Glomus Tumor Masquerading for 22 Years as Osteoarthritis of the Hip

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Glomus tumors are rare benign mesenchymal neoplasms that account for less than 2% of soft tissue tumors. These neoplasms typically are small nodules less than 1 cm in diameter, associated with pain that is exacerbated by tactile stimulation and cold hypersensitivity.

We present a case of a large glomus tumor of the left lateral hip associated with a long history of severe pain of the left hip interfering with ambulation. Chronic pain as a result of a subcutaneous glomus tumor is rare and frequently misdiagnosed. In the case reported, a solid glomus tumor presented with 22 years of unilateral hip pain attributed to posttraumatic degenerative joint disease. Excision of a 4×3-cm nodule resulted in complete resolution of tenderness and joint pain. Subcutaneous glomus tumors can have unusually large size and location and should be considered in the differential diagnosis of chronic, atypical, or treatment-resistant joint pain.

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Glomus tumors are rare benign mesenchymal neoplasms that account for less than 2% of soft tissue tumors.[1] They originate from the modified smooth muscle cells of the neuromyoarterial plexus or glomus body, which is involved in thermoregulation.[2] The most common location of presentation is subungually on the hands; however, glomus tumors may arise in other soft tissue where glomus bodies are sparse or absent.[3] The classic diagnostic triad consists of lancinating pain, tenderness to tactile stimulation, and cold hypersensitivity.[4] Unusual presentations have been reported, including chronic pain syndromes and osteoarthritis.[5,6] While there have been previous reports of glomus tumors causing intractable pain, we report a case of a 4×3-cm hemorrhagic glomus tumor arising at a site of trauma 22 years prior to diagnosis, which was repeatedly misattributed to painful degenerative joint disease of the left hip.

Case Report

A 57-year-old man presented to the emergency department complaining of worsening foot pain of a year's duration and a gradual onset of inability to ambulate. He had severe psoriasis that was controlled with methotrexate (20 mg weekly administered intramuscularly) until he was lost to follow-up 2 months prior. He was admitted to the hospital with a diagnosis of bilateral lower extremity cellulitis and started on intravenous antibiotics.

On physical examination, the patient had generalized psoriasis over the trunk and extremities, with hyperkeratosis and fissuring of both soles. A 4×3-cm firm tumor was present over the left lateral hip (Figure 1), with marked tenderness, warmth, and overlying erythema. The lesion was nonfluctuant, noncompressible, and without any evidence of drainage. The patient had pain with movement of the left hip, which was localized to the tumor itself.

Prior hospitalization records referred to a history of post traumatic degenerative joint disease of the left hip. The patient reported that he had been struck on the left hip with a baseball bat 22 years prior and the lesion had been slowly growing since the trauma. He denied any history of hip pain or skin lesion prior to the traumatic event and also denied any fevers, chills, weight loss, or drainage from the lesion. Several times in the past 2 years he had been advised by his outpatient dermatologist to have radiologic imaging of the lesion but had missed multiple appointments for these studies.

The clinical differential diagnosis included an organized hematoma or abscess, cutaneous metastasis,

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dermatofibrosarcoma protuberans, and other soft tissue sarcomas.

Magnetic resonance imaging of the lower left extremity demonstrated a lobulated ovoid lesion in the subcutaneous fat adjacent to the skin at the level of the greater trochanter. The signal characteristics and enhancement pattern were suggestive of a proteinaceous or hemorrhagic collection or abscess but could not rule out neoplasm. There was no radiologic evidence of arthritis.

Needle aspiration of the mass revealed only minimal return of bloody aspirate. The lesion was excised in its entirety.

On histopathologic examination, the mass measured 3.1×2.2×1.5 cm. The subcutaneous tissue was composed of uniform appearing cells with a mild amount of eosinophilic cytoplasm and small minimally pleomorphic nuclei (Figure 2). No mitotic figures were evident. The neoplastic cells were arranged in small clusters and sheets around the blood vessels. Centrally, the lesion was hemorrhagic with extensive fibrosis and granulation tissue consistent with a glomus tumor with organizing thrombus. There was no involvement of the overlying skin. Immunohistochemistry of the tumor cells was positive for smooth muscle actin and negative for c-kit, S100, desmin, CD34, and chromogranin, consistent with glomus tumor cells. The histology and immunohistochemistry were compatible with a glomus tumor, with no characteristics of malignancy.

