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Case Report

Intrapelvic melanocytic schwannoma resection with computer-assisted navigation ☆

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ABSTRACT

Melanocytic schwannoma is a rare nerve tumor characterized by melanin-producing neoplastic Schwann cells. Wide surgical resection is the management of choice for this tumor; however, anatomical location and proximity to nerve roots can make locating this tumor and the surgical resection challenging. Here we describe the case of 49-year-old male with a melanocytic schwannoma in the presacral area adjacent to the second sacral nerve root that was managed by wide resection aided by computer-assisted navigation due to the difficulty in identifying its location intraoperatively. The utilization of computer-assisted navigation improves accuracy and precision through the creation of a virtual continuous tridimensional map, particularly useful when oftentimes tumor margins may seem equivocal and further resection would compromise the patient's functionality. The value of computer-assisted navigation for soft tissue tumor resections in orthopedic oncology is still in its infancy, though, in certain scenarios it may advance the technique for some soft tissue resections.

Introduction

Melanocytic schwannoma (MS) is a rare nerve sheath tumor with roughly 200 reported cases [1]. Wide surgical resection is the management of choice however owing to the rarity of this tumor and that its relatively unclear clinical course there is no consensus regarding postoperative surveillance or adjuvant therapies [2]. Obtaining negative margins is especially imperative within MS's management due to the risk of local recurrence and metastatic potential. Local recurrences have been associated with incomplete resections and invasion of the surrounding tissues [3]. Although the bulk of the literature portrays the use of this technique for the purpose of osseous resections, during the past decades computer-assisted navigation has been incorporated within the musculoskeletal oncology realm as a strategy to ensure adequate oncologic resection in a precise manner by limiting resection of nonmalignant tissue [4,5].

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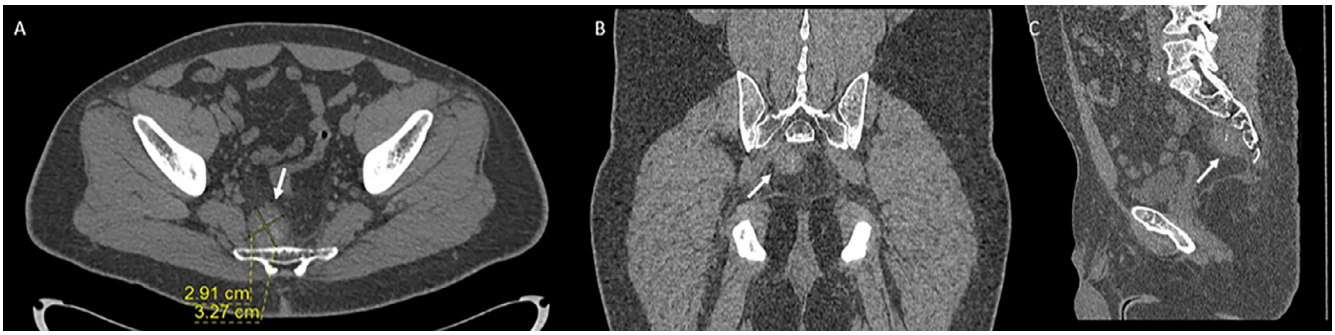

Fig. 1 – Computed tomography depicting a solitary mass (arrow) on the right anterior aspect of the sacrum. (A) Axial image, (B) Coronal image, (C) Sagittal image.

We describe a case of melanocytic schwannoma in the presacral area managed by wide resection aided by computer-assisted navigation. To the best of our knowledge this is the first such case to be described in literature.

Case report

A 49-year-old male presented to the emergency department with a complaint of abdominal pain. A computed tomography (CT) was obtained and the patient was diagnosed with a renal calculus. The symptoms resolved without intervention. However, in this CT scan a presacral mass was incidentally found and described as a rounded presacral soft tissue mass just to the right of midline, measuring 3.3 × 2.9 cm with punctate foci of mineralization, approximating the right S2 and S3 nerve roots (Fig. 1). The patient was referred to the Sarcoma Multidisciplinary Clinic of our institution, an NCI-designated comprehensive care center. The patient subsequently underwent a magnetic resonance imaging (MRI) study that confirmed a mass located lying over the right sacral ala, medial to the piriformis, at the level of S2-S4 (Fig. 2). The patient denied any complaints and examination revealed no neurological deficits.

