Glomus tumor of the hip

An unusual location

Sibel Hakverdi, *MD,* ***Aydiner Kalaci,*** *MD,* ***Mehmet Yaldiz,*** *MD,* ***Bülent Akansu,*** *MD.*

ABSTRACT

تعتبر الأورام الدبقية في العادة أورام حميدة وشاذة، التي تظهر في الأدمة أو في الأنسجة تحت الجلد للأطراف. ونادراً ما تظهر في الورك. نستعرض في هذا تقرير حالة مريض يعاني من آلام شديدة وتألم باللمس في الورك الأيسر، خاصة عند الجس وعند موضع الجلوس. خلال الفحص كان هناك كتلة طرية قابلة للجس تحت الجلد، وتألم شديد عند اللمس في الورك الأيسر. أظهرت نتيجة الأشعة بالموجات فوق الصوتية وجود كتلة كثيرة الأوعية الدموية تحت الأدمة يبلغ قطرها cm1.2. تم استئصالها بالكامل تحت التخدير الموضعي. كان التشخيص المرضي النسيجي "ورم دبقي". أصبح المريض خالٍ من الأعراض لمدة ثلاثة أشهر من المتابعة الطبية.

Glomus tumors are usually benign neoplasms that occur in the dermis or subcutaneous tissues of the extremities. They occur very rarely in the hip. We report a patient with severe pain and tenderness in the left hip, especially on palpation, and in the sitting position. On physical examination, there was a soft palpable subcutaneous mass and severe tenderness in the left hip. Ultrasound revealed a hypervascular subdermal mass that was 1.2 cm in diameter, which was subsequently totally excised under local anesthesia. The histopathologic diagnosis was a 'glomus tumor'. The patient has been symptom-free for 3 months of follow-up.

Saudi Med J 2009; Vol. 30 (2): 299-301

From the Departments of Pathology (Hakverdi, Yaldiz, Akansu), and Orthopedic Surgery (Kalaci), Faculty of Medicine, Mustafa Kemal University, Antakya, Turkey.

Received 3rd September 2008. Accepted 25th January 2009.

Address correspondence and reprint request to: Dr. Sibel Hakverdi, Bahçelievler Sitesi Kardelen Sok. No:3, 31030 Antakya, Hatay, Turkey. Tel. +90 (326) 2415515. Fax. +90 (326) 2217618. E-mail: hakverdisibel@yahoo.com

Glomus bodies are special arterio-venous anastomoses located in the reticular dermis, and they play an important role in thermoregulation. The wall of these arterio-venous anastomoses, also named Sucquet-Hoyer canals, are composed of endothelial cells and surrounded by multiple layers of uniform glomus cells. Accounting for their contractility, they are thought to be modified smooth muscle cells. Therefore, they can react to thermal changes and regulate blood flow to the skin. Glomus bodies can be found anywhere else in the body, but are mostly concentrated in the extremities especially, digits, palms, and the soles of the feet.[1,2] Glomus tumors are rare neoplasms originating from these special smooth muscle cells. Nowadays, they are accepted as real neoplasms instead of being hamartomatous or hyperplastic lesions. The glomus tumor, which is a neoplasm of the glomus apparatus, was originally described by Masson in 1924.[2,3] These lesions are grossly bluish-gray in color and painful. Pain, pinpoint tenderness, and hypersensitivity to thermal changes are major symptoms. In this report, we discuss an unusual case of subcutaneous glomus tumor of the hip in an older patient in order to highlight the clinical and histopathological features.

Case Report. A 52-year-old male was admitted to our hospital with the complaint of pain in his left hip. He reported that he had suffered from this pain for 25 years and had visited different physicians several times. He stated that his pain had worsened over the last 10 days.[4] He also added that, he was uncomfortable while sitting because of pain, and felt tenderness in this region with cold exposure. On ultrasonography and MRI, a well-demarcated, hypoechoic, and solid lesion, 1.2 cm in diameter located in the subcutaneous area, surrounded by edema was reported. Total excision with surrounding normal tissue was performed under local anesthesia by surgeons, and the extracted tissue was sent for histopathological examination. Grossly, it was

a small tissue section with a gray-bluish lesion in the subcutaneous area. On microscopic examination, the tumor consisted of round to oval cells with a uniform appearance, and these cells formed multiple layers around the vascular structures (**Figure 1**). Neoplastic cells had well-defined cell borders with densely eosinophilic cytoplasm (**Figure 2**). There was no evidence of malignancy potential. Immunohistochemical studies performed with vimentin were positive (**Figure 3**), whereas smooth muscle actin, desmin, and epithelial membrane antigen (EMA) were negative. Due to these findings, the lesion was diagnosed as 'glomus tumor'. He has been symptom free for 3 months of follow-up.

