CASE REPORT

Primary Intraosseus Glomus Tumour of the Thumb: A Rare Presentation and its Management

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Abstract

A 23 year old female presented with excruciating pain in the right thumb of 6 months duration, with an acute exacerbation of 10 days, mainly on exposure to cold. On examination, a bluish hue was present in the subungual region of the right thumb, with pin point tenderness. Hildreth's test and cold sensitivity test was positive. Radiographs and MRI showed a well defined osteolytic lesion in the distal phalanx with cortical thinning but no cortical breach. Surgical resection of the lesion was done and the histopathological examination confirmed the diagnosis of intraosseous glomus tumour. At one year follow up, she showed an excellent functional outcome.

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Introduction

Glomus tumour is a rare benign vascular lesion arising from the glomus cells, which are modified perivascular smooth muscle cells without an intermediary capillary bed. Normal glomus bodies are meant for thermo regulation of the skin which are located in the deep dermis and subungal region of the finger tips[1–3].

The most common site for glomus tumour is the finger tip which accounts for 1–5% of all hand tumors[1–3]. The glomus tumour was named by Masson in 1942[4]. It usually presents with excruciating pain and a bluish solitary nodule in the subungual region of the finger tips. Although they are said to arise from glomus bodies, they have also been reported to arise from regions which dot have glomus cells like cervix, eyelid, middle ear, lung and rectum[1–3,5].

Primary intraosseous glomus tumours are very rare with only 21 cases reported in the literature so far, five of which invoved the spine[6–10]. 4 cases had been reported in the long bones, one in the proximal tibia, two in the ulna, one in fibula[6–10]. We are reporting a rare presentation of a primary intraosseus glomus tumor of thumb. On an extensive review of literature, less than ten similar cases were found to be reported so far[6–10].

Case Report

A 23 year old female presented with pain in the right thumb of 6 months duration, which was excruciating in nature and aggravated on exposure to cold. She had undergone conservative management initially, but her symptoms were getting worse with time. There was no history of local trauma or infection. On examination a bluish hue was present in the

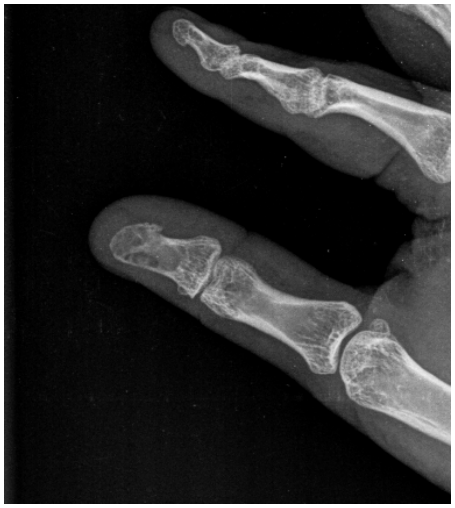


Figure 1. X-ray showing lytic lesion in the distal phalanx.

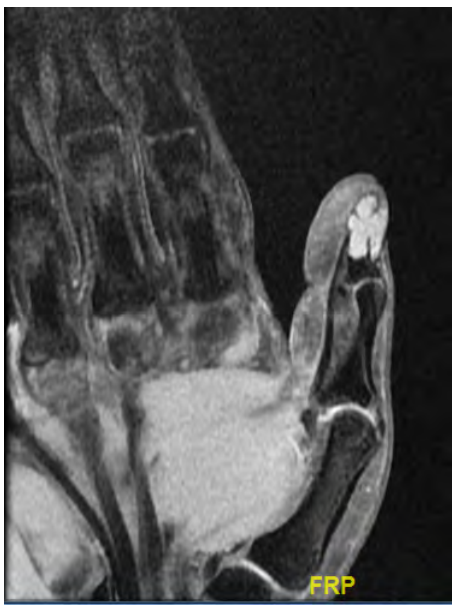
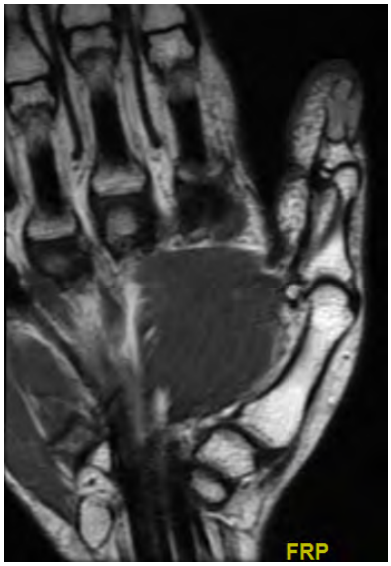
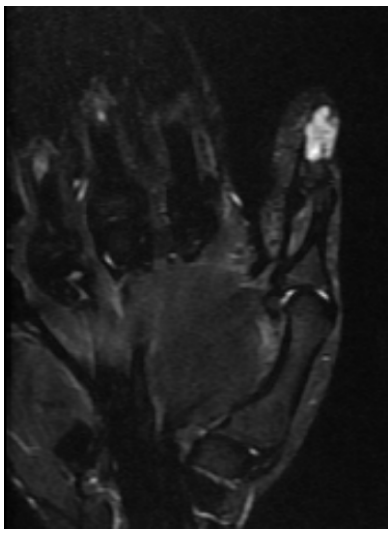


