Case Report

Malignant Glomus Tumor: A rare and locally aggressive cancer in a young male

Deepak K. L[1], Pragnya Coca[2*], Aswathy Ashok Menon[3]

[1]Kauvery Hospital, Electronic City, Bangalore, Karnataka, India
[2]Manipal Hospitals, Kanakapura Road, Bangalore, Karnataka, India
[3]Neuberg Anand Reference Laboratory, Bangalore, Karnataka, India
*Correspondence

Abstract

A young male presented with a recurrent left thumb nail base swelling, that needed multiple procedures to be excised to negative margins, that was diagnosed to be a malignant glomus tumor which is a rare form of soft tissue neoplasm that tends to be locally aggressive.

Keywords: Malignant Glomus tumor; Soft tissue neoplasm

1. Case Presentation

A 23-year-aged male was first seen a year earlier at another center, with complaints of pain and swelling at the base of the left thumb. A mass was identified over the left thumb base. MRI of the part showed a well-defined well encapsulated lesion 1cm X 0.8 cm X 1.5 cm in the subungual plane on the dorsal aspect of the digital phalanx patient with T 1 hypo intensity and T2 hyper intensity and restricted diffusion. He was operated upon with excision of the mass by the treating surgeon. Histopathology of the excised tissue was reported elsewhere as necrotic fragments of crushed lymphoid or vasoformative tumor? Glomus tumor. Further testing or imaging was not done and patient was observed.

He later presented to us with similar complaints of a recurrent mass with pain and swelling over the left thumb base nearly a year after the first surgery. On examination, left thumb nail plate was deformed with a mass measuring 1.75 cm × 1.5 cm × 1.25 cm involving the nail bed including the germinal matrix and sterile matrix. Almost the whole of the nail bed was involved except the nail bed on the radial margin for 0.5 cm. MRI of the part was suggestive of again a subungual mass lesion? Recurrent Glomus Tumor.

He underwent excision of the nail bed tumor with nail bed graft. Postoperative histopathology was initially reported as malignant small round blue cell tumor with positive nail bed margins. Immunophenotype of the tumor showed patchy smooth muscle actin (SMA) positivity and focal paranuclear dot like positivity for synaptophysin. Tumor cells were negative for H Caldesmon, Desmin, CK, INSM1, NKX2.2, Myogenin and S-100. In view of a predominantly undifferentiated small blue cell morphology with the additional presence of necrosis, moderate nuclear atypia and brisk mitotic activity, a diagnosis of malignant glomus tumor was made.

Patient was then subjected to a full body staging investigation with PET CT scan to rule out metastatic disease given the history of recurrent disease and long course of illness.

Citation: Deepak K. L, Pragnya Coca, Aswathy Ashok Menon. Malignant Glomus Tumor: A rare and locally aggressive cancer in a young male. Kauverian Med J. 2025;2(11):22-26
Academic Editor: Dr. Venkita S. Suresh
ISSN: 2584-1572 (Online)

PET-CT was suggestive of only post procedural changes without any evidence of metastatic disease.

As the post-surgical margins were positive for tumor, and given the preponderance of the tumor for local recurrence, patient was once again subjected to wide excision of the nail matrix with full thickness skin grafting. Option of amputation of the digit was discussed with the patient. However, patient preferred a thumb sparing procedure. Postoperative histopathology showed residual tumor clusters within the proximal margin of the area of excision. Distal margin, ulna margin and radial margin were free from tumor cells.

The patient was again recalled for an excision the skin over the proximal margin with frozen biopsy control for tumor clearance.

Frozen section of the excised skin was once again reported as positive for malignancy. Again the patient was counseled for amputation of the digit, which he refused. However, the final histopathology review report of the excised skin bits was negative for malignancy. The patient is now on surveillance for recurrence.

Histopathology specimens of the tumor were sent for Next Generation Sequencing (NGS) for BRAF V600E mutation. No clinically relevant mutations were detected.

