Title:


Abstract:

Malignant transformation of glomus tumor is exceedingly rare and most common reported in lower extremity and abdominal viscera. We could find only 6 previous case report of malignant glomus tumor in the hand. Although large and deeply located glomus tumors are considered to be malignant, evidence has shown that most of these cases were clinically benign. These lesions are better considered as glomus tumor of uncertain malignant potential. Due to rarity of malignant glomus tumor, decision for operative treatment must be based on few case reports. In this article we review the literature for malignant glomus tumor of hand and their management, also we report a 49 year-old man with glomus tumor of uncertain malignant potential in thumb who was treated by wide amputation

Key words: Glomus tumor; malignancy; Bone involvement

Level of evidence: 4
Introduction

Glomus Tumors are benign vascular neoplasms that originate from neuromyoarterial structures in the reticular dermis called glomus bodies. Digital glomus tumors consist approximately 1% to 2/6% of all hand tumors and are more commonly seen in middle aged women. In contrast extra digital cases are more common in older men (1).

Malignant transformation of glomus tumors is exceedingly rare. Till now only few cases of malignant glomus tumors of hands were reported in the literature, moreover, only in one case bony involvement has been reported. In current presentation, we reported a case of glomus tumor of uncertain malignant potential in the thumb with severe bony destruction.

Case report

A 49 year-old man with history of right thumb episodic severe paroxysmal pain and cold hypersensitivity visited our hand clinic. Pain has started secondary to blunt trauma since three years ago. Past medical history and drug history were unremarkable. He was operated two-times before final surgery. Three years ago, in first operation we excised the tumor with curettage of distal phalanx lytic lesion (figure 1 A,B) but the symptoms were persistent for more two years. Therefore, in second operation, we curettage the tumor and filled it with iliac bone graft. Pathology report of both operations indicated on glomus tumor (figure 1 C,D). After these two operations the symptoms were still persisting. Physical examination revealed point tenderness, swelling and deformity of the thumb. Other findings were soft tissue mass in volar aspect of distal and proximal phalanges of the thumb with venous engorgement (figure 1G,H). No lymphadenopathy was detected. Plain X-ray showed massive destruction of distal
phalanx with soft tissue enlargement (figure 1E,F). MRI findings were abnormal soft tissue mass with loss of normal configuration of distal phalanx and encasement of adjacent tendon (figure 1I, J). Triphasic whole body bone scan by TC 99m-MDP showed intense radiotracer uptake of distal phalanx of the thumb. Chest x-ray, abdominopelvic sonography and bone scan didn’t show any distant metastasis. Due to two episodes of tumor recurrence and severe functional impairment we decided to treat the patient by amputation of the thumb through middle part of proximal phalanx and cover the stump of amputation with greater dorsal flap instead of palmar flap because soft tissue invasion of tumor in palmar surface was more than dorsal (figure 1M, 1N).

Histopathological sections shows infiltration of a neoplasm composed of tight convolutes of capillary sized vessels surrounded by collars of round uniform cells with punched out nuclei with mild to moderate atypical and eosinophilia cytoplasm arranged in hemangiopericytoma like pattern with unclear pleomorphic and atypia in some region and infiltrating border (figure 1K, L). Mitotic figures were scant and no tumor necrosis was identified. All these findings were compatible with glomus tumor of uncertain malignant potential. A margin of amputation was free of tumor and the patient was symptoms free in 6 month follow up.

Discussion

Glomus tumors are benign hamartomas that arise from the normal glomus apparatus, located in subcutaneous tissue. The normal glomus body is a contractile neuromyoarterial receptor that controls blood pressure and temperature by regulating flow in the cutaneous microvasculature
Glomus bodies are highly concentrated in the tips of digits especially under the nail, so the tumor is usually in the subungual area.

Distinguishing features of the glomus tumor is the classic triad of symptoms: Hypersensitivity to cold, paroxysmal pain and point tenderness. Ultrasonography and MRI can be a valuable method of imaging to detect glomus tumors.

Malignant transformation of glomus tumors is exceedingly rare and has been often reported in many different anatomic locations such as the lower extremities and abdominal viscera. Malignant transformation in the hand is extremely rare. However, hand surgeons should be aware of the possible diagnosis of malignant glomus tumor in the hand. There are only 6 previous reports of malignant glomus tumors in the hand.

Wide local excision has traditionally been recommended as the adequate treatment, as most malignant glomus tumors are considered to be unlikely to metastasized. Although metastasis is an unusual event in malignant glomus tumor, locally aggressive behavior and local recurrence appears to be more common. Histopathologically malignant glomus tumor is defined as those that: 1) have marked nuclear atypia and elevated mitotic rates greater than 5 mitosis per 50 high power fields. Or 2) display atypical mitotic figures. Classification of glomus tumor with atypical features was showed in table 2.

Although large (greater than 2 cm) and deeply located glomus tumors are considered to be malignant, evidence has shown that most of these cases behave in clinically benign fashion. Therefore, these lesions are better to considered as glomus tumors of uncertain malignant
potential. Due to rarity of malignant glomus tumor, operative treatment must be based on few case reports.

In our case report the patient did not fulfill complete criteria of malignant glomus tumor, but due to large size (3cm), cellular atypia and locally recurrence, considered as glomus tumor of uncertain malignant potential so we decided to do wide amputation through proximal phalanx of thumb because of radiographic bony involvement of distal phalanx and soft tissue involvement of volar aspect of distal part of proximal phalanx. In previous case reports only one case had bony involvement of distal phalanx who was operated by ray amputation (Table-1) Our patient was symptom free 6 months after surgery.

References:


Table 1: Review of previous case reports. (F: female, Lt: left, M: male, Rt: right, Y: year)

Table 2: Classification of glomus tumor with atypical features