CASE REPORT

Primary Intraosseous Malignant Glomus Tumor of the Finger with a Different Radiological Appearance: a Case Report and Review Literature

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Abstract

Primary intraosseous malignant glomus tumor is very rare and is often presented as an osteolytic lesion. The present case study reports a malignant intraosseous glomus tumor of the index finger that was presented as an osteosclerotic lesion and invaded adjacent tissues, leading to ray amputation. We also reported result of a five-year follow-up and presented a short literature review for the lesion.

Level of evidence: IV

Keywords: Glomus tumor, Intraosseous, Osteosclerotic lesion

Introduction

Glomus tumor is a benign and rare vascular tumor that is the cause of 1.5% to 2% of all soft tissue tumors (1). This lesion occurs in a digital subungual location and is presented with pain that can be exacerbated by temperature change. It is presumed that glomus tumors are composed of glomus cells. However, they have been observed in extracutaneous sites that do not contain glomus cells, including the nose, eyelid, middle ear, lung, mediastinum, stomach, rectum, cervix, penis, and bone (2-3). A possible explanation is that these tumors may originate from perivascular cells that can differentiate into glomus cells (4).

The primary intraosseous glomus tumor, which is usually reported in the distal phalanx, has a role in the differential diagnosis of osteolytic lesion of the distal phalanx (5-7). It has been reported in the middle phalanx as a well-circumscribed radiolucent lesion that involves the proximal aspect of the middle phalanx of the 5th finger and leads to the endosteal erosion of the adjacent cortex, misdiagnosed as enchondroma (8).

Here we present an intraosseous malignant glomus tumor of the proximal phalanx of the index finger with a unique and different radiographical appearance resembling osteoid osteoma. The pathology, imaging appearance, treatment, and five-year follow-up results of the condition are described, and a short literature review is presented as well. The patient gave informed consent for doing the study and pulshing its report.

Case report

A healthy 34-year-old man was referred to a healthcare center due to a painful mass in the proximal phalanx of the left index finger. The mass, that had been noticed in August 2012, caused occasional pain and a slight range of motion restriction in the proximal interphalangeal joint. The patient alleviated the pain using painkillers. There was a small radiolucent lesion with significant sclerosing areas surrounding it in the radiographic images, indicating osteoid osteoma [Figure 1]. On the magnetic resonance images (MRIs), the lesion was observed as an area with prolongation at T1 and T2. The lesion was suspected to be osteoid osteoma, based on radiographic images.

The patient underwent partial resection and curettage for sampling at the center. The pathologic examination showed nonspecific bony trabecula, possibly due to inadequate sampling. The symptoms did not relieve and swelling increased within the next two years.

The patient's index finger was quite swollen and had

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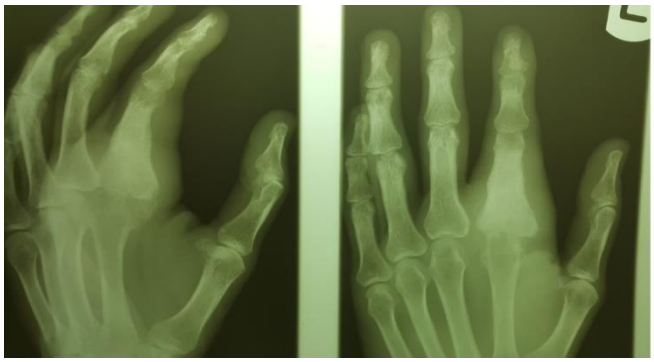

Figure 1. Radiographic images of the proximal phalanx of the index finger before the first biopsy

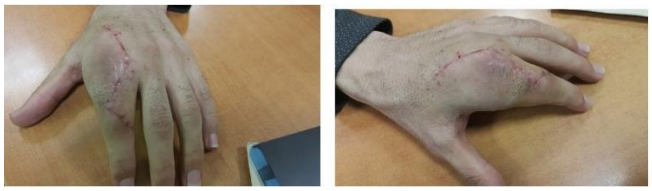

Figure 2. Swollen and painful finger two years after the first surgery

severe motion restriction in the metacarpophalangeal and proximal interphalangeal joint at the time he was referred to our center [Figure 2]. The patient used pain killers for relieving his pain during the past two years which had led to gastric bleeding due to excessive use. Therefore, he had been referred to our center for diagnosis and treatment.