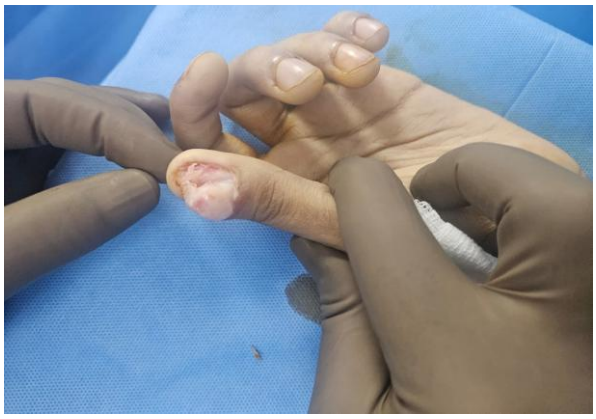

Fig (1): Clinical photograph of a left nail base mass preoperatively

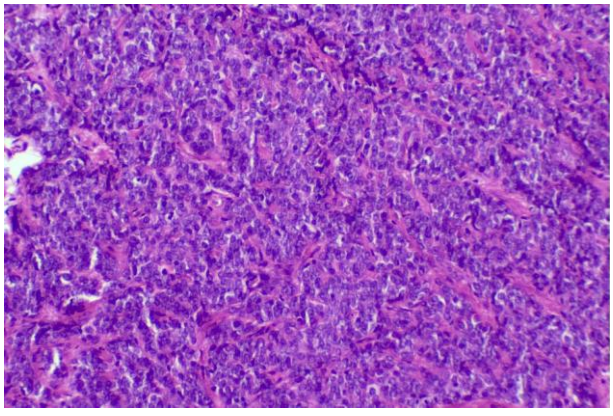

Fig (2): Compactly arranged tumor cells with small round blue cell morphology (H&E, 10x)

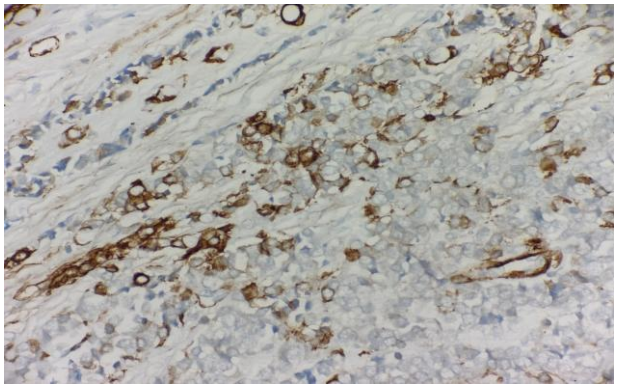

Fig (3): Tumor cells positive for SMA IHC (Smooth Muscle Actin) (H&E 10x)

2. Discussion

Malignant glomus tumors are exceedingly rare soft tissue neoplasms comprising of about 2.9% of all glomus tumors. They typically arise from the neuromyoarterial glomus body, most common in distal extremities but can occur viscerally (e.g., lung, kidney, breast).

In a case series of five patients by Wood et al [1], both males and females were equally represented, with an age distribution of 9 to 49 years.

They have been classified by Folpe et al. [2] as malignant glomus tumors, symplastic glomus tumor, glomus tumor of uncertain malignant potential (GT-UMP) or glomangiomatosis.

This classification forms the basis for the current WHO diagnostic criterion for malignant glomus tumors. Histological markers for malignancy include atypical mitoses and marked nuclear atypia regardless of mitotic activity. These malignant cells show intense staining for vimentin with retained smooth muscle actin (SMA) and muscle-specific action.

In a study of 102 patients with glomus tumors by Dashti et al [3], a BRAF V600E mutation was detected in 6% of patients, all of whom were malignant or GT-UMP. This mutation may be associated with a malignant phenotype. In patients with progressive disease, BRAF could be a promising therapeutic target.

Malignant glomus tumors tend to be locally aggressive and have the potential to metastasize rarely. When metastasis is seen, it is often fatal. [4]

In the same case series by Wood et al, four out of five patients required re-resection after initial surgery done elsewhere, like in our case. Recurrence and metastasis each occurred in ~20 % of their patients.

Wide local excision with negative margins is the primary treatment for malignant glomus tumors. Data for adjuvant radiation is limited; one case with positive margins in the same case series, showed late recurrence despite radiotherapy, suggesting minimal long-term benefit. Chemotherapy has been rarely used; regimens typically similar to those for soft-tissue sarcomas—but no standardized protocol due to scarcity of cases.

We could only find only one other case report of malignant glomus tumor in the extremity from India by Kumar et al [5], where the presentation was similarly a painful mass on the finger, that was recurrent and excised completely.

While the overall prognosis of completely excised tumors is good, local recurrence is common with inadequate excision. Metastasis is rare but usually associated with poor prognosis.

References

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