The left hip pain resolved postoperatively and has remained asymptomatic since removal of the glomus tumor.

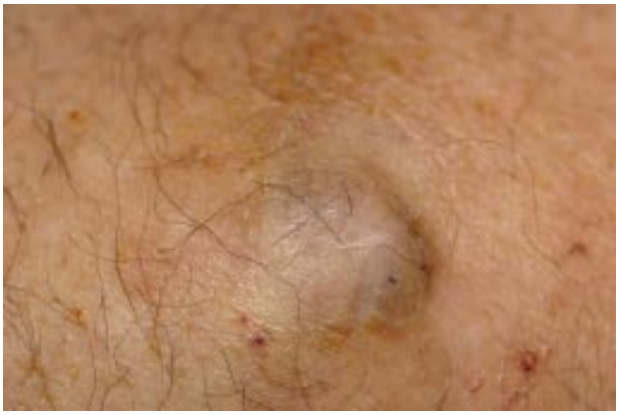

Figure 1. A 4×3-cm violaceous firm tender tumor on the left lateral hip.

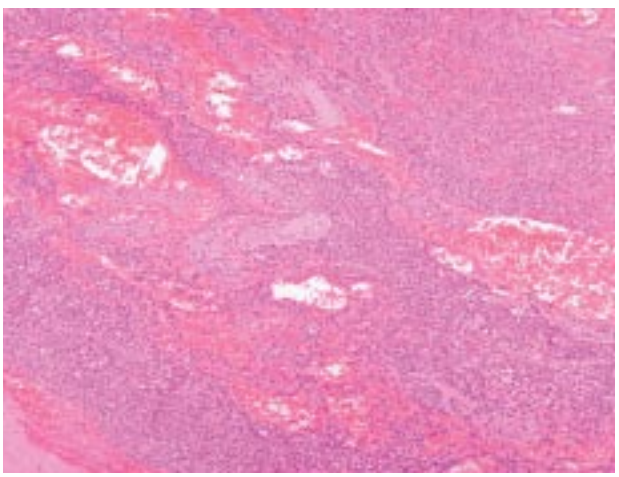

Figure 2. Uniform appearing cells, containing eosinophilic cytoplasm, arranged in nests adjacent to capillaries with central hemorrhage, fibrosis, and granulation tissue consistent with a glomus tumor with organizing thrombus (H&E, original magnification ×10).

Comment

Glomus bodies, situated in the stratum reticularis of the dermis, are neuromyoarterial receptors that detect fluctuations in temperature and regulate arteriolar flow.[1] The arterial end of the glomera, the Sucquet-Hoyer canal, is surrounded by small, spherical, uniform glomus cells that histologically resemble smooth muscle cells. Immunophenotypic and electron microscopic features of glomus tumors suggest that they arise from the modified smooth muscle cells of the glomus body.[7]

The glomus tumor was first described by Wood[8] in 1812, with further clarification of the histologic aspect of glomus tumors outlined by Masson[9] in 1924. These hamartomas are rare benign neoplasms clinically distinguished by their small size and the severity of pain that they cause. The clinical presentation is typified by paroxysms of pain radiating away from the lesion.[3] Although primarily found subungually on the hands, they have been described in other sites, including the head and neck, trunk, and lower extremities.[10] Glomus tumors most commonly occur as solitary red-blue lesions less than 1 cm in diameter. Multiple lesions are uncommon but have been reported.[9] While there is frequently a point of maximal tenderness, there is seldom a palpable mass in the early stages. In our patient, the tumor was unusually large with a diameter of 4 cm, and it was palpable over the left lateral hip.

