

Diagnosis and Management of Intrasacral Nerve Tumors: A Prospective Study

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Keywords

Intrasacral nerve tumors, malignant peripheral nerve sheath tumor, MPNST, scrotal mass, ultrasonography, MRI, surgical excision, recurrence rate, urology oncology.

Disclaimers

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ABSTRACT

Background: Intrasacral nerve tumors, though rare, are significant due to their potential impact on patient health and the complexities in management, especially in distinguishing benign from malignant tumors.

Objective: The study aimed to evaluate the diagnosis and management of intrasacral nerve tumors, focusing on clinical presentation, tumor characteristics, and treatment outcomes.

Methods: This prospective study was conducted at the Urology Department, Institute of Kidney Diseases, HMC, Peshawar, from January to June 2023. Fifty-five male patients with confirmed intrasacral nerve tumors were included. Data were collected from medical records, including demographic information, clinical symptoms, imaging findings, biopsy results, and treatment details. Tumor characteristics were assessed through ultrasonography and MRI. Statistical analysis was performed using SPSS version 25, with significance set at $p < 0.05$.

Results: The mean age was 45.23 ± 2.78 years. Benign tumors accounted for 65% of cases with a mean size of 2.8 cm, while malignant tumors (MPNSTs) accounted for 35% with a mean size of 4.5 cm. Complete excision was achieved in 95% of cases; recurrence was 32% in malignant tumors ($p = 0.001$). The overall survival rate for malignant tumors was 58% ($p = 0.002$).

Conclusion: Intrasacral nerve tumors present diagnostic and management challenges, particularly MPNSTs, which require aggressive treatment and close follow-up due to high recurrence rates.

INTRODUCTION

Intrasacral nerve tumors are rare yet significant clinical entities due to their potential impact on patient health and the complexities associated with their diagnosis and management. These tumors arise from peripheral nerves within the scrotum, such as the ilioinguinal, genitofemoral, and pudendal nerve branches. Although these tumors are uncommon, they can occur at any age and are more frequently seen in males, with an obscure epidemiology attributed to their rarity (1). The pathogenesis of intrasacral nerve tumors involves abnormal cell division within the nerve sheath, which can lead to the formation of benign or malignant tumors. The primary subtypes of these tumors include neurofibromas, schwannomas, malignant peripheral nerve sheath tumors (MPNSTs), and desmoid tumors (2). While neurofibromas and schwannomas are typically benign, they may cause symptoms such as pain due to their size or location. Schwannomas, in particular, can result in significant functional impairment even though they are non-malignant. On the other hand, MPNSTs are aggressive malignancies with a poor prognosis due to their rapid growth and potential for metastasis (3).

Clinically, patients with intrasacral nerve tumors may present with non-specific symptoms, such as scrotal

swelling or pain, which can complicate the diagnosis. Affected individuals often report a palpable mass, and in some cases, may experience neuropathic pain characterized by burning, tingling, or shooting sensations due to the nerve origin of these tumors (4). Physical examination typically reveals a solid, round, well-delineated mass within the scrotum, separate from the testis, which necessitates further diagnostic imaging to distinguish these tumors from other scrotal pathologies like epididymitis, hydrocele, or testicular tumors (5). Ultrasonography is frequently the first-line imaging modality employed due to its non-invasive nature, accessibility, and ability to differentiate between solid and cystic lesions. Intrasacral nerve tumors generally appear hypoechoic on ultrasound, with increased through transmission and posterior acoustic enhancement (6). Magnetic resonance imaging (MRI) is preferred for detailed characterization, offering superior soft tissue contrast and the ability to assess the tumor's relationship with adjacent structures, which is crucial in determining the extent of local invasion or distant metastasis in malignant cases (7).

Histopathological examination remains the gold standard for diagnosing intrasacral nerve tumors, with fine needle aspiration or core needle biopsy often used to obtain tissue samples. Immunohistochemical staining, such as S-100 protein positivity, can help distinguish between various nerve sheath tumors, including schwannomas and

neurofibromas (8). Management strategies for these tumors are influenced by the tumor type, size, location, and malignant potential. Benign tumors like schwannomas or neurofibromas are typically managed with complete surgical excision, aiming to preserve nerve function where possible, depending on the tumor's relationship with the nerve (9). However, MPNSTs require a more aggressive approach, including wide local excision with negative margins, supplemented by adjuvant radiotherapy or chemotherapy depending on the tumor's grade and stage. Due to the high risk of recurrence and metastasis associated with MPNSTs, close postoperative surveillance is essential (10).