The incisional biopsy of the lesion was carried out subsequently. The microscopic examinations revealed that neoplastic tissue consisted of branching vascular channels lined by intact endothelial cells, interspersed by round to ovoid glomus cells. The tumor cells had a lightly eosinophilic to amphiphilic cytoplasm with indistinct cell borders forming solid sheets of cells interrupted by different-sized vessels and a hemangiopericytoma pattern. On the high-power field, nuclei showed a hyperchromatic to vesicular chromatin pattern with inconspicuous to well-defined nucleoli exhibiting nuclear pleomorphic. Increased cellularity and mitotic activity up to 5 mitoses/50HPF were evident. Infiltrative growth of vessels invested by glomus cells in necrotic cancellous bone was obvious [Figure 3].

Tumor size, mitotic activity, and tumor pleomorphism led us to the possibility of a primary interosseous malignant glomus tumor. Immunohistochemistry assessment was performed and tumors cells were positive for actin-SM and collagen 4 and negative for CK-PAN, DESMIN, S100P, and CD34.

There was no sign of metastasis to any other bone or area based on the whole-body bone scan and systemic workup. Ray amputation was the preferred treatment due to the involvement of the metacarpophalangeal joint and metacarpal head. A microscopic examination of the amputated part confirmed the diagnosis. The proximal margin of the amputated part was free of tumor cells. In the five-year follow-up, the patient was healthy and normal, without any problem with his hand [Figure 4].

Discussion

Glomus tumor is a neoplasm composed of round cells of the same size, margin, and eosinophilic cytoplasm. Increased mitosis and atypia are commonly observed, and there is vascular perfusion around the lesion (9). The symptoms of glomus tumor include spot tenderness pain and sensitivity to cold. Other symptoms, including palpable and bluish nodules, can be observed on examination in cases where the lesion is superficial (10). Although glomerular tumors are usually soft tissue lesions that do not involve the bone, the associated bone abnormalities are not uncommon. Completely cortical shallow bone defects, usually involving distal phalanx tuft adjacent to the tumor, are considered to be the result of cortical pressure caused by the lesion (8, 11).

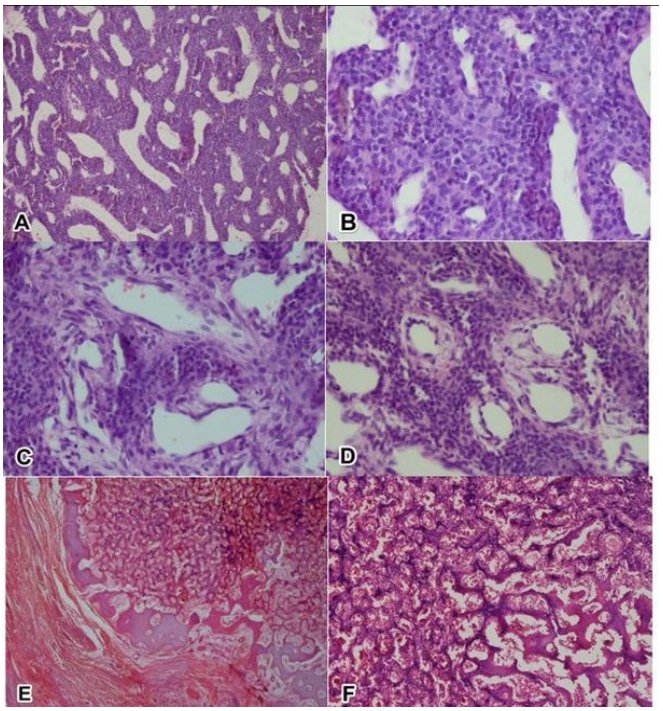

Figure 3. Microscopic sections of malignant glomus tumor

A. Solid sheets of cells interrupted by irregular or "staghorn" vessels show glomangiopericytomatous differentiation (hematoxylin-eosin, original magnification×100)