Figure 2. Coronal T1 MRI image.



(a)



(b)

Figure 3. (a) Coronal PD MRI image. (b) MRI image.

subungual region of right thumb, with pin point tenderness. On further examination the Hildreth's test and the cold sensitivity test were positive. With a provisional diagnosis of glomus tumor, radiography of the thumb was done and to our surprise, there was a 1x1 cm well defined osteolytic lesion in the distal phalanx with thinned out cortex and no cortical break. MRI was done, which showed hyper to isointense lesion in the distal phalanx on T1 weighted image and homogenous high signal intense lesion in T2 weighted images (Figures 1-3).

Excisional biopsy was done under local anesthesia through a midline nail splitting approach. After complete excision of the tumor which was 1x1 cm, a thin shell of intact cortex was present anteriorly. So primary bone grafting was not done. Post operative period was uneventful and the patient was totally free of her pain. Sutures were removed after 2 weeks and gradual mobilization was started. By 3 months, she had resumed all her activities. One year follow up of the case showed an excellent clinical outcome (Figures 4–8).

Histopathological Examination

On gross examination, it was a grey white lesion aggregate measuring 0.6 × 0.6 cm. There was no foci of hemorrhage or necrosis identified grossly.

On microscopic examination under scanning power, multiple fragments of a cellular neoplasm was seen which under low power was arranged as nests surrounded by thin walled vessels. There was hyalinised stroma in between. Under high power, the

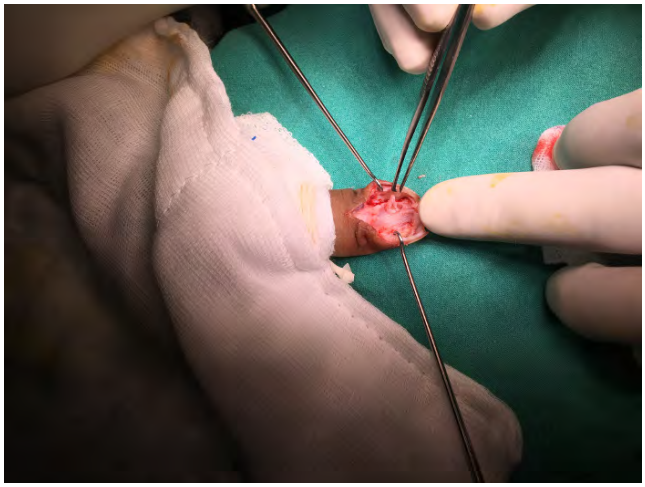


Figure 4. Midline nail splitting approach.

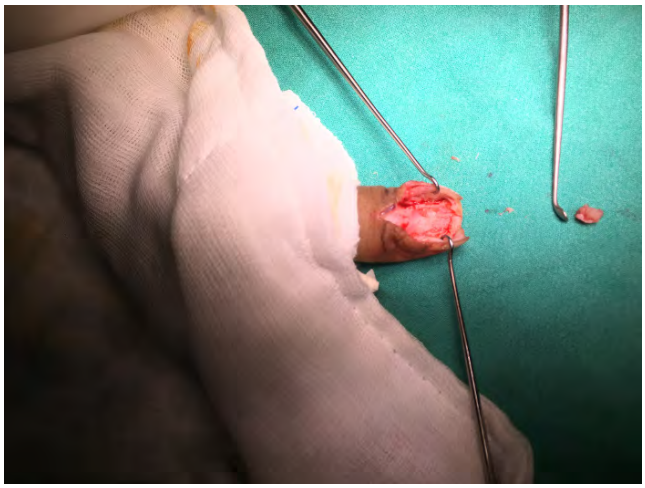


Figure 5. Curetting the lesion.