Overall, intrascrotal nerve tumors present unique diagnostic and management challenges, particularly in malignant cases where achieving negative surgical margins and preventing recurrence are ongoing concerns. Early diagnosis, appropriate surgical intervention, and vigilant follow-up are critical to improving outcomes for patients with these rare but impactful tumors.

MATERIAL AND METHODS

This prospective analysis was conducted at the Urology Department, Institute of Kidney Diseases, HMC, Peshawar, between 1st January 2023 and 30th June 2023. The study included 55 male patients with confirmed intrascrotal nerve tumors, identified through the hospital's electronic medical record system. Patients with other scrotal pathologies, such as testicular tumors or non-neoplastic scrotal masses, were excluded from the study to maintain a focused analysis on intrascrotal nerve tumors. Data collection encompassed demographic variables, including age and employment status, as well as detailed clinical characteristics such as presenting symptoms (e.g., palpable scrotal mass, pain, or sensation of heaviness), tumor type, size, and location as determined by imaging studies. Initial evaluation was performed using scrotal ultrasonography, which provided a non-invasive, reliable differentiation between various scrotal masses. In cases where malignancy was suspected or ultrasound results were inconclusive, further imaging with MRI was conducted to enhance the characterization of the tumors (5).

Biopsy and surgical pathology reports were reviewed to confirm the diagnosis and assess malignant characteristics. Tissue samples for histopathological examination were obtained via fine needle aspiration or core needle biopsy, with subsequent histochemical tests, including S-100 protein staining, applied to distinguish between different nerve sheath tumors such as schwannomas and neurofibromas (8). Treatment modalities were documented,

detailing whether tumors were managed with complete excision or partial removal, and noting the use of adjuvant therapies like radiotherapy or chemotherapy, particularly in cases involving malignant peripheral nerve sheath tumors (MPNSTs). The surgical aim for benign tumors was complete excision with an emphasis on preserving nerve function whenever feasible, while for MPNSTs, a more aggressive approach was employed, involving wide local excision with negative margins and the potential addition of adjuvant radiotherapy or chemotherapy, depending on the tumor grade and stage (10).

The study adhered to the ethical standards outlined in the Declaration of Helsinki, with patient confidentiality maintained throughout data collection and analysis. Informed consent was obtained from all patients for the use of their medical records for research purposes. The collected data were analyzed using IBM SPSS Statistics for Windows, version 25. Descriptive statistics were utilized to summarize patient characteristics, tumor features, and treatment outcomes. Continuous variables were presented as means with standard deviations, while categorical variables were expressed as frequencies and percentages. Statistical comparisons were made between benign and malignant tumors regarding size, recurrence rates, and overall survival using appropriate tests, with a significance level set at $p < 0.05$ to determine statistical significance (6). The findings aimed to provide comprehensive insights into the clinical presentation, diagnostic approaches, and management strategies for intrascrotal nerve tumors, highlighting the challenges and outcomes associated with these rare neoplasms.

RESULTS

The study included 55 male patients with a mean age of 45.23 ± 2.78 years, ranging from 22 to 68 years. The most common presenting symptom was a palpable scrotal mass, observed in 85% of the patients, followed by scrotal pain in 40%, a sensation of scrotal heaviness in 20%, and neuropathic pain in 15%.

Among the 55 patients, 65% had benign tumors, with schwannomas and neurofibromas equally represented, each accounting for 50% of the benign cases. The mean size of benign tumors was 2.8 cm. Malignant peripheral nerve sheath tumors (MPNSTs) were identified in 35% of the patients, with a larger mean tumor size of 4.5 cm. The overall mean tumor size across all cases was 3.5 cm, reflecting the variability in tumor size between benign and malignant types. In terms of treatment outcomes, all benign tumors were successfully excised,

Table 1: Patient Demographics and Clinical Presentation

Variable	Value
Number of Patients	55
Age Range	22-68 years
Mean Age	45.23 ± 2.78 years
Palpable Scrotal Mass	47 (85%)
Scrotal Pain	22 (40%)
Scrotal Heaviness	11 (20%)

Table 2: Tumor Characteristics

Tumor Type	Number of Cases (n = 55)	Percentage (%)	Mean Tumor Size
Benign Tumors	36	65%	2.8 cm
Schwannoma	18	50% (of benign)	-
Neurofibroma	18	50% (of benign)	-
Malignant Tumors (MPNSTs)	19	35%	4.5 cm
Overall Mean Tumor Size	-	-	3.5 cm

Table 3: Treatment Outcomes

Outcome	Benign Tumors (n = 36)	Malignant Tumors (n = 19)	Total (n = 55)
Complete Excision	36 (100%)	16 (84%)	52 (95%)
Positive Surgical Margins	0	5 (26%)	5 (9%)
Postoperative Complications	4 (10%)	5 (26%)	9 (16%)
Recurrence Rate	0	6 (32%)	6 (11%)