The patient underwent a CT-guided biopsy of the mass. The histopathology demonstrated a malignant melanocytic schwannoma. Microscopically strips of neoplastic tissue with elongated cells exhibiting moderate nuclear atypia and pleomorphism with enlarged nucleoli were observed, no obvious psammomatous bodies were visualized.

Due to the tumor grade and local recurrence risk (35%), preoperative external beam radiation was recommended to aid in local disease control. Radiation was recommended in the preoperative setting as opposed to the adjuvant setting for 2 reasons. First, a lower dose is required in the preoperative setting and the treatment volume is restricted. Second, adjuvant radiation treatment is often precluded in the pelvis and retroperitoneum due to the bowel which fills the resection bed in the postoperative state. Intensity Modulated Radiation Therapy and Image-Guided Radiation Therapy technique was utilized to maximize the dose to the tumor and reduce the risk of microscopic spread of the disease while sparing critical organs nearby. The patient received 5000 cGy in 25 fractions over the course of 5 weeks.

Due to proximity of the tumor to the S2 nerve root and altered anatomy from preoperative radiotherapy, computer-assisted navigation was used to aid in the resection. Three holes were created in the right iliac wing for the computer navigational pins which were then merged with the preoperative CT scan using the Stryker Navigation System. The procedure began with gaining exposure through a lower abdomen midline incision. The viscera were mobilized out of the pelvis and the peritoneal reflection was divided lateral to the rectum, allowing exposure of the presacral space. Dissection was completed with a combination of sharp technique using Metzenbaum scissors and then with electrocautery through the mesorectal fat. As the tumor was approached, there was uncertainty about the gross tumor margins, as well as the vicinity with the S2 nerve root. Computer navigation was utilized to establish the precise location of the distal edge of the tumor, as well as the foramen for the S2 nerve root (Fig. 3). Special attention was taken to ensure that the S2 nerve root was not divided. The resection concluded without complications. Total anesthesia time was 202 minutes. The final pathology report confirmed the diagnosis of malignant melanocytic schwannoma tumor (Fig. 4). The specimen had adequate negative margins.

The patient recovered uneventfully presenting no neurological deficits. Currently, 29 months after the tumor resection, there is no evidence of local recurrence on MRI scans, as well as no evidence of metastatic disease on the CT scans of the chest. The patient will be continued to be monitored at measured intervals due to the unpredictable clinical course of melanocytic schwannoma.

Discussion

Melanocytic schwannoma is characterized by melanin-producing neoplastic Schwann cells [6]. MS primarily occurs in posterior cervical and thoracic spine roots, sympathetic chain, and gastrointestinal tract; however, it can occur anywhere in the peripheral nervous system [1,7,8]. This tumor occurs in 2 forms, depending on the presence of psammoma bodies at histological examination [9]. It presents in young individuals, without sex predominance [2]. Early literature reported MS to have a low tendency to metastasize, whereas, more recently

