Urology Case Reports 55 (2024) 102774

Contents lists available at ScienceDirect

Urology Case Reports

journal homepage: www.elsevier.com/locate/eucr

Oncology

Glomus tumor with malignant features: A case report and review of the literature

Andrew Allen *, Andrew Watts, Isaac Melin, Peter Langenstroer

Department of Urology, Medical College of Wisconsin, Milwaukee, WI, USA

ARTICLE INFO

Keywords:
Glomus tumor
Malignant renal neoplasm
Nephrectomy

ABSTRACT

Glomus tumors are rare mesenchymal neoplasms of the subcutaneous tissue, most frequently found in the distal extremities. They are typically benign, but malignant glomus tumors have been described in the literature. Here we present a patient found to have a unilateral renal mass with pathology displaying a primary renal glomus tumor with malignant features. Review of the literature reveals only three cases of malignant glomus tumors and five glomus tumors with malignant potential. As such, previous initial presentations, current criteria for glomus tumor malignancy, and previous treatment outcomes of these cases were reviewed.

1. Introduction

Glomus tumors are rare mesenchymal neoplasms that make up less than 2% of all soft tissue tumors and are most often seen in young adults.[1] They resemble modified smooth muscle cells of the normal glomus body, and as such, are typically found at sites with abundant glomus bodies including the skin and soft tissues.[2] Visceral organs are especially infrequent sites for glomus tumor growth, as glomus bodies are not typically present in these tissues. Most glomus tumors are benign, with just 1% of reported cases meeting criteria for malignancy.[1] Assessment of the current literature reveals only eight cases of primary renal glomus tumors with any malignant characteristics.[3–9] In this report, we discuss the case of a renal mass that was pathologically identified as a glomus tumor with malignant features.

2. Case presentation

A 69-year-old female presented to a community hospital for evaluation following routine lab work notable for elevated liver function tests. Ultrasound incidentally identified a nearly 8 cm, mildly vascular, heterogeneous mass in the mid-lower pole of the right kidney. MRI of the abdomen without contrast showed a solid enhancing right lower pole mass measuring 7.8 x 7.0 x 5.6 cm with areas of necrosis (Fig. 1). CT of the chest was also ordered to complete the workup, which was negative for any metastatic disease. The patient subsequently underwent right robot-assisted radical nephrectomy without any notable intraoperative complications. The patient was discharged on postoperative day one following an uneventful hospitalization.

Pathologic review showed a nodular tumor composed of expanded nests to solid sheets of cells with clear cytoplasm and accentuated cytoplasmic membranes. The nuclei were generally round and uniform with some smaller cells with conspicuous atypia and greatly increased mitotic activity (up to 30 mitoses per 10 HPF). Prominent blood vessels were present, surrounding and embedded within nests of tumor cells. Areas of myxoid and fibrotic background were also present (Fig. 2). The immunohistochemical panel revealed the tumor cells were positive for SMA, calponin, desmin, vimentin, and cyclin D1. There was weak focal staining for synaptophysin and CD10. All remaining stains were negative. Given the histologic features, SMA expression, and other exclusionary stains, our pathology colleagues reported this to be most consistent with a diagnosis of glomus tumor with malignant features.

The patient most recently completed a 21-month postoperative follow up visit. She is now over two years out from initial presentation and is in good health at this time. All follow-up MRIs have shown no concern for local recurrence or metastatic disease.

3. Discussion

There is an exceptionally limited number of primary renal glomus tumor cases. Our review of the literature yielded approximately 30 total cases, only eight of which displayed any malignant features. Initial presentation of primary renal glomus tumors appears to vary significantly. Glomus tumors are most often diagnosed incidentally during imaging obtained for other reasons.[4] Reports note microscopic

* Corresponding author.
E-mail address: anallen@mcw.edu (A. Allen).

https://doi.org/10.1016/j.eucr.2024.102774
Received 13 June 2024; Accepted 20 June 2024
Available online 21 June 2024

