CASE REPORT

Congenital nodular multiple glomangioma: a case report

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A 13 year old girl presented with recurrent painful ''varicosities'' on her right calf. These lesions were subsequently clinically diagnosed as ''cavernous haemangiomas'' after normal duplex scanning and were excised. Histological examination revealed multiple glomangiomas (glomus tumours). A literature review revealed only two reported cases of nodular multiple glomangioma, so that this is the third case to be reported in the literature.

Glomus tumours are benign tumours of the perivasculature. They arise from modified smooth muscle cells, called glomus cells, located in the walls of the Sucquet-Hoyer canal, a specialised arteriovenous anastomosis central to thermoregulation.[1] Most tumours are less than 1 cm in diameter, occurring in the dermis or subcutis in the upper and lower extremities.[1] There are two forms of glomus tumour, with the more common solitary variant accounting for most of the cases (90%), and a rarer multiple variant accounting for 10% of cases[2]; this second form is seen most often in children and is thought to be inherited in an autosomal dominant fashion.[1] The multiple variant is subdivided clinically into nodular and plaque-like lesions. Histologically, glomus tumour is composed of varying proportions of glomus cells, blood vessels, and smooth muscle.[1] Depending on the predominant component, there are three variants of glomus tumour, namely: (1) angiomatoid (glomangioma) with predominant blood vessels; (2) solid (predominantly glomus cells); and (3) glomangiomyoma (predominantly smooth muscle). Multiple glomus tumours generally correspond to glomangioma.

''A literature review revealed only two reported cases of this rare variant''

In 2001, Carvalho *et al* reported a congenital plaque-like variant and reviewed 14 other cases. Of these 15 cases, 13 were described as the plaque-like type and two were nodular.[2] In 2001, an additional case of congenital plaque-like multiple glomus tumour was reported by Lin *et al*.[3]

We report a case of congenital multiple glomangioma in a 13 year old girl, with the nodular variant. A literature review revealed only two reported cases of this rare variant.

CASE REPORT

A 13 year old girl was seen in the paediatric plastic surgery outpatient clinic in Birmingham Children's Hospital, UK, after being referred for painful varicosities on her right calf, recurring after three previous phlebectomies (histological examination was not performed). These bluish ''venous'' lesions had been present since birth. The patient was experiencing pain at the site of the lesions. On standing the lesions would dilate. After each excision the lesions reappeared. The patient's mother, grandmother, great grandmother, and great aunt each had similar lesions at different anatomical locations. Although there was no histological diagnosis for these lesions, the clinical picture supports an autosomal dominant inheritance.

On clinical examination, there appeared to be varicosities along the long saphenous axis in her right calf (fig 1). Duplex scanning showed normal superficial and deep veins in the right leg. At this point it was assumed that the lesions were in fact scattered cavernous haemangiomata. Excision of the lesions was carried out by the vascular surgery department. At surgery, there appeared to be multiple subcutaneous haemangiomata, ''like isolated knotted varicose veins'', which were excised with skin ellipses and subcutaneous tissue from the right upper part of the thigh, popliteal fossa, and medial calf. Eight lesions were excised in total.

PATHOLOGICAL FINDINGS

Macroscopic examination showed 10 pieces of fibro-fatty tissue, the largest of which measured 2.5 × 2.5 × 2 cm. The smallest measured 1.3 × 0.7 × 0.4 cm. Most of them showed bluish vessels. Microscopy revealed multiple diffuse lesions, present both within the dermis and deep within the subcutaneous tissue fragments, and unlikely to be excised completely. These lesions were formed of dilated vessels, with a surrounding collar of regular round cells with round nuclei

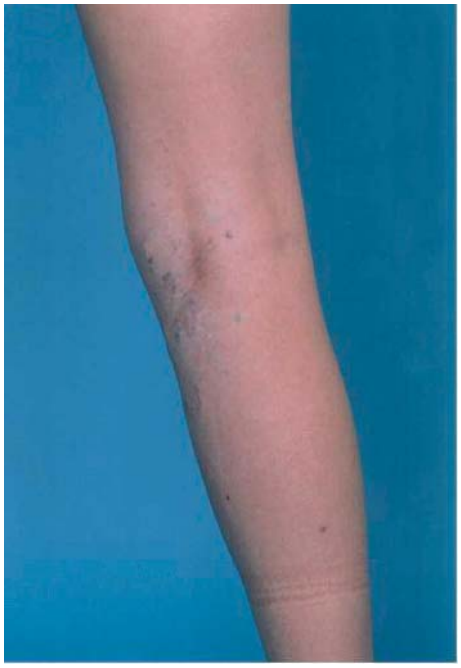

Figure 1 The patient's right calf, showing varicosities along the long saphenous axis. This photograph is reproduced with the full consent of the patient.