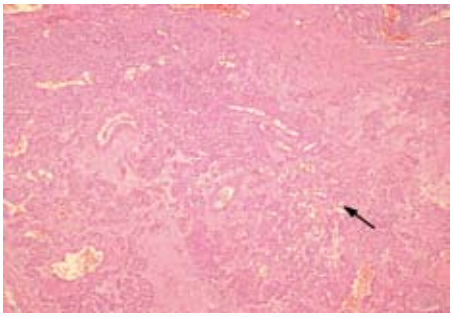

Figure 1 - The neoplasm is composed of a trabeculated network of vessels (arrow) surrounded by bland, small tumor cells (Hematoxylin-Eosin, original magnification x 100).

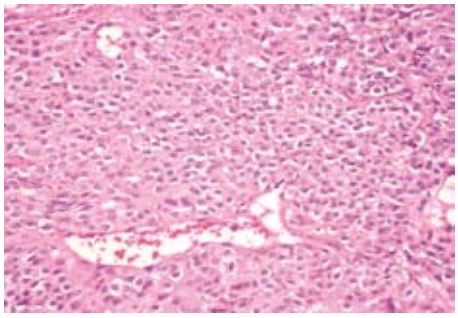

Figure 2 - The neoplastic cells have sharply outlined round-to-oval nuclei with bland chromatin and scant-to-moderate eosinophilic cytoplasm (Hematoxylin-Eosin, original magnification x 400).

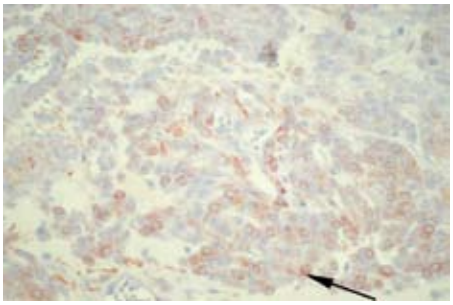

Figure 3 - Immunostaining for vimentin that shows scattered tumor cells with cytoplasmic positivity (arrow) (Vimentin, original magnification x 400).

Discussion. Glomus tumors are very uncommon lesions occurring most frequently in the extremities, especially the subungual area of the digits.[1,2] However, they were also reported in atypical locations where they do not exist physiologically, such as kidney, nasal cavity, stomach, esophagus, trachea, lung, pancreas, ovary, and vagina.[1] Scheifer et al[2] documented extra digital glomus tumors that were diagnosed over a 20-year period, and showed that most of these lesions were located in the extremities, as well as one in the buttock, as in our patient. In the literature as we know, this location for a glomus tumor is very rare.[4,5] Clinically, a glomus tumor is equally common in males and females, but there is female predominance in subungual lesions as 3:1.[1] In one series,[2] pain, hypersensitivity to cold exposure, and tenderness to blunt pressure were detected as the most common symptoms, and the duration of these complaints ranged from 7-11 years.[2] In another study, it was reported that the patient had suffered from pain for 40 years.[4] Similarly, these 3 major symptoms and chronic duration of 25 years were present in our patient.

Histopathologically, the glomus tumor can include vascular structures like hemangiopericytoma or paraganglioma, as well as being more cellular and solid.[1] Glomus tumors can be classified as solid glomus tumor, glomangioma, glomangiomyoma, and glomangiosarcoma.[6] Folpe et al[7] attempted to define the criteria of atypic and potentially malignant ones based on cytological and histological features. These features included deep location, size larger than 2 cm, atypical mitotic figures, and marked atypia with mitotic activity. None of these findings were present in our case. Instead of being thought as rarely metastasizing, there is a case report of a highly aggressive tumor with distant metastases, which originated from the gluteal region.[8] Cutaneous adnexal tumors such as eccrine spiradenoma and solid forms of hidradenoma, hemangioma, and hemangiopericytoma should be kept in mind in the differential diagnosis. None of these contain glomus cells, and based on the distinctive architecture and microscopic morphology, can therefore be excluded on histological grounds alone. If intradermal nevus or melanocytic nevus is suspected, S-100 protein and HMB-45 can help in the diagnosis. However, some metastatic carcinomas can be seen as similar to glomus tumors in hematoxylin-eosin slides. Pleomorphic adenoma of the cellular type is also included in the differential diagnosis. A total lack of ductal differentiation, the absence of cytokeratins and epithelial membrane antigen, and the presence of vimentin on immunostaining exclude pleomorphic

adenoma, which is a mixed tumor with a high content of myoepithelial cells.[1,9] For the differential diagnosis, immunohistochemical techniques are very important, as well as history and histopathological examination. In many studies, smooth muscle actin and vimentin were stained positive, whereas cytokeratin, desmin, myoglobin, S-100 protein, neurofilaments, and factor VIII-related antigen were negative.[4,5,10] However, there are also studies on vimentin and actin negativity in some of the cases.[11,12] In our case, only vimentin was positive. Most of the glomus tumors were thought to be sporadic. However, some studies showed that there might be familial transition in some cases, especially, chromosome 1 and 11 were indicated, and heredity was thought to be as autosomal dominant and paternal.[13-18] Generally, total excision is enough for treatment, but very rarely recurrence is reported. In the presence of malignant features, a wide excision is needed with a close follow-up of the patient for regional or distant metastases.[1,2,6,18]

In conclusion, our study presents a glomus tumor located in an unusual location. Our case, and similar ones with an unusual location presented in literature, illustrates that the glomus tumor should be kept in mind in the differential diagnosis anywhere in the body, especially in terms of patients with complaints of pain, hypersensitivity to cold exposure, and tenderness to blunt pressure.