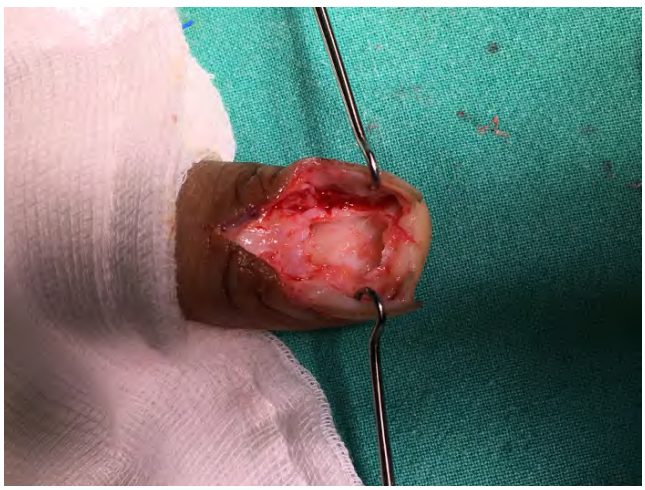


Figure 6. After complete curettage.

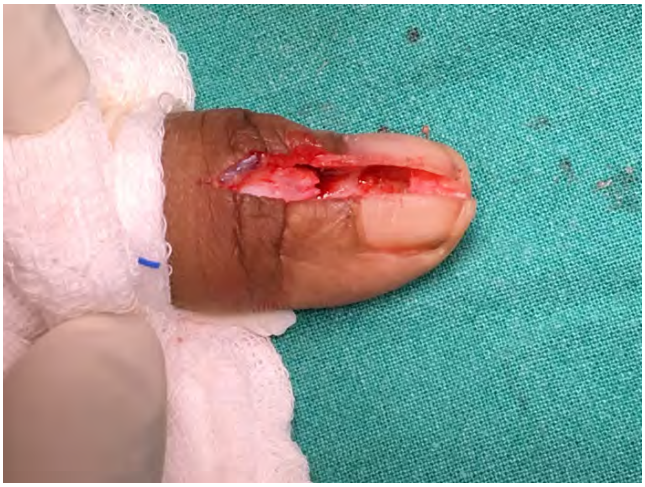


Figure 7. Before closure.

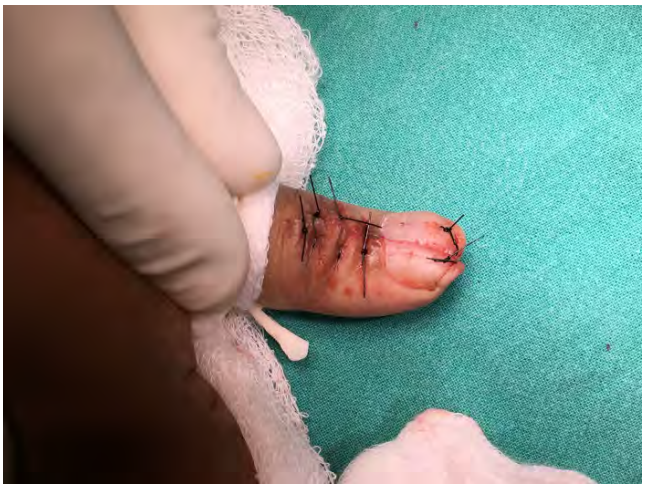


Figure 8. After closure.

cells were uniform, polygonal shaped, with moderate amount of eosinophilic cytoplasm, round central nucleus with fine chromatin. There was scant mitosis and no necrosis. On IHC staining, the tumour cells were positive for smooth muscle actin (Figure 9).

Discussion

Glomus tumour is a rare benign vascular tumour arising from the glomus cells, which are modified perivascular smooth muscle cells without an intermediary capillary bed. Glomus bodies are neuromyoarterial structures meant for thermo regulation and regulation of normal blood flow, located in the deep dermis and subungal regions of finger tips[1–3].