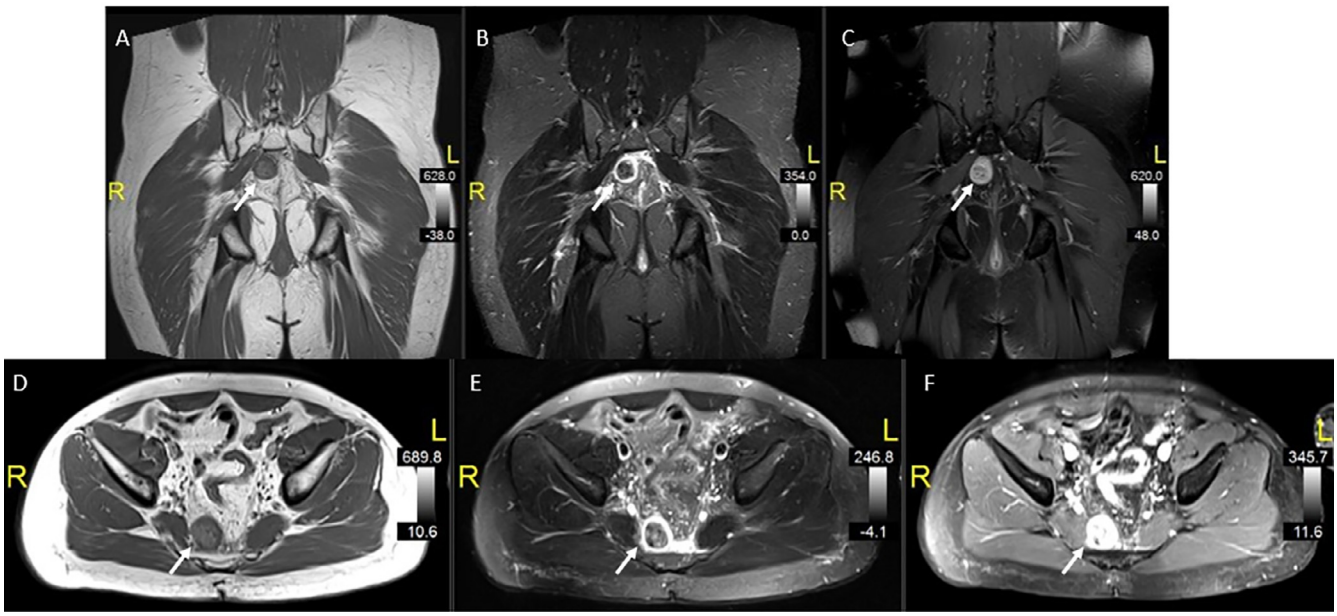

Fig. 2 – Magnetic resonance imaging showing a soft tissue mass (arrow) hypointense in T1 (A,D), with peripheral hyperintensity on STIR (B,E) and with gadolinium heterogenous enhancement (C,F).

MS's metastasizing potential has been emphasized [1,7,8]. In the largest case series with 40 patients, Torres-Mora report local recurrences in 35% and metastases in 44% of patients [8]. While there is currently no standard of care protocol for the management of MS, due to the rarity of this tumor, most patients are treated surgically [1,10]. Imperfect resections can lead to local recurrences, with some authors advocating for adjuvant radiotherapy [1,2]. In such uncertain scenarios, the surgeon's attention needs to be focused on resecting the tumor with adequate tumor-free margins.

Computed tomography (CT) and MRI are used to assess tumor characteristics and margins preoperatively while fluoroscopy, rulers and the naked-eye have historically been the only available intraoperative tools to locate and assess the adequacy of resection margins in orthopedic oncology. Integration of preoperative imaging information during the procedure can often be problematic and cumbersome especially in a difficult and irregular location, such as the pelvis, sacrum, or when anatomy has been altered from prior operations or radiation. Computer-assisted navigation, while initially developed for neurosurgical application, has been used within orthopedic surgery for a few decades now [5]. Recently, the utility of computer-assisted navigation has been explored within the musculoskeletal oncology subspecialty [4]. Computer-assisted navigation integrates preoperative and intraoperative information and provides real-time high-definition 3-dimensional feedback [11]. Once obtained, the computer-assisted navigation images have higher resolution than fluoroscopy and are also radiation free for the remaining of the procedure which is an added benefit to the surgeons and other surgical staff [12].

While there are reports of increased procedure time, this is to be expected with any new operative technique [4]. As surgeons gain familiarity with the use of computer-assisted navigation, surgical times are expected to decrease [4]. Computer-assisted navigation has been shown to improve precision and accuracy within bone tumor resections [5,13,14]. Clinically, negative margins with the use of computer-assisted navigation can decrease local recurrences; however, this has not been shown to lead to increased overall survival which is determined by metastatic disease [5].

Most of the current literature regarding computer-assisted navigation in orthopedic oncology refers to either bone tumors or bone adjacent soft tissue sarcomas that require a bony resection to achieve a margin-free excision [4,5,11–14]. In this report, the aim is to depict the usefulness of computer-assisted navigation for soft tissue tumors where gross identification of the tumor was not possible but the resection does not necessarily involve a bone cut. In this reported example, our patient presented with a tumor overlying a sacral nerve root which preoperatively was assessed as salvageable. As often seen in sacral and pelvic surgery, particularly after radiotherapy, assessing the tumor margins was challenging during the procedure. Specifically, the proximity of the distal aspect of the tumor to the nerve root made the gross intraoperative assessment of the margins tenuous, thus we resorted to this technique previously proven to improve the adequacy of bone resections. Our margins were close as expected, when dealing with a tumor adjacent to a vital structure, but negative providing the patient with all the benefits of a margin-free resection in terms of local recurrence and with the sparing of the nerve root. Some limitations were of concern prior to the procedure such as limited visualization of a soft tissue structure on CT imaging, but computer-assisted navigation was able to provide sufficient visualization of the tumor to allow a margin-free safe resection. Computer navigation is a promising technique that needs to be further explored within the musculoskeletal oncology field for soft tissue resections where the literature is currently sparse.

