

Intraosseous glomus tumor of distal phalanx, thumb :A rare case reportRuchi Rathore¹, Deepshikha Arora², Ankit Jain³, Sarla Agarwal⁴¹Assistant professor, Department of pathology, NDMC Medical college, New Delhi, India² ex junior resident pathology, University college of medical sciences, current SMO CGHS, New Delhi, India³ Ex senior resident, Pathology, university college of medical sciences, New Delhi, India⁴ Retired professor and Head, pathology, university college of medical sciences, New Delhi, India**ABSTRACT**

Glomus tumors are rare benign neoplasms of glomus bodies. Commonly encountered in the subungual region of digits these can be found anywhere in the body. Although some rarely encountered sites including stomach liver synovium etc. have been reported, intraosseous location of these tumors have been described in less than 25 cases in the literature. We present a case of a 60 year old female presenting with complaints of stabbing pain for past 2-3 years in the distal phalanx of the right thumb. Radiography revealed a well demarcated cystic lesion in the distal phalanx. Microscopy and immunohistochemistry were suggestive of an intraosseous glomus tumor. Since the occurrence of these tumors in the bone is very rare and there are 10% chances of recurrence and even smaller chance of malignant transformation, correct diagnosis and complete excision is essential for better prognosis of the patients.

Key Words: Glomus Tumor, Bone, Intraosseous lesion, Lytic Lesion, Distal Phalanx**Introduction**

Glomus tumors except for glomangiosarcomas are rare benign neoplasms derived from the neuromyoarterial glomus cells. First described by Mason in 1924, they account for only upto 1.6% of the 500 consecutive soft tissue tumors as reported by Mayo Clinic. [1, 2] The glomus body is located in the stratum reticularis of the dermis and plays a key role in thermal regulation. Majority of these cases occur in distal extremities particularly involving the subungual region and the forearm as glomus bodies are more common in the palmar and plantar regions especially the nailbeds and fingertips.

Although these tumors have been reported at various other sites including stomach mediastinum, liver and synovium in the past, intraosseous locations have been reported in less than 25 cases so far.

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E Mail: ruchirathore13@gmail.com**Case report**

A 60 year old female presented with complaints of gradual onset of stabbing pain for past 2-3 years in the distal phalanx of the right thumb. This pain had recently increased in intensity and it worsened at night. There was no history of trauma or infection. Radiographic examination revealed a 7mm well demarcated round to oval radiolucent cystic lesion in the distal phalanx of the thumb. The lesion also thinned the cortex out. In view of the clinical and radiological picture a possibility of enchondroma was thereby suggested. (Figure 1) The blood chemistry and serological investigations of the patient were within normal limits. The cyst was excised and sent for histopathological examination. It was reported as round cell tumor from a private lab. We received the microscopy slides and block for review at our department. On review, the lesion consisted of branching vascular channels lined by endothelial cells, interspersed by uniformly round to ovoid cells forming nests, sheets, and trabeculae. The tumor cells have lightly eosinophilic to amphophilic cytoplasm with well-demarcated borders. Some of the tumor cells had vacuolated cytoplasm. The nuclei were round to oval with uniform dense chromatin and inconspicuous nucleoli. There was no prominent nuclear atypia, and mitotic activity was low. On morphology, a diagnosis

of glomus tumor was made and immunohistochemistry was applied for confirmation. The tumor was found to be positive for smooth muscle actin (SMA) and vimentin and negative for S-100 and neuron specific enolase (NSE) thus confirming the diagnosis.(Figure 2) Post-operative course of the patient was uneventful as she was free of any clinical manifestations and post op PET scan showed absence of any residual hypermetabolic focus. The patient was under follow up for 2 years and did not develop any signs of recurrence so far.

Discussion

Glomus is a neuromyoarterial structure located in the skin whose function is to regulate the temperature via arteriovenous shunting of blood. It is a specialized vascular anastomotic complex which is surrounded by various nerve endings. Although these glomus bodies are present in the stratum reticulare throughout the body, they are more concentrated palmar and plantar regions. Fingertips especially the nailbeds are particularly rich in such glomus structures [3]. Glomus tumors occur mostly in the corium and subcutaneous tissues, usually of the finger tips and present with classical symptoms such as aching pain, exquisite point tenderness, and temperature sensitivity [4]. However since glomus structures are present through the body, their occurrence has been described at various common and uncommon sites including the digits, mediastinum, rectum, cervix, mesentery, eyelid, nose, and chest wall [4]. The occurrence of this tumor within the bone, as seen in our case, is distinctively rare. On our search of literature very few cases have been reported so far. The incidence of these tumors is estimated to be merely 1.6% among the consecutive 500 soft tissue tumors reported by Mayo Clinic [3]. The origin of intraosseous glomus tumors however still remains controversial. While some studies hypothesize that they originate from soft tissue (of the normal arteriole and normal venule), and erode through the opening in the bony cortex, or the others say they may arise within the bone itself [5,6,7]. According to Jaffe, intraosseous glomus tumors can originate in a normal glomus organelle, which occasionally can be located within the medullary cavity of the bone [8]. Some authors also suggest that the pericytes located within the walls of the intraosseous blood vessels may also be the potential cells of origin. As in our case, the average age range of affected individuals is 14 to 61 years, with a slight female predominance [5]. The most common site of the

primary intraosseous glomus tumor is the distal phalanx of the finger [8]. The presence of glomus tumor describes in an intraosseous location of thumb also makes this case uncommon. Grossly these tumors are usually small, ranging in size from a few millimeters to 2 cm. [5] On histopathology the key features are the presence of nests or masses of glomus cells surrounding thin-walled vessels. Characteristically there is excessive vascular proliferation with endothelial thickening and presence of large cells known as glomus cells associated with the muscular layer of these vessel walls. However if the cellularity of such tumors is high they can rarely be misdiagnosed as hemangiopericytoma, adnexal tumor, apudoma, metastatic deposits of prostatic carcinoma, or even a microfollicular thyroid carcinoma. [9]