B & C. Sheets of round to polygonal cells with pale eosinophilic to amphophilic cytoplasm and indistinct cytoplasmic boundaries closely associated with variably sized blood vessels. Mild atypia in the form of nuclear hyperchromasia, pleomorphism, and mitotic count exceeding 5 mitoses/50HPF has been observed (hematoxylin-eosin, original magnification×400)

D. Focal mild perivascular hyalinization giving the vasculature a prominent appearance (hematoxylin-eosin, original magnification×400)

E. At low magnification, an infiltrative growth pattern is evident in degenerated and necrotic trabecular bone (hematoxylin-eosin, original magnification×40)

F. Diffuse glomus tumor with infiltrative growth of vessels which are encircled by ectomesenchymal cells infiltrates bony trabeculae (hematoxylin-eosin, original magnification×100)

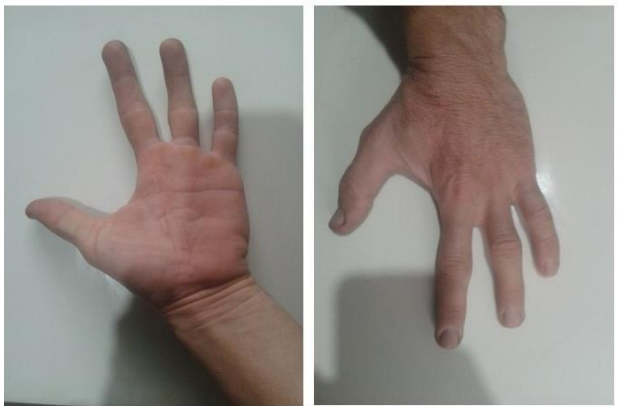

Figure 4. Five years after the second ray amputation

Glomus tumor is often benign; however, if it is deep or larger than 2 cm or has a pathology of malignant behavior, such as increased mitosis, necrosis, or atypia, the lesion should be suspected of being malignant. Glomus tumors can be found in uncommon sites, such as the gastrointestinal tract and intramural and intra-osseous sites. Although most cases of glomus tumors are diagnosed in the ages of 20-40 years, sometimes the symptoms lasted for several years (11).

The first published report of intraosseous glomus tumor, as a tumor in the distal phalanx of the thumb, appeared in 1939 (12). Intraosseous localization of the glomus tumor is very rare. To the best of our knowledge, only 20 cases of glomus tumor have been reported in the literature (4) which usually resemble enchondroma or epithelial obstruction cyst in the radiographic image. The correct diagnosis is made only after surgical removal and pathological examination of the specimen (13).

In the interpretation of a lesion's radiographic image, there is a high risk of confusion with enchondroma due to their similarity. Other differential diagnoses include aneurismal bone cysts, epithelial cysts, and giant cell tumors. In our case, interestingly, radiographic images showed sclerosing and radiopacity, rather than radiolucency which might be due to excessive bone reaction to the lesion. It should be noted that MRI can also show lesions with an increased signal in the T2 sequence (14).

Immunohistochemistry can be very helpful in making the diagnosis in cases with an abnormal tissue appearance. Glomerular tumors are typically stained for muscle-specific actin and vimentin, due to the fact that they are generally considered to be modified smooth muscle cells. Reaction to keratin, protein S 100, factor VIII antigen, and myoglobin has been shown to be negative (15). Intraocular glomus tumors behave the same as soft-tissue glomus. They are usually benign and do not metastasize or undergo malignant changes. Large and deep lesions and lesions with malignant changes are more prone to malignancy (16).

In conclusion, although intraosseous malignant glomus tumor is very rare and is considered in the differential diagnosis of a radiolucent lesion, it should be taken into account as a differential diagnosis of a radiopaque and sclerosing lesion, including osteoid osteoma. Although there is no report concerning this tumor's metastasis, local invasion of tumor cells from proximal phalanx to metacarpophalangeal joint and metacarpal head in our case suggested the invasiveness of the tumor over time.

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