

Case Report

Glomus tumor: an atypical presentation

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ABSTRACT

Glomus tumors are rare soft-tissue neoplasms of the neuromyoarterial glomus body, typically present in adults as small, blue-red papules or nodules of the distal extremities, with most cases involving subungual sites. The majority of glomus tumors are benign, malignant cases have been rarely reported, with such cases typically being locally invasive. 27 years old patient presented with four years history of intermittent swelling, pain in left lower leg. Biopsy of the lesion revealed lobulated vascular tumour composed of endothelial cells showing intracytoplasmic lumina and solid aggregates of tumour cells, divided by intervening fibrocollagenous septae. Glomus tumour of uncertain malignant potential was diagnosed after excision biopsy. Multispeciality team approach was done for complete cure of the condition.

Keywords: Glomus tumor, Locally invasive, Team approach

INTRODUCTION

Glomus tumours are rare benign neoplasms, solitary and small, and commonly found under the fingernails. These are specialized arteriovenous anastomosis usually found in the skin of the extremities.^{1,2} Incidence is 1.6% among the 500 consecutive soft tissue tumors report from Mayo Clinic. If there is no underlying inherited condition then the tumour is considered “sporadic” or “random”. Glomus tumours are one of the painful tumours of skin others being leiomyoma, angioliipoma, eccrine spiradenoma and neurilemmoma. Pain is produced when exposed to cold. They are thought to represent neoplastic proliferation of modified smooth muscle cells originating from pre-existing normal glomus cell populations. They can be present at sites not known to contain glomus cells. These tumours may arise from perivascular cells that can differentiate into glomus cells. An atypical presentation of glomus tumor due to its unusual location is reported.

CASE REPORT

Twenty-seven years old male patient presented with intermittent swelling and pain in the left lower leg, for four years and inability to walk and bear weight for 20 days. There was no history of trauma, fever, weakness in any other part, or skin lesions. Dermatological examination revealed solitary 6×8 cm, ill defined, excruciatingly tender swelling in lateral aspect of left lower 1/3 of leg. Touch sensation was lost over an ill defined area overlying swelling. Skin over the swelling was normal. Wasting of left leg was present. Muscle power of dorsiflexors of foot was grade 3 out of 5, with impending foot drop. Diagnosis of Hansen's disease pure neuritic in type I lepra reaction with probable nerve abscess (left) was entertained. Multi drug therapy (MDT) (MB) and oral steroids in the dose of tab prednisolone 40 mg was started.

Table 1: Classification of glomus tumours with atypical features.

S. no.	Type	Characteristics
1	Malignant glomus tumour	Marked atypia+mitotic activity (>5/50 HPF or atypical mitotic figures or large size (>2 cm) + deep location
2	Glomus tumour of uncertain malignant potential	Superficial location +high mitotic activity (>5/50 HPF) or large size only or deep location only
3	Symplastic glomus tumour	Lacks criteria for malignant glomus tumour and marked nuclear atypia only
4	Glomangiomas	Lacks criteria for malignant glomus tumour or glomus tumour of uncertain malignant potential and diffuse growth resembling angiomas with prominent glomus component

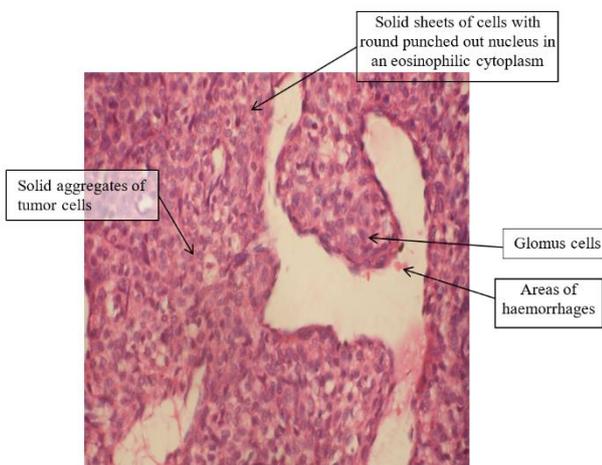


Figure 1: Glomus tumour.



Figure 2: MRI left leg: residual ill defined infiltrating lesion in lateral aspect of the left ankle.



Figure 3: MR Angiography, multifocal delayed enhancing lesion in the intermuscular planes in distal 1/3 of leg.

However, after 3 weeks of MDT and steroids, patient was not relieved of swelling and pain. He was still unable to bear weight. Ultrasonography revealed multiple subcutaneous abscess involving left lower leg. Incisional biopsy revealed lobulated vascular tumour composed of endothelial cells showing intracytoplasmic lumina and solid aggregates of tumour cells, divided by intervening

fibrocollagenous septae. Large vascular channels were seen suggestive of low grade vascular tumour. Vascular surgeon's opinion was sought. Excision biopsy of the tumour done. Histopathological examination revealed glomus tumour of uncertain malignant potential based on deep location (Table 1 and Figure 1). Immunohistochemistry showed actin and vimentin positive. Despite surgical excision patient was not relieved of symptoms. Magnetic resonance imaging (MRI) left leg showed residual ill defined infiltrating lesion in the lateral aspect of left leg (Figure 2). MR angiography showed multifocal delayed enhancing lesion in the inter muscular planes in distal 1/3rd of left leg (Figure 3). Embolisation of feeding vessels of the residual tumour was done by interventional radiologist. Patient was relieved of pain immediately. However, post procedure the tumour did not shrink completely. Over a period of two months swelling recurred at exision site. Debulking of residual tumour with local flap cover was done by reconstructive surgeon since the lesion was extremely painful. A final diagnosis of infiltrative glomus tumour in intermuscular planes was entertained. Patient is on regular follow up and presently asymptomatic.

DISCUSSION

Glomus tumours are classified as glomus tumour proper, glomangioma, and glomangiosarcoma.³ Glomus tumour proper is well circumscribed or encapsulated dermal tumour which may extend into subcutis, composed of solid aggregates of glomus cells surrounding inconspicuous vessels. Glomangiomas are usually multiple, mostly seen in children, thought to be inherited as autosomal dominant trait with incomplete penetrance linked to chromosome 1p21-infiltrating glomus tumours, as seen in present case, is a rare variant of histologically otherwise typical glomus tumour that is deep seated.⁵ They can be seen in deep soft tissue, bone vagina, trachea, lung, gastrointestinal tract, veins and nerves. About 10% recur following excision.^{6,7} Glomangiosarcomas are very rare, <10% of all glomus tumours are malignant.

CONCLUSION

Index case is an uncommon presentation of deep infiltrative glomus tumour masquerading clinically as

Hansen's disease neuritic with nerve abscess due to signs of lower limb wasting, pain and possibility of impending foot drop. Diagnostic dilemma was solved by investigations in the form of MRI, MR angiography and surgical biopsies. The treatment rendered was by multispeciality team approach of dermatologist, interventional radiologist, vascular and reconstructive surgeons. Embolisation of the tumour relieved the patient of the distressing pain but debulking was found necessary as it did not shrink despite embolization. This combined conservative treatment helped save the patient from undergoing a mutilating below knee amputation.

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