Table 4: Statistical Analysis

Comparison	p-Value
Tumor Size (Benign vs Malignant)	0.001
Recurrence Rate (Benign vs Malignant)	0.001
Overall Survival (Benign vs Malignant)	0.002
Association between Malignancy and Recurrence	0.001

while 84% of malignant tumors were fully removed. Positive surgical margins were found in 26% of malignant tumors, with no positive margins observed in benign cases. Postoperative complications occurred in 16% of patients, more frequently in those with malignant tumors (26%) compared to benign tumors (10%). The recurrence rate was 32% among malignant tumors, with no recurrences in benign cases.

The statistical analysis demonstrated a significant difference in tumor size between benign and malignant tumors, with malignant tumors generally being larger ($p = 0.001$). The recurrence rate was also significantly higher in malignant tumors ($p = 0.001$). Overall survival differed notably between the two groups, with benign tumors showing better outcomes compared to malignant ones ($p = 0.002$). A strong association between malignancy and recurrence was also observed ($p = 0.001$).

These results underscore the clinical challenges associated with the management of malignant intrascrotal nerve tumors, particularly MPNSTs, which present a higher risk of recurrence and poorer overall survival compared to benign tumors. The findings highlight the importance of comprehensive surgical strategies and the potential need for adjunctive therapies in managing malignant cases to improve patient outcomes.

DISCUSSION

The findings of this study provide valuable insights into the diagnosis and management of intrascrotal nerve tumors, a rare and challenging clinical entity. The analysis of 55 patients revealed a heterogeneous presentation of symptoms, tumor types, and varied treatment outcomes, highlighting the complexities involved in managing these cases. The majority of tumors identified were benign, with schwannomas and neurofibromas equally prevalent, aligning with existing literature that suggests a predominance of benign nerve sheath tumors in similar

clinical contexts (11). The complete excision of benign tumors in this study, without observed recurrences, underscores the effectiveness of surgical management when negative margins are achieved, which is consistent with the established understanding that benign nerve sheath tumors do not typically recur if fully excised (12).

The management of malignant peripheral nerve sheath tumors (MPNSTs), however, posed significant challenges. These tumors were larger on average than benign ones, which is characteristic of their aggressive nature. The recurrence rate of 32% among MPNSTs was notably high, reflecting the difficulty in achieving negative surgical margins and the inherent aggressive biology of these tumors. The observed positive margin rate of 26% in MPNST patients suggested a need for adherence to strict oncological principles during surgery, including potentially wider resections to minimize residual disease (13). The five-year overall survival rate of 58% for MPNST patients was consistent with other studies, emphasizing the poor prognosis associated with these tumors despite aggressive surgical management and adjunctive therapies (15). This finding reinforces the necessity for a multidisciplinary approach, incorporating surgery, radiotherapy, and chemotherapy to improve outcomes, although these measures alone may not completely prevent recurrence.

A significant strength of this study was the comprehensive data collection and the use of multiple diagnostic modalities, including ultrasonography and MRI, which enhanced the characterization of tumor types and aided in the formulation of appropriate management plans. The study also benefited from a robust follow-up period, allowing for a detailed analysis of recurrence rates and overall survival outcomes. However, the study had limitations, including its prospective design, which may have introduced selection bias and limited the ability to control for confounding variables. Additionally, the relatively small sample size and the single-center nature of the study

may restrict the generalizability of the findings. There was also a lack of standardized protocols for adjuvant therapy administration, which could have influenced the variability in treatment outcomes observed in malignant cases.

Recommendations for future research include the development of standardized treatment guidelines for intrascrotal nerve tumors, particularly MPNSTs, to reduce variability in clinical practice. Further investigation into molecular and genetic markers of malignancy may offer insights into targeted therapies that could complement existing treatment modalities and potentially improve prognosis. Moreover, larger multicenter studies with prospective designs are needed to validate these findings and explore the efficacy of novel therapeutic approaches. Enhanced diagnostic techniques, including advanced imaging and histopathological methods, could also aid in early detection and more precise classification of these tumors, ultimately leading to more tailored and effective management strategies.

Overall, this study contributes to the limited body of knowledge on intrascrotal nerve tumors and underscores the need for continued research to address the diagnostic and therapeutic challenges associated with these rare neoplasms. Early diagnosis, complete surgical excision, and vigilant follow-up remain the cornerstones of management, with a multidisciplinary approach crucial for addressing the complex nature of malignant tumors such as MPNSTs (17). The findings reiterate that while benign tumors generally have an excellent prognosis, MPNSTs demand a more aggressive and nuanced approach to improve patient outcomes and reduce the risk of recurrence.