In such cases of diagnostic dilemma, the combination of actin and vimentin, positively coupled with a negative reaction for cytokeratin, factor-8 related antigen, and S-100 protein confirms the diagnosis of glomus tumor over other differentials. [10,11] Since in our case the tumor was intraosseous, other differentials like epithelial inclusion cyst, simple bone cyst, aneurysmal bone cyst, enchondroma or rarely metastasis especially from a bronchogenic carcinoma were also considered on radiography. However the classical histopathology of the lesion ruled out the above. Glomus tumor has been subclassified histopathologically into three variants namely vascular, myxoid and solid forms depending on the amount of vascularity and cellularity of these tumors. The vascular form consists of prominent vascular component with abundant cavernous spaces, myxoid form consists of broad bands of loose, textured tissue between the cords of glomus cells, and solid form consists of solid mass of epitheloid cells with relatively small amount of connective tissue component and few vessels with a small lumen [4]. In our case due to the presence of myxoid change we subclassified it as the myxoid variant of primary intraosseous glomus tumor. Glomus tumors are usually treated by curettage thereby meticulously shelling out the entire lesion as has been done in our case [3]. The postsurgical recurrence rate is approximately 10%. In our case the patient was under follow up for 2 years and did not show any signs of recurrence so far. Since in exceptionally rare cases a malignant transformation after recurrence has been described we also did a PET Scan after 3 months of surgery of the patient [8]. It further ruled out any possibility of recurrence of the same.

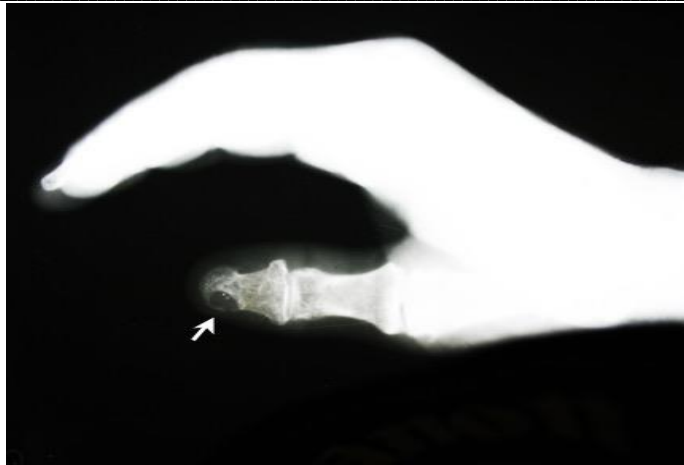


Fig 1: X-ray showing a well demarcated lytic lesion in the distal phalanx of thumb.

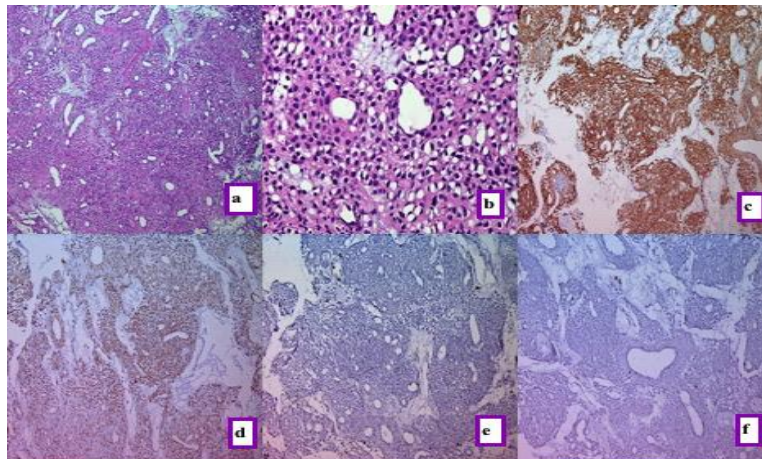


Fig 2 : **a)-** Tumor with uniform round to ovoid glomus cells and interspersed blood vessels [Hematoxylin and Eosin (H & E) ,100X]. **b)-** Cells with uniform nuclei and no atypia (H & E, 400X). **c)-** Diffuse cytoplasmic positivity for SMA (IHC, 400X) . **d)-** Vimentin positive tumor cells (IHC, 400X). **e)-** S-100 stain, negative (IHC, 400X), **f)-** NSE negative (IHC, 400X).

Conclusion

Primary intraosseous glomus tumors are very rare especially in the thumb. Though benign, there are 10% chances of recurrence and even smaller chances of malignant transformation in these tumors . So a correct diagnosis and complete excision is essential for better prognosis of the patients. The possibility of this tumor, amongst the other more commonly described intraosseous tumors, must also be kept in the list of differential diagnosis .

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