The glomus tumor usually is a well-circumscribed lesion found in the dermis or subcutis and is often delineated by a well-defined fibrous capsule.[7] Typical glomus tumors are subcategorized as solid tumors, glomangiomas, or glomangiomyomas depending on the relative prominence of glomus cells, vascular structures, and smooth muscle.[1] Histologically, glomus tumors comprise the following 2 components: branching vascular channels separated by the

connective tissue stroma. The stroma contains aggregates, nests, and masses of small, bland-appearing, round glomus cells with scant eosinophilic granular cytoplasm. The cells lack nuclear atypia and mitotic activity is rare. Immunohistochemical staining is characteristically positive for vimentin and smooth muscle actin, negative for desmin and S100 protein, and variably positive for CD34.[11] In accordance with typical glomus tumor histology, the biopsy specimen from our patient demonstrated clusters and sheets of round cells containing scant eosinophilic cytoplasm arranged around blood vessels; these cells also stained positive with smooth muscle actin. Unlike a classic glomus tumor, however, the presence of hemorrhagic material, fibrosis, and granulation tissue seen within our patient's tumor is of uncertain significance but may represent the effect of repetitive trauma. There are no reports in the literature of glomus tumors complicated by central hemorrhage or fibrosis, which may explain the unusually large size of the tumor in our patient.

Glomus tumors may be solitary or multiple, painful or painless, idiopathic or inherited as an autosomal dominant trait.[12] Folpe et al[13] proposed a new classification system for glomus tumors after studying 52 cases with unusual features previously diagnosed as atypical. Malignant tumors were defined to have deep location (size greater than 2 cm) or atypical mitotic figures, or nuclear atypia with more than 5 mitotic figures per 50HPF (high-power field). Symplastic tumors demonstrated high nuclear grade with no other malignant features. Tumors that lack criteria for malignancy but have increased mitotic activity with superficial location, large size, or deep location were categorized as glomus tumors of uncertain malignant potential.[13] These neoplasms are thought to be benign, though infiltrative and malignant glomus tumors have been reported.[14]

Standard radiographic imaging of glomus tumors usually is normal. High-resolution magnetic resonance imaging is useful for defining the full extent of the lesion.[15,16] Angiography can confirm the vascular origin of the tumor and its location but is not a standard diagnostic tool.[9] Scintigraphy with labeled red blood cells does not show accumulation of radioactivity in the tumor over time.[17]

Although typically small in size, there have been reports of a number of glomus tumors with diameters greater than 1 cm. The largest reported glomus tumor was a 14-cm giant mass in a vein of the forearm. This mass did not demonstrate any aggressive clinical behavior, recurrence, or metastasis.[18] Abou Jaoude et al[10] reported 2 glomus tumors of the neck; one tumor simulated a parotid tumor and the second tumor originated from the internal perijugular space, with maximum diameters of 4 and 5 cm, respectively. Hall and Odell[6] presented a case of an 8-cm glomangioma of the hip that masqueraded as osteoarthritis of the hip for nearly 8 years and resolved after surgical extirpation of the tumor. A number of massive visceral glomus tumors also have been documented.[19,20] On histologic examination, none of these glomus tumors demonstrated any malignant or infiltrative characteristics. We now report the case of a 4-cm glomus tumor on the hip that also lacked features of a malignant process, despite the large size of the tumor.

In addition to the classic presentation of excruciating point tenderness of glomus tumors with or without a palpable subcutaneous mass, glomus tumors in close proximity to joints may present with arthralgia. Ghaly and Ring[5] reported a supraclavicular glomus tumor that presented with a 20-year history of undiagnosed shoulder pain, despite multiple pharmacologic treatments, physical therapy, and 2 corrective surgeries. Removal of a 9-mm encapsulated mass resulted in complete relief of joint pain.[5] Caughey and Highton[21] reported a patient with a 3-cm red-blue swelling over the left patella that was histologically consistent with a glomus tumor; the patient originally presented with a 13-year history of intractable knee pain. A popliteal glomangioma mimicking a tender Baker cyst was described in a 9-year-old girl who complained of pain and tenderness of her posterior left knee.[22] Monoarticular arthralgia or arthritis, although rare, may be the presenting symptom of periarticular glomus tumors and thus the diagnosis should be considered in the differential diagnosis of chronic, atypical, or treatment-resistant joint pain.

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