specific damage to the elastic fibres with calcium deposition is unknown. Moreover, the specific genetic biochemical defect that allows for the calcification of elastic fibres is also unknown (11).

REFERENCES

1. Neldner KH. Pseudoxanthoma elasticum. Clin Dermatol 1993; 1: 13.
2. Christensen OB. An exogenous variety of pseudoxanthoma elasticum in old farmers. Acta Derm Venereol (Stockh) 1978; 58: 319–321.
3. Otkjaer Nielsen A, Christensen OB, Hentzer B, Johnson E, Kobayasi T. Salpeter-induced dermal changes electron-microscopically indistinguishable from pseudoxanthoma elasticum. Acta Derm Venereol (Stockh) 1978; 58: 323–327.
4. Dupre A, Bonafe JL, Christol B. Chequered localized pseudo-xanthoma elasticum: a variety of Christensen's exogenous pseudo-xanthoma elasticum? Acta Derm Venereol (Stockh) 1979; 59: 539–541.
5. Rongioletti F, Rebora A. Pseudoxanthoma elasticum-like papillary dermal elastolysis. J Am Acad Dermatol 1992; 26: 648–650.
6. Oppenheim M. Hautveraenderungen durch Chlorkalzium bei der Herstellung der Sogennanten Eislutscher. Wien Klin Wschr 1935; 48: 207–209.
7. Sneddon IB, Archibald RM. Traumatic calcinosis of the skin. Br J Dermatol 1958; 70: 211–214.
8. Wheeland RG, Roundtree JM. Calcinosis cutis resulting from percutaneous penetration and deposition of calcium. J Am Acad Dermatol 1985; 12: 172–175.
9. Clendenning WE, Auerbach R. Traumatic calcium deposition in skin. Arch Dermatol 1964; 89: 360–363.
10. Schoenfeld RJ, Grekin JN, Mehregan A. Calcium deposition in the skin: a report of four cases following electroencephalography. Neurology 1965; 15: 477–480.
11. Walsh JS, Failley JA. Calcifying disorders of the skin. J Am Acad Dermatol 1995; 33: 693–706.

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I. Neri, S. Marzaduri, F. Bardazzi and A. Patrizi
Department of Dermatology, University of Bologna, Via Massarenti 1, I-40138 Bologna, Italy.

Congenital Multiple Annular Glomus Tumors

Sir,

The glomus is a specialized arteriovenous shunt, involved in thermal regulation. It is made up of an efferent arteriole, which extends into thick-walled arteriolar segments called Suquet-Hoyer canals, and a venule. Glomus cells are found in layers in the walls of the Suquet-Hoyer canals – the site of A-V anastomosis. Glomus bodies are normally concentrated in fingertips and nail beds. Glomus tumors, first described by Masson (1) in 1924, are rare benign proliferations of glomus bodies. We describe the first case of congenital multiple glomus tumors arranged in an annular pattern and review the literature on congenital multiple glomus tumors.

CASE REPORT

A 3-year-old Hispanic male was evaluated for multiple asymptomatic skin lesions, present since birth, which were slowly increasing in size and number. Past medical history was negative, as was family history of similar lesions. Physical examination revealed multiple bluish-purple macules, 2–3 mm in diameter, in an annular configuration on the right upper (Fig. 1) and lower back and buttock. In addition, small individual bluish macules were scattered bilaterally across the trunk and shoulders, with a total of about 20 lesions. The macules were non-tender and did not blanch on diascopy. Laboratory values, including a CBC, SMA-20, and PT/PTT, were normal. A biopsy of an annular lesion revealed within the superficial and deep dermis, extending to the deep margin of the specimen, irregularly shaped interconnected cystic spaces that in areas had jagged outlines and dissected in between collagen bundles (Fig. 2a). These spaces were empty and were lined by cuboidal cells having round, small and uniform nuclei. The cells were arranged as a single row or multiple layers. In some areas one could appreciate widely spaced slender spindle-shaped cells on top of the cuboidal cells, separating these cells from cystic lumina (Fig. 2b). Immunohistochemical studies demonstrated the cuboidal cells to be vimentin-positive, desmin-negative. These findings were diagnostic of multiple glomus tumors.