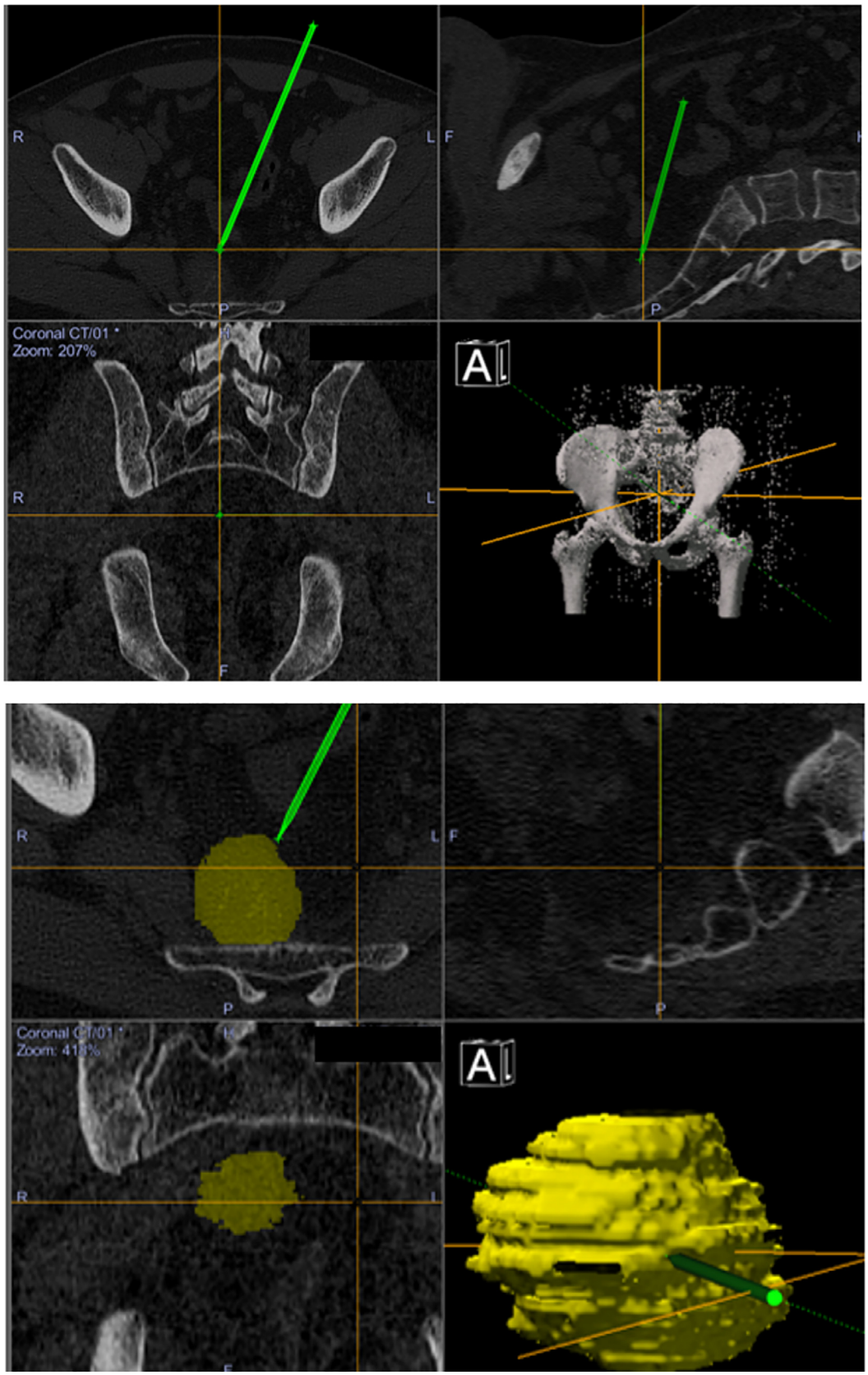

Fig. 3 – Navigation images depicting tumor location and visualization intraoperatively.

