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Surgical Approach for a Gluteal Neurofibroma

AUTHORS:
Lemons W[a]; Markovich B[b]; Lindsay B[c]; Murken D[d]

CORRESPONDING AUTHOR:
Wesley Lemons, MD
VCU School of Medicine
1200 E. Broad Street
9th Floor
Box 980153
Richmond, VA 23298
Email: wesley.lemons@vcuhealth.org

AUTHOR AFFILIATIONS:
a. West Virginia University School of Medicine
J.W. Ruby Memorial Hospital
Morgantown, WV 26506

b. Department of Radiology
West Virginia University School of Medicin
J.W. Ruby Memorial Hospital
Morgantown, WV 26506

c. Department of Orthopedic Surgery
West Virginia University School of Medicine
J.W. Ruby Memorial Hospital
Morgantown, WV 26506

d. Department of Surgery
West Virginia University School of Medicine
J.W. Ruby Memorial Hospital
Morgantown, WV 26506

Background	Localized neurofibromas typically demonstrate a benign course and are often followed unless the patient desires removal for cosmetic reasons or symptoms related to mass effect. The workup of neurofibromas largely consists of some form of biopsy to confirm the diagnosis. After the diagnosis is confirmed via histology, the tumor can either be surgically removed or followed until the patient requests removal. The surgical removal of these tumors can be quite complex due to the local invasion of surrounding structures.
Summary	A 74-year-old male presented with an enlarging right gluteal mass that he noticed several months prior following a fall from standing. The mass continued to enlarge during the next several months resulting in impingement on pelvic structures leading to need for surgical evaluation. Preoperative biopsy was consistent with neurofibroma. The patient subsequently underwent resection of the mass by a team of multiple specialists from fields including urology, orthopedic oncology, and colorectal surgery. This case highlights the crucial role that preoperative imaging, and specifically interdisciplinary imaging review, plays in building an operative team and directing its surgical approach.
Conclusion	The surgical approach to resection can be complicated by the locally advanced nature of neurofibromas. Interdisciplinary review of high-quality preoperative imaging is vital for surgical planning. The distortion of pelvic tissue planes and the presence of a "levator ani cap" on magnetic resonance imaging in this case supported the need for a team of surgical subspecialties and the feasibility of a pure perineal approach.
Key Words	mass; surgery

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Case Description

A 74-year-old male patient presented to his primary care physician with an enlarged, painful mass in his right buttock that he believed was a hematoma resulting from a fall (which occurred a few months prior to presentation). The patient was referred to general surgery for possible drainage, but ultrasound was concerning for a mass. An ultrasound-guided biopsy was consistent with a spindle cell proliferation, indicating a peripheral nerve sheath tumor (PNST). The patient was referred to orthopedic surgery and underwent an magnetic resonance imaging (MRI) of the pelvis, which revealed a large 12 × 10 × 16 cm heterogeneous T2 bright mass centered within the right buttocks and extending into the right ischioanal fossa. On imaging, the mass was observed to exert leftward on the rectum and the base of the prostate and was in close approximation to the right inferior pubic symphysis and the right corpus cavernosum. An interdisciplinary surgical team consisting of an orthopedic surgeon, colorectal surgeon, and urologist assembled and planned to resect the mass. The patient was counseled as to the benefits of resection as well as the risks including positive margins and/or need for abdominoperineal resection and colostomy, prostatectomy, or penectomy.

Discussion

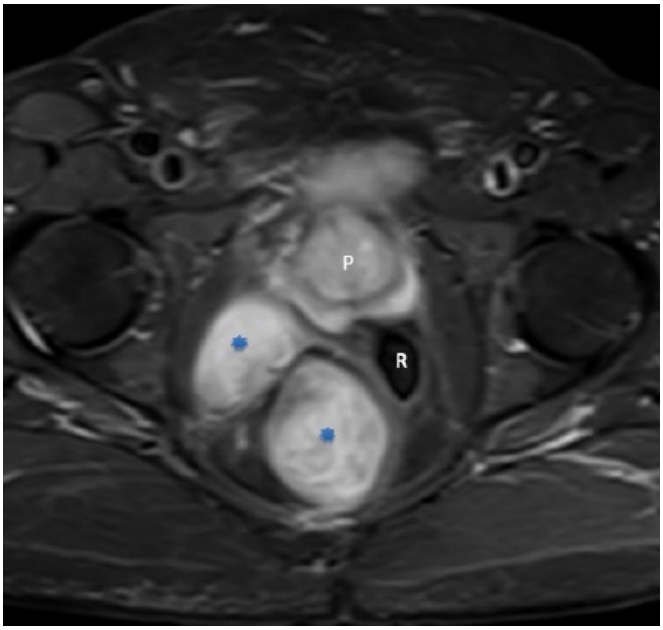
Neurofibromas are the most common type of PNST.[1] Neurofibromas arise from the endoneurium and other nearby connective tissues of the peripheral nerve and expand outward exerting mass effect on nearby structures.[1] In this case, the patient presented with a sporadic localized tumor with a low risk of malignant transformation.