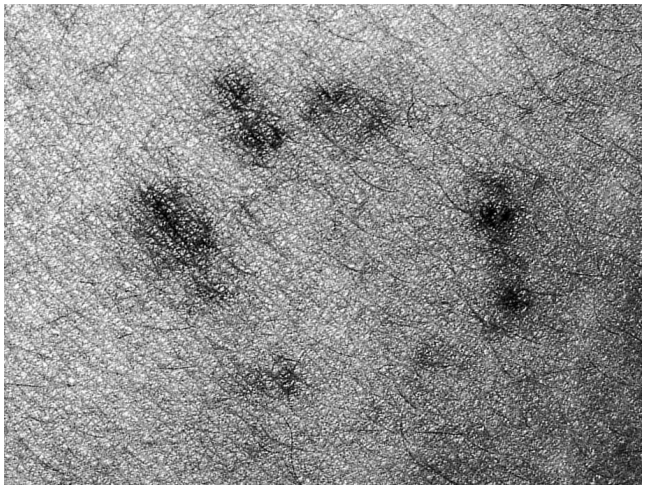

Fig. 1. Annular glomus tumors on the back.

DISCUSSION

Glomus tumors, also known as glomangiomas, are uncommon neoplasms that occur as a solitary tumor or as the rare multiple type. Solitary glomus tumors are usually small bluish painful nodules, situated in the nail bed or on an extremity. The pain can be exacerbated by changes in temperature, pressure and trauma. Subungual solitary tumors are more common in females and develop at an average age of 40, whereas solitary tumors at other sites occur equally in both sexes and occur earlier, at an average age of 25 (2).

In 1937, Weidman & Wise (3) described multiple glomus tumors. Unlike the solitary type, they are often inherited as an autosomal dominant trait and are more common in childhood, with a male predominance. They are generally asymptomatic and not situated in the nail bed. Multiple glomus tumors

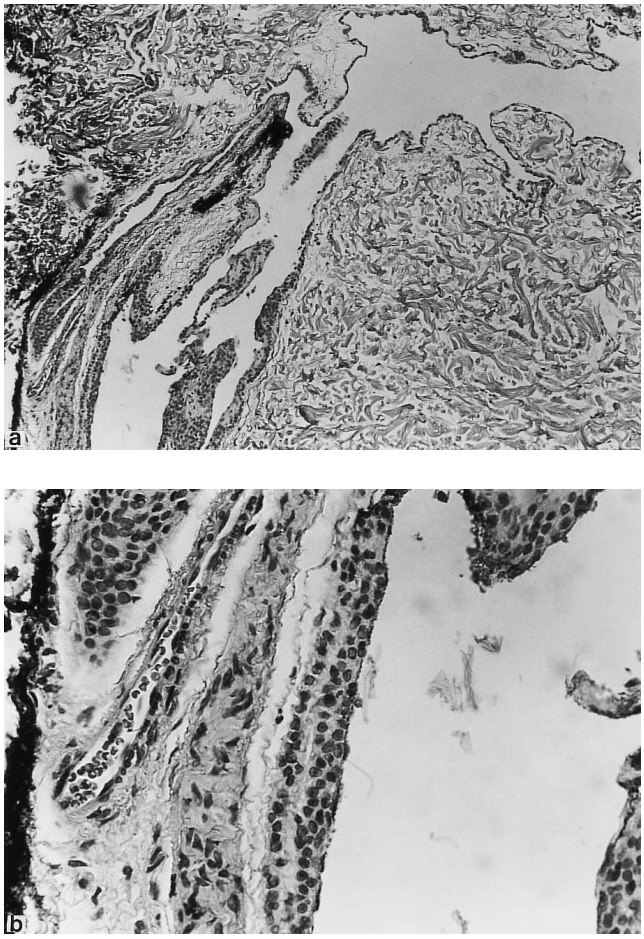

Fig. 2. Irregular cystic spaces lined by cuboidal cells (*a*) and multiple layers of cuboidal glomus cells (*b*).