References

1. Weiss SW, Goldblum JR. Perivascular Tumors. In: Enzinger and Weiss's Soft Tissue Tumors. 4th ed. St. Louis (MO): Mosby Inc; 2001. p. 985-1001.
2. Schiefer TK, Parker WL, Anakwenze OA, Amadio PC, Inwards CY, Spinner RJ. Extradigital glomus tumors: a 20-year experience. ***Mayo Clin Proc*** 2006; 81: 1337-1344.
3. Mason P. Le glomus neuromyo-arterieldes regions tactil et ses tumeurs. ***Lyon Chir*** 1924; 21: 257-280.
4. Gencosmanoglu R, Inceoglu R, Kurtkaya-Yapicier O. Glomangioma of the hip. ***Dermatol Surg*** 2003; 29: 1244-1247.
5. McDonald JM, Moonka R. Extradigital glomus tumor as a cause of buttock pain. ***Orthopedics*** 2000; 23: 851-852.
6. Mentzel T, Hügel H, Kutzner H. CD34-positive glomus tumor: clinicopathologic and immunohistochemical analysis of six cases with myxoid stromal changes. ***J Cutan Pathol*** 2002; 29: 421-425.
7. Folpe AL, Fanburg-Smith JC, Miettinen M, Weiss SW. Atypical and malignant glomus tumors: analysis of 52 cases, with a proposal for the reclassification of glomus tumors. ***Am J Surg Pathol*** 2001; 25: 1-12.
8. Watanabe K, Sugino T, Saito A, Kusakabe T, Suzuki T. Glomangiosarcoma of the hip: report of a highly aggressive tumour with widespread distant metastases. ***Br J Dermatol*** 1998; 139: 1097-1101.
9. Graadt Van Roggen JF, Joekes EC, Welvaart K, Van Krieken JH. Unusual presentation of multiple subcutaneous glomus tumours of the lower limb with extensive glomus cell hyperplasia. ***Histopathology*** 1999; 34: 474-475.
10. Dervan PA, Tobbia IN, Casey M, O'Loughlin J, O'Brien M. Glomus tumours: an immunohistochemical profile of 11 cases. ***Histopathology*** 1989; 14: 483-491.
11. Bertalot G, Falchetti M, Parafioriti A. Glomus tumour: the immunohistochemical characteristics of twenty-three cases. ***Pathologica*** 1994; 86: 509-512.
12. Kessaris P, Klimis T, Zanakis S. Glomus tumour of the hard palate: case report and review. ***Br J Oral Maxillofac Surg*** 2001; 39: 478-479.
13. van der Mey AG, Maaswinkel-Mooy PD, Cornelisse CJ, Schmidt PH, van de Kamp JJ. Genomic imprinting in hereditary glomus tumours: evidence for new genetic theory. ***Lancet*** 1989; 2: 1291-1294.
14. Struycken PM, Cremers CW, Mariman EC, Joosten FB, Bleker RJ. Glomus tumours and genomic imprinting: influence of inheritance along the paternal or maternal line. ***Clin Otolaryngol Allied Sci*** 1997; 22: 71-76.
15. Calvert JT, Burns S, Riney TJ, Sahoo T, Orlow SJ, Nevin NC et al. Additional glomangioma families link to chromosome 1p: no evidence for genetic heterogeneity. ***Hum Hered*** 2001; 51: 180-182.
16. Brouillard P, Ghassibé M, Penington A, Boon LM, Dompmartin A, Temple IK, et al. Four common glomulin mutations cause two thirds of glomuvenous malformations ("familial glomangiomas"): evidence for a founder effect. ***J Med Genet*** 2005; 42: 13.
17. Kuru I, Oktar SO, Maralan G, Yaycioğlu S S, Bozan ME. [Familial glomus tumor encountered in the same finger and localization in four family members]. ***Acta Orthop Traumatol Turc*** 2005; 39: 365-368. Turkish.
18. Ozdemir O, Coşkunol E, Ozalp T, Ozaksar K. [Glomus tumors of the finger: A report on 60 cases]. ***Acta Orthop Traumatol Turc*** 2003; 37: 244-248. Turkish.