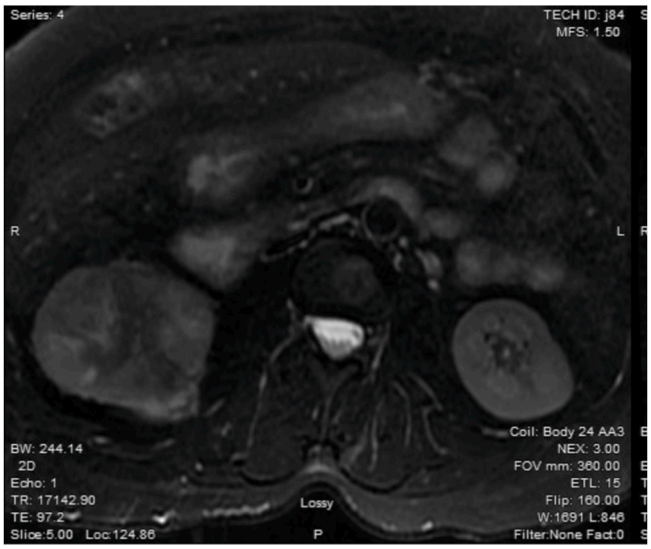

Fig. 1. Abdominal MRI: MRI Abdomen without contrast showing a solid enhancing right lower pole mass measuring 7.8 x 7.0 x 5.6 cm.

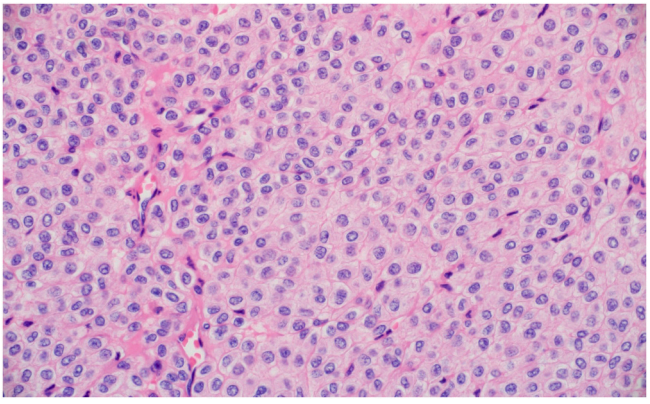

Fig. 2. Glomus Tumor: High power microscopy revealed pathology consistent with typical glomus tumor.

hematuria,[6–8] gross hematuria and flank pain,[3] and metastatic sources of pain[5] as reasons for initial patient presentation. There does not appear to be a difference in presenting symptoms for benign tumors compared to tumors with malignant features.[10]

There are no established radiological features of renal glomus tumors, given they present as heterogeneous, nonspecific enhancing lesions. No cases have undergone preoperative biopsy, so diagnosis has been made post-operatively with histological and immunochemical analysis. This typically shows modified perivascular smooth muscle cells arranged in sheets and nests.[1] In accordance with guidelines for management of renal masses, treatment of these lesions is contingent on size, consisting primarily of partial[7,8] or total nephrectomy,[3,4,6] both of which have been shown to be successful.

As previously mentioned, there are only eight reported cases of primary renal glomus tumors harboring malignant features. Of these cases, three were labeled as malignant,[3–5] while five were described as having uncertain malignant potential.[3,6–9] When diagnosing glomus tumors, Folpe and colleagues originally suggested that the term "malignant glomus tumor" should be reserved for "lesions with a marked risk of metastasis." As such, this requires meeting at least one of the following parameters: size >2 cm, deep location, atypical mitotic figures, or moderate to high-grade nuclear atypia, and 5 or more mitoses per 50 HPFs.[11]

Malignancy criteria of glomus tumors was later revised in the 2013 WHO Classification of Tumors of Soft Tissue and Bone, where it was decided that marked nuclear atypia or atypical mitotic figures satisfied requirements for malignancy, while size and location features would only meet requirements for "tumor of uncertain malignant potential."[12] The glomus tumor of our patient met several of these criteria: conspicuous atypia and greatly increased mitotic activity (30 per 10 HPF), large size (8.9 x 6.0 x 5.5 cm) and deep location.