have been subdivided into regional, disseminated, and plaque-like types (4, 5). Multiple glomus tumors vary from small discrete bluish macules and papules to large nodules and plaques measuring a few centimeters in diameter. The number of tumors is quite variable, ranging in the majority of cases from less than 10 to as many as 400 (6). The age of onset of multiple glomus tumors varies from birth through adolescence, with congenital tumors being extremely rare. Kohout & Purdy-Stout (4) in 1961 reviewed 731 cases of all types of glomus tumors in children and found 6 that were congenital and only one that was congenital and multiple. In 1990 Landthaler et al. (5), in their report of congenital multiple glomus tumors, only described their 2 patients with large plaques and nodules and Leu's patient (7) who had just nodules.

In a review of the literature, we documented only 14 cases of multiple congenital tumors (4, 5, 7). The lesions occur primarily as a few isolated bluish nodules and plaques and rarely as clusters of multiple small papules and macules as in our patient. The tumors occur predominantly on the trunk and occasionally on the face and extremities. There is a male predominance and a positive family history is not always present.

The clinical course of multiple glomus tumors is usually benign. McEvoy et al. (6), however, reported that patients with diffuse or generalized distribution of lesions may show evidence of the Kasabach–Merritt syndrome.

Histologically, solitary glomus tumors are well circumscribed and surrounded by a fibrous capsule. A single layer of flattened endothelial cells lines narrowed vascular lumina. Multiple layers of glomus cells are found peripheral to the endothelial cells (2). In contrast, multiple glomus tumors have large irregularly shaped vascular spaces and no fibrous capsule. A single layer of flattened endothelial cells lines the vascular spaces, and the glomus cells at the periphery are only 1 to 3 cell layers thick or even absent. Immunohistologic studies suggest that glomus cells are similar to vascular smooth muscle, since both are vimentin-positive and desmin-negative.

Previously the primary treatment for glomus tumors has been surgical excision of painful lesions and reassurance and observation of those that are asymptomatic (4). Newer treatment modalities include the CO_2 and argon laser (8), electron beam irradiation, and sclerotherapy using hypertonic saline (9) and sodium tetradecyl sulfate (10).

REFERENCES

1. Masson P. Le glomus neuromyoarterielles regions tactiles et ses tumeurs. Lyon Chirurgical 1924; 21: 257–280.
2. Tsuneyoshi M, Enjoji M. Glomus tumor – a clinicopathologic and electron microscopic study. Cancer 1982; 50: 1601–1607.
3. Weidman FD, Wise F. Multiple glomus tumors of the order of telangiectases. Arch Dermatol 1937; 35: 414–426.
4. Kohout E, Purdy-Stout A. The glomus tumor in children. Cancer 1961; 14: 555–565.
5. Landthaler M, Braun-Falco O, Eckert F, Stolz W, Dorn M, Wolff HH. Congenital multiple plaque-like glomus tumors. Arch Dermatol 1990; 126: 1203–1207.
6. McEvoy BF, Waldman PM, Tye MJ. Multiple hamartomous glomus tumors of the skin. Arch Dermatol 1971; 104: 188–191.
7. Leu HJ. Multiple kongenitale Glomustmoren. Morphol Med 1982; 2: 149–156.
8. Barnes L, Estes S. Laser treatment of hereditary multiple glomus tumors. J Dermatol Surg Oncol 1986; 12: 912–915.
9. Siele RJ, Spencer DM, Davis LS. Hypertonic saline destruction of multiple glomus tumors. J Dermatol Surg Oncol 1994; 20: 347–348.
10. Gould EP. Sclerotherapy for multiple glomangiomata. J Dermatol Surg Oncol 1991; 17: 351–352.

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Tamara Moss, Philip C. Don, Jeffrey M. Weinberg, Soren M. White, Mark Kaufmann and Wojciech Szianiawski
Department of Dermatology, New York Medical College-Metropolitan Hospital Center, 1901 First Avenue, New York, New York 10029, U.S.A.