Glomus tumour was first described by Wood in 1812, and in 1924 named by Masson[4]. The usual site for the glomus tumour is the finger tips, 1–5% of all hand tumors. It usually presents with a bluish

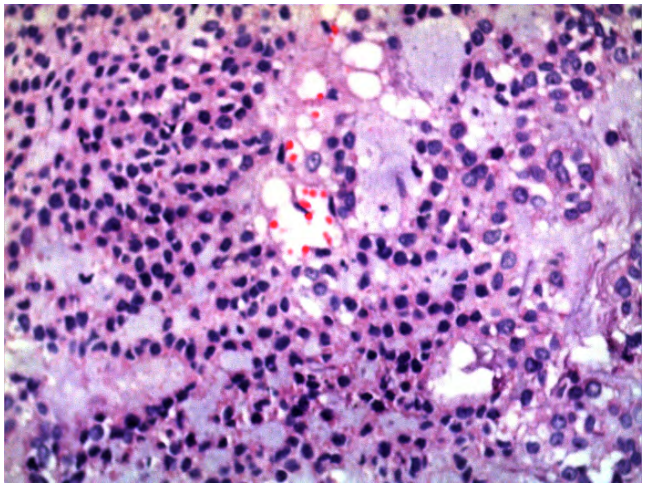


Figure 9. High power microscopic image of the lesion.

or a pinkish red visible or palpable solitary nodule in the subungual regions of finger tips[1–4]. They may also present as a curvature or deformity of the nail plate[4,5]. The characteristic feature of the tumor is the clinical triad of intense paroxysmal pain, exquisite point tenderness, and sensitivity to cold[4]. The Love test which has a sensitivity of 100% but a specificity of 0% involves probing the lesion with a pointed instrument which triggers pain in the affected area but not the area immediately adjacent. Reproducing the pain by induction of ischemia with a tourniquet to the affected limb refers to the Hildreth sign. Cold water or ethanol applied to the affected area will also induce the symptoms. This test has a sensitivity and specificity of 100%[1–4]. Glomus tumours should be differentiated from other painful conditions such as neuroma and gouty arthritis[11]. Occasionally multiple glomus tumours have been found in patients with type 1 neurofibromatosis. These are often harder and painless and are rarely found in the digits[11].

Even though the condition can be diagnosed clinically, standard radiography, ultrasound, and magnetic resonance imaging (MRI) can be used to localize the tumour and determine its size preoperatively. Radiography can detect large lesions and long-standing tumors characterised by thinning or erosion of the cortical bone in the phalanges. In MRI the lesions appear as slightly hypointense or hyperintense in T1images and hyperintense in T2 images. The treatment of choice is complete excision of the lesion and the commonly used surgical approaches are the transungual and the lateral[4].

Even though they are said to arise from glomus bodies, these have also been reported to arise from regions which do not have glomus cells like the cervix, eyelid, middle ear, lung and rectum. 20 to 40 years age is the most commonly involved age group though it can be found in any age group. There is a female predisposition of 3:1 in subungual lesions. Large soft tissue glomus tumors in the subungual region may cause extrinsic erosion of distal phalanx with clear sclerotic margin. Primary intraosseous glomus tumors are very rare with only 21 cases reported in the literature, among which five involved the spine[6–10]. Intra osseous glomus of the long bones have been described in tibia, ulna and fibula[6–10]. To our knowledge, less than ten cases of primary intraosseus glomus tumor of thumb have been reported in the literature so far[7,8,10].

Radiologically, differential diagnoses for such a lesion in the phalanx include enchondroma, epithelial inclusion cyst and a simple bone cyst. MRI can suggest the possibility of glomus tumour, but definitive diagnosis can be established only after excision of the lesion and histopathological examination. Microscopically, glomus tumour is a benign neoplasm composed of polygonal cells with moderate amphophilic to eosinophilic cytoplasm and central round nucleus. Glomus cells are arranged as nests surrounded by capillary sized vessels. Glomus tumours are positive for smooth muscle actin and H-caldesmon. Ultrastructurally glomus cells have bundles of thin actin-like filaments with dense bodies[12].

Though a rare disease, proper knowledge about the condition, thorough clinical evaluation along with appropriate radiological investigation and histopathological examination can help in the diagnosis. Selection of appropriate treatment method and a good post-operative rehabilitation protocol can assure an excellent functional result as in our case.

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