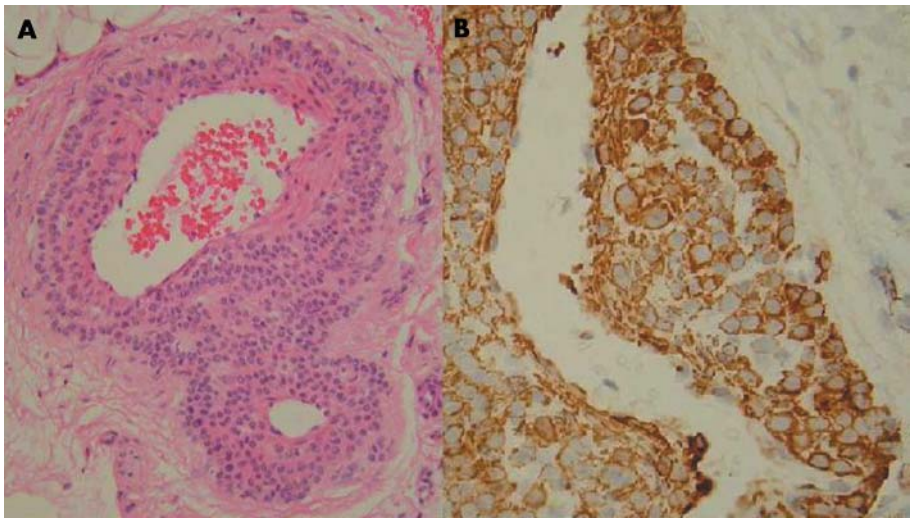

Figure 2 (A) Low power view of a lesion formed from dilated vessels, with a surrounding collar of regular round cells with round nuclei and eosinophilic cytoplasm (haematoxylin and eosin stain). (B) The tumour cells were positive for smooth muscle actin (immunohistochemical stain).

and eosinophilic cytoplasm (fig 2A). There was no mitotic activity or atypia. These features were consistent with the angiomatoid variant of glomus tumour (glomangioma). The presence of the tumour within the subcutaneous tissue is in keeping with what has been described as ''infiltrating glomus tumour'', which implies difficulty in complete excision and the possibility of recurrence.

The tumour cells were immunoreactive for smooth muscle actin (fig 2B). The periodic acid Schiff stain distinctively highlighted the cytoplasmic membrane of the tumour cells. These immunohistochemical and special stains added support to the diagnosis.

DISCUSSION

Glomangioma has an early onset, with one third of cases presenting before 20 years of age.[4] Familial cases have been reported with autosomal dominant transmission, incomplete penetrance, and variable expression. In 2000, Pena-Penabad *et al* reported two cases of familial multiple glomangiomas, but the lesions were not present from birth in the first patient and the second patient presented for consultation as an elderly adult.[5] There have been several papers describing glomangiomas in the knee region,[6–9] but all four papers describe a solitary tumour, which is different from the multiple type reported here. Clinically, glomangiomas appear as red to blue compressible papulonodules.[2] Although solitary glomus tumours often produce pain, multiple glomangioma usually does not. In our case, the patient presented with pain. In most cases there are fewer than 10 lesions. Of the 15 cases of congenital glomangioma presenting in childhood found in the literature, 13 were described as the plaque type and two as nodular. Our patient presented in childhood with painful, bluish nodules in her right lower extremity, fitting in with the nodular multiple variant.

This is the third case in the literature of congenital multiple glomangioma, nodular type, presenting in childhood. The presence of the tumour within the subcutaneous tissue explains why it recurred.

Take home messages

- We describe a rare case of congenital multiple glomangioma, nodular type, presenting in childhood—only the third to be reported
- The tumour was present within the subcutaneous tissue, causing it to recur

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The patient gave full consent for the reproduction of the photograph.

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