosinophilic cytoplasm (figure 2). These features were consistent with the diagnosis of glomuvenous malformation (GVM).

GVM, previously known as glomangioma or multiple glomus tumour, is a vascular malformation that accounts for 5% of all venous-type anomalies.1 It is frequently inherited (63.8%) and usually present at birth with a slow expansion during childhood. Lesions are nodular and scattered or plaque-like and segmental, with colour varying from pink to purplish dark blue, with a distinct raised, often hyperkeratotic cobblestone-like appearance. They are not compressible and are typically painful by compression. GVMs involve skin and subcutis, with no extracutaneous involvement, and are mainly located on the extremities but may also involve the trunk.1,2 Accurate diagnosis of GVM and distinction from other venous malformations (VM) can be challenging. Histology makes the definite diagnosis, revealing the presence of undifferentiated smooth muscle cells (glomus cells) surrounding convoluted venous channels, but history and physical findings help early recognition.3

Distinguishing GVM from other VM is crucial for the management of the patients, as the treatment differs. In fact, elastic compressive garments, which often decrease discomfort associated with common VM in a limb, can aggravate pain in GVM and so are contraindicated in these patients. Small GVM can also be surgically excised because they are more superficial and invade less the adjacent structures. In contrast, complete surgical excision of VMs is rarely possible because they permeate surrounding tissues and often involve deep structures. Sclerotherapy has been found to be less effective in GVMs than in VM. A small number of isolated cases have been successfully treated with ablative laser therapy. Asymptomatic lesions can just be kept under observation, since most of the patients never experience any related medical problems.4

The authors intend to emphasise that awareness of GVMs especially during childhood and adolescence, most often with a positive family history, help to establish the correct diagnosis and consequently a properly management.

Figure 1 Clinical presentation. Multiple well-defined bluish to purple coalescent soft and slightly tender nodules on the right breast.

Figure 2 Histopathological findings. Dilated capillaries containing erythrocytes. Layers of glomus cells surrounding capillaries (H&E, original magnification ×100).

Learning points

- Distinguishing glomuvenous malformation (GVM) from other venous malformations is crucial for the management of the patients.
- Awareness of GVMs in special during childhood and adolescence, most often with a positive family history, help to establish the correct diagnosis.

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