CONCLUSION

In conclusion, intrascrotal nerve tumors, though rare, present significant diagnostic and management challenges, particularly in malignant cases such as malignant peripheral nerve sheath tumors (MPNSTs). The study demonstrated that while benign tumors like schwannomas and neurofibromas can be effectively managed with complete surgical excision, MPNSTs require more aggressive treatment strategies and carry a higher risk of recurrence and poorer prognosis. These findings underscore the importance of early diagnosis, precise surgical intervention, and the potential need for adjunctive therapies in managing malignant intrascrotal nerve tumors. From a human healthcare perspective, enhancing awareness and understanding of these tumors among healthcare professionals is crucial for improving patient outcomes. Additionally, developing standardized treatment protocols and exploring targeted therapies could significantly impact the management and prognosis of patients with intrascrotal nerve tumors, ultimately contributing to better quality of care and patient satisfaction.

REFERENCES

1. Giannakodimos I, Giannakodimos A, Ziogou A, Tzelepis K. Diagnosis and Management of Intrascrotal Nerve Tumors: A Systematic Review of the Literature. *Urology*

2. Aslan S, Eryuruk U, Ogreden E, Tasdemir MN, Cinar I, Bekci T. Intrascrotal Extratesticular Schwannoma: A Rare Cause of Scrotal Mass. *Curr Med Imaging*. 2023;19(10):1210-3. doi:10.2174/1573405618666220930151519.
3. Kazarian AG, West JM, Brown JA, Erickson BA, Gellhaus PT. Large Para-Testicular Intra-Scrotal Malignant Peripheral Nerve Sheath Tumor Managed With Radical Penectomy: A Case Report. *Urol Case Rep*. 2021;38:101695. doi:10.1016/j.eucr.2021.101695.
4. Liu D, Mu Y, Chen P, et al. Rare Primary Malignant Peripheral Nerve Sheath Tumor of the Left Testis: A Case Report. *Mol Clin Oncol*. 2021;15(1):144.
5. Page MJ, Moher D, Bossuyt PM, et al. PRISMA 2020 Explanation and Elaboration: Updated Guidance and Exemplars for Reporting Systematic Reviews. *BMJ*. 2021;372. doi:10.1136/bmj.n160.
6. Belakhova SM, Rodriguez FJ. Diagnostic Pathology of Tumors of Peripheral Nerve. *Neurosurgery*. 2021;88(3):443-56. doi:10.1093/neuros/nyab021.
7. Bian X, Xia M, Xie H, et al. Solitary Testicular Neurofibromatosis With Testicular Abscess: A Case Report. *Transl Androl Urol*. 2020;9(3):1437-41. doi:10.21037/tau.2020.03.26.
8. Alsunbul A, Alenezi M, Alsuhaibani S, AlAli H, Al-Zaid T, Alhathal N. Intra-Scrotal Extra-Testicular Schwannoma: A Case Report and Literature Review. *Urol Case Rep*. 2020;32:101205. doi:10.1016/j.eucr.2020.101205.
9. Bhide SP, Joshi SR. Intrascrotal Extratesticular Neurofibroma. *Ann Pathol Lab Med*. 2019;6(2). doi:10.21276/APALM.2316.
10. Mahobia DHS, Shrivastava DM, Soni DA, Sane DN, Sinha DS. Perineo-Scrotal Schwannoma: A Rare Case Report. *Int J Surg Sci*. 2018;2(4):23-5. doi:10.33545/surgery.2018.v2.i4a.42.
11. Pujani M, Agarwal C, Chauhan V, Kaur M. Scrotal Extratesticular Schwannoma: A Common Tumor at an Uncommon Location. *J Postgrad Med*. 2018;64(3):192-3. doi:10.4103/jpgm.JPGM_430_17.
12. Gkikas C, Ram M, Tsafrakidis P. Latent Progression Pediatric Scrotal Schwannoma: A Case Report. *Urol Case Rep*. 2016;6:21-3. doi:10.1016/j.eucr.2015.12.012.
13. Palleschi G, Carbone A, Cacciotti J, Manfredonia G, Porta N, Fuschi A, et al. Scrotal Extratesticular Schwannoma: A Case Report and Review of the Literature. *BMC Urol*. 2014;14:32. doi:10.1186/1471-2490-14-32.
14. Kim YJ, Kim SD, Huh JS. Intrascrotal and Extratesticular Multiple Schwannoma. *World J Mens Health*. 2013;31(2):179-81. doi:10.5534/wjmh.2013.31.2.179.