Neurofibromas typically present as an asymptomatic cosmetic distortion to areas such as the neck, chest, back and extremities, although they can also present as a symptomatic, enlarging palpable mass.[1,2] Core needle or excisional biopsy is usually required for definitive diagnosis.[3] Histological examination will demonstrate scattered spindled cells with poorly defined borders in the background of a collagenous matrix with mast cells.[1,4] The nuclei of these tumors are often described as small, hyperchromatic and wavy with minimal mitoses present.[1] Immunohistochemical evaluation demonstrating CD 34, Myelin basic protein and S100 (in 50% of tumors) positivity can further aid in diagnosis of neurofibromas.[1,5]

Localized neurofibromas have an excellent prognosis. In many cases, the tumors are clinically monitored; however surgical excision is recommended if the patient is experiencing undesirable cosmetic changes or symptoms including pain/discomfort from mass effect or peripheral neurologic symptoms that range from nerve pain to gait disturbances and weakness.[4]

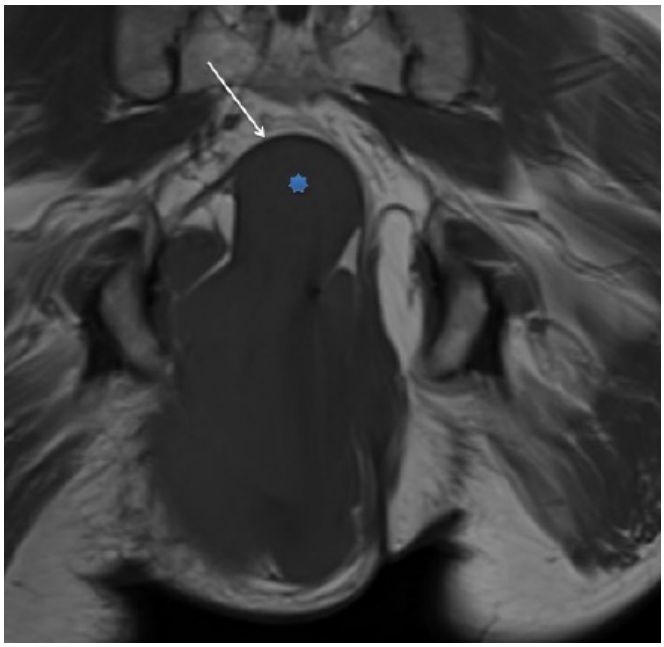
In this case, surgery was indicated due to the patient's symptom of pain and the proximity of the mass to adjacent pelvic sutures. While there was no evidence of involvement or invasion of adjacent structures on imaging, the proximity to the anorectum and genitourinary system warranted sub-specialty involvement. The questions of best surgical approach and how to balance complete R0 resection with increased morbidity remained.

Figure 1. Axial Fat-Suppressed T2-weighted MRI. Published with Permission



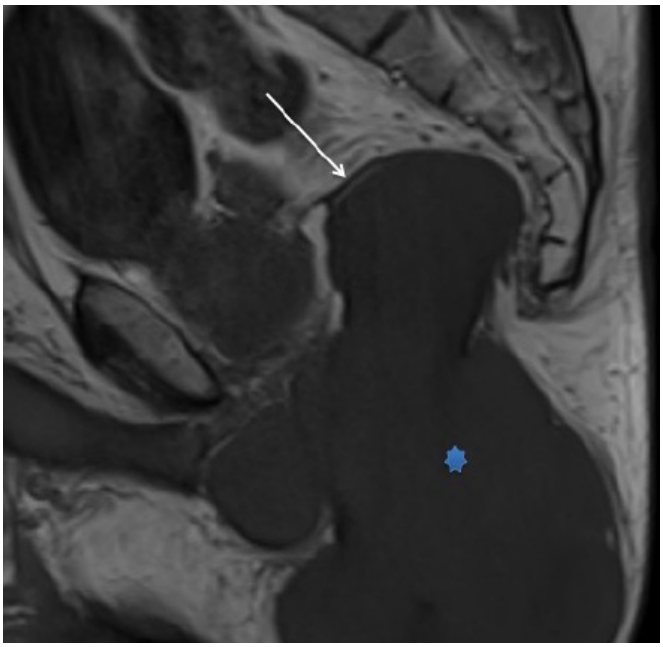
The large pelvic neurofibroma (asterisk) causes mass effect and leftward displacement of the rectum (R). The mass also distorts the right peripheral zone of the prostate (P) without evidence of tumor invasion.