Due to the scarce number of cases, there are no established criteria for defining glomus tumors of the kidney as malignant. Some reports have suggested that metastasis should be considered the only defining feature of malignancy.[10] Likewise, other sources have emphasized that despite meeting the size, depth, and mitotic activity criteria for a malignant glomus tumor of the skin and soft tissue, there is inadequate data on primary renal glomus tumors and therefore diagnosis of malignant glomus tumor should be cautioned.[6,13] As such, without coexisting metastasis, diagnosis of glomus tumor with uncertain malignant potential may be favored in this case. However, there is one case of a glomus tumor with extensive metastasis labeled as a tumor with uncertain malignant potential.[8] The authors elected to make this specific diagnosis secondary to a lack of atypical mitotic features, 1 mitosis per 50 HPF, and successful clinical response to temozolomide treatment, noting disease stability 18 months later.

Considering the limited data, renal glomus tumor diagnosis with any concern for possible malignancy warrants close postoperative follow-up. Our patient is currently two years post-op and has remained otherwise healthy with normal follow-up imaging. There are currently no case reports of glomus tumors with uncertain malignant potential that include postoperative follow up greater than two years.

4. Conclusion

To our knowledge, this case is consistent with the longest follow-up of a renal glomus tumor with uncertain malignant potential, following radical nephrectomy. Given this uneventful postoperative course, lack of disease recurrence, and no metastasis, we conclude that glomus tumor "harboring malignant potential" is likely the most appropriate diagnosis in this case.

Funding

This research did not receive any specific grant from funding agencies in the public, commercial, or not-for-profit sectors.

CRediT authorship contribution statement

Andrew Allen: Investigation, Writing – original draft. **Andrew Watts:** Writing – review & editing. **Isaac Melin:** Writing – review & editing. **Peter Langenstroer:** Supervision, Writing – review & editing.

References

1. Gombos Z, Zhang PJ. Glomus tumor. *Arch Pathol Lab Med.* 2008;132:1448–1452.
2. Weiss SW, Goldblum JR, Enzinger FM. *Enzinger and Weiss's Soft Tissue Tumors*. fourth ed. St. Louis: Mosby; 2001.
3. Li R, Petros FG, Davis CJ, et al. Characterization of glomus tumors of the kidney. *Clin Genitourin Cancer.* 2018;16:e253–e256.
4. Chen Y-A, Li H-N, Wang R-C, et al. Malignant glomus tumor of the kidney: a case report and review of the literature. *Clin Genitourin Cancer.* 2017;15:e151–e153.
5. Lamba G, Rafiyath SM, Kaur H, et al. Malignant glomus tumor of kidney: the first reported case and review of literature. *Hum Pathol.* 2011;42:1200–1203.
6. Gill J, Van Vliet C. Infiltrating glomus tumor of uncertain malignant potential arising in the kidney. *Hum Pathol.* 2010;41:145–149.
7. Zhao M, Yang M, Gu W, et al. Glomus tumor of the kidney in a Child with Tuberous Sclerosis. *Pediatr Dev Pathol.* 2020;23:230–234.
8. Kapogiannis F, Tsiampa E. Glomus tumor of the kidney harboring malignant potential. *Cureus.* 2021;13, e19479.
9. Vasilevska-Nikodinovska V, Samardjiski M, Jovanovik R, et al. Low-grade malignancy glomus tumor in a Setting of multiple glomus tumors - case report. *Open Access Maced J Med Sci.* 2019;7:4082–4088.

10. Bhardwaj S. Haines Iii GK and Si Q: primary renal glomus tumor with concurrent papillary renal cell carcinoma and multiple papillary adenomas in a patient with end stage renal disease: a case report and clinicopathologic analysis. *Int J Clin Exp Pathol*. 2022;15:459–466.
11. Folpe AL, Fanburg–Smith JC, Miettinen M, et al. Atypical and malignant glomus tumors: analysis of 52 cases, with a Proposal for the Reclassification of glomus tumors. *Am J Surg Pathol*. 2001;25:1–12.
12. Organisation mondiale de la santé and Centre international de recherche sur le cancer. In: *WHO Classification of Tumours of Soft Tissue and Bone*. fourth ed. Lyon: International agency for research on cancer (IARC) press; 2013.
13. Sirohi D, Smith SC, Epstein JI, et al. Pericytic tumors of the kidney—a clinicopathologicanalysis of 17 cases. *Hum Pathol*. 2017;64:106–117.