Spindle cell hemangioendothelioma of the sacrum

Case report

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Spindle cell hemangioendothelioma is a rare benign vascular tumor that is not known to involve the sacrum. The authors describe the case of a 31-year-old woman presenting with low-back and radicular pain without weakness or bowel or bladder dysfunction. Admission CT and MRI studies revealed a large S1–3 lytic sacral lesion. The patient initially underwent a nondiagnostic percutaneous biopsy. She subsequently underwent an open biopsy, during which the lesion was found to be highly vascular. Histological investigation revealed a vasoformative lesion consistent with spindle cell hemangioendothelioma. Preoperative embolization followed by resection via intralesional curettage resulted in resolution of symptoms up to 9 months postintervention. Despite the authors’ recommendation, the patient became pregnant at that time and requested no additional follow-up imaging. The authors present the first reported case of a spindle cell hemangioendothelioma of the sacrum and review the current literature.

Key Words • spindle cell hemangioendothelioma • spindle cell hemangioma • hemangioma • vascular tumor • vascular malformation • sacral tumor • oncology • vascular disorders

This article contains some figures that are displayed in color online but in black-and-white in the print edition.
Contrast T1-weighted images. On precontrast imaging, the lesion appeared hypointense on T1- and mildly hyperintense on T2-weighted images. This lesion involved most of the S-1 vertebra superiorly and the S-3 inferiorly. Laterally on both sides this lesion was seen extending into the neural foramina. On the left side, this lesion involved the S1–2 and S2–3 neural foramina. On the right side this lesion extended into the S2–3 neural foramina.

The measurements of this lesion were 3.76 × 7.6 cm in anteroposterior and transverse dimensions, respectively. The differential diagnosis based on the neuroradiologist’s reading included a metastatic deposit versus primary bone lesion. The patient underwent a percutaneous biopsy that was nondiagnostic. Open biopsy with resection of the biopsy tract revealed a low-grade spindle cell hemangioendothelioma. The patient received consultation about the surgical option of en bloc sacrectomy versus intralesional curettage. Because of her age and lack of bowel or bladder symptoms, the patient chose the latter.

Operation. Because the open biopsy revealed that the lesion was quite vascular, embolization was performed preoperatively, with treatment of the bilateral iliolumbar arteries. The patient was taken to the operating room the following day for resection of the biopsy tract and an L5–S4 laminectomy with intraoperative neuromonitoring. The majority of tumor was noted to be soft, with a somewhat firm capsule. Complete exposure of the thecal sac was achieved (Fig. 2).

As stated above, the patient desired preservation of bowel and bladder function. Because en bloc resection would have required removal of the S2–4 and possibly the S-1 nerve roots, the tumor was resected via intralesional curettage into the sacral ala bilaterally. Tumor was resected up to the cortical bone adjacent to the sacroiliac joint, because the tumor did not invade beyond this point laterally. Minimal tumor bleeding was noted secondary to the embolization. Tumor was densely