Figure 2. Sagittal Fat-Suppressed T2-weighted MRI. Published with Permission



The large pelvic neurofibroma (asterisks) causes an abutment of the urethra (white arrow).

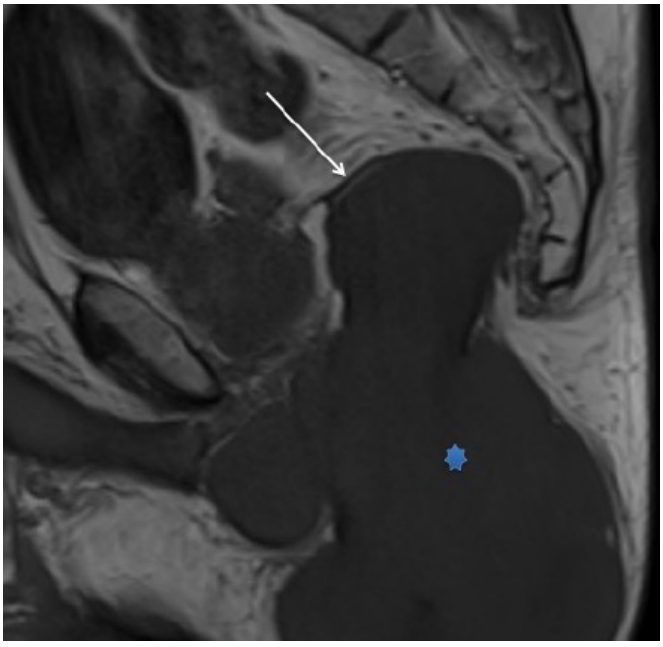
Figure 3. Sagittal Nonfat-Suppressed T1-Weighted MRI. Published with Permission



The large pelvic neurofibroma (asterisk) distorts and displaces the levator ani muscle (white arrow).

Preoperatively, the surgical team met to choose the best positioning and approach. There was some concern, based on the proximal extent of the tumor, that a combined transabdominal transperineal approach would be needed. The question of whether a minimally invasive or robotic approach might facilitate transabdominal dissection was also raised. Careful imaging review with a radiologist facilitated surgical planning—this step was crucial to the success of the procedure. There was clear evidence of a "levator ani cap" (indicated by the arrow in Figure 3 and Figure 4) that resulted from the neurofibroma pushing up inferiorly on the levator ani muscles. This observation suggested that the mass was entirely extra-pelvic. This imaging distinction therefore indicated a pure perineal approach in prone positioning.

Figure 4. Sagittal Nonfat-Suppressed T1-Weighted MRI. Published with Permission



The large pelvic neurofibroma (asterisk) distorts and displaces the levator ani muscle (white arrow).

The patient was placed in the prone position, and his entire perineum and bilateral buttocks were prepped and draped in the usual fashion. A curvilinear longitudinal incision was performed just to the right of the sacrum and coccyx with extension directly over the gluteal mass. Dissection was carried down through the deep fascia, at which point the encapsulated mass was appreciated. It was circumferentially dissected free with a combination of sharp and blunt dissection. Great care was taken to identify and protect the pudendal nerve, the sciatic notch and sciatic nerve, the rectum and external sphincter complex, and the prostate and the crus of the penis. Digital rectal examination and flexible sigmoidoscopy was then performed to rule out anorectal injury. The prostate, urethra, and base of the penis were similarly examined to rule out injury. The operation was uncomplicated. The tumor was removed as a single intact specimen without injury to adjacent structures. Pathology confirmed neurofibroma with tumor present at the margins. The patient had an uncomplicated postoperative course and remains without evidence of recurrence at 10 months.

Conclusion

This case demonstrates the importance of preoperative planning and the necessity of radiology involvement in providing adequate surgical treatment. In this case, there was a dilemma regarding the most appropriate surgical approach. The MRI imaging clearly demonstrated a cap present on the superior aspect of the tumor, which can be readily identified as the levator ani complex. This radiologic finding completely changed the surgical planning direction by clearly indicating the need to employ a perineum approach. In addition to the surgical avenue, the imaging clearly demonstrated that the tumor was encroaching on the rectum, pudendal nerve, and the urethra. These findings indicated the necessity for colorectal and urologic consultations during the preoperative period to allow for a smooth operative course.

Lessons Learned

Complex pelvic tumors often require the assembly of an interdisciplinary surgical team and meticulous preoperative planning to facilitate safe oncologic resection; to best identify the ideal surgical route, the importance of a detailed review of high-quality imaging with a radiologist cannot be underestimated.

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