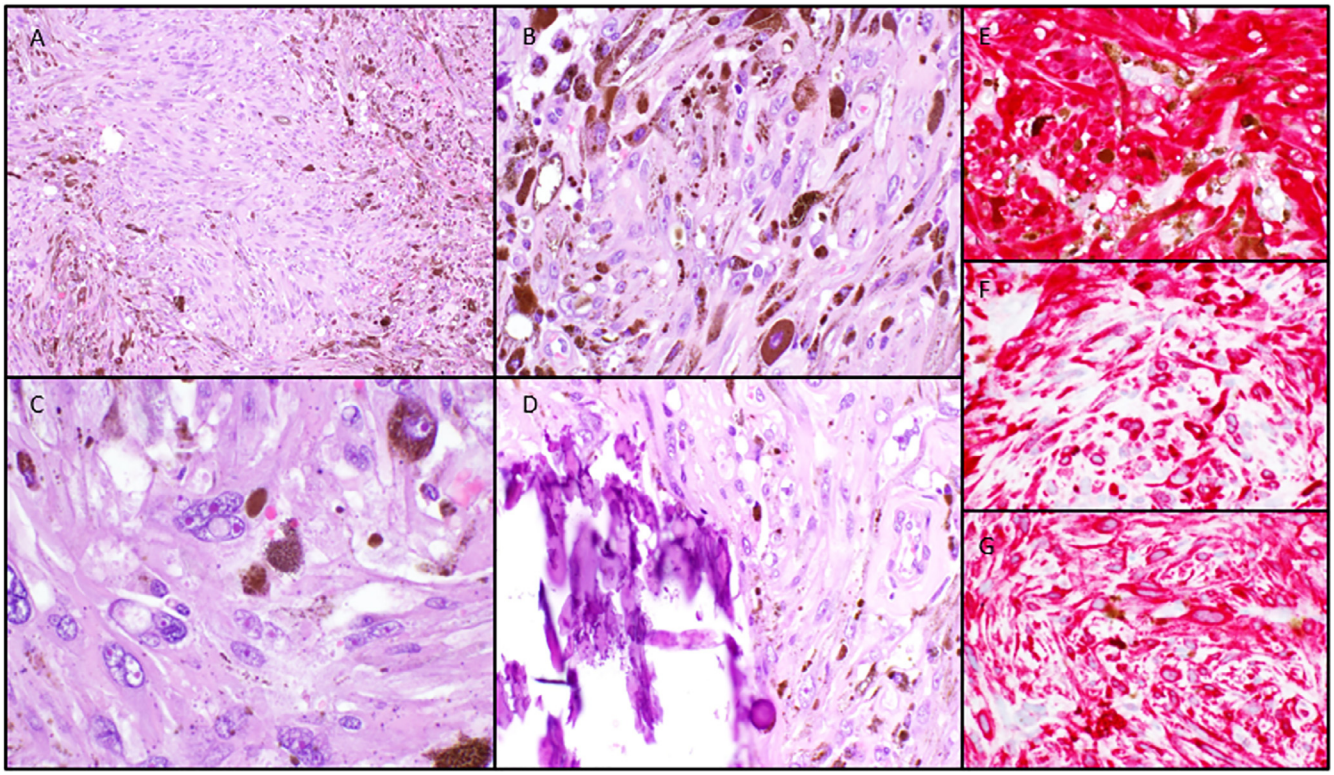

Fig. 4 – Tumor microscopic analysis. (A) Melanotic schwannoma, consisting of short fascicles of spindled to epithelioid, some of which are heavily pigmented (H&E, 10x). (B) High power view of melanotic schwannoma, showing plump spindle to epithelioid cells with indistinct amphophilic cytoplasm, round/ovoid nuclei (some with prominent nucleoli) and abundant melanin pigment (H&E, 40x). (C) Melanotic schwannoma with marked nuclear enlargement, nuclear cytoplasmic inclusion, and prominent nucleoli (H&E, 60x). (D) Melanotic schwannoma showing calcifications and small calcospherite (H&E, 40x). Immunohistochemistry demonstrating S100 (E), Melan-A (F) and HMB-45 (G) positive expression in melanotic schwannoma.

Conclusion

We believe this is the first reported case of the use of computer-assisted navigation for the resection of an intrapelvic melanocytic schwannoma. When treating rare neoplasms with an uncertain clinical course, it is paramount to achieve negative margins. The utilization of computer-assisted navigation improves accuracy and precision through the creation of a virtual continuous tridimensional map of the intraoperative field. Computer-assisted navigation improves the surgeon's confidence and decision-making process, particularly when tumor margins may seem equivocal and further resection would compromise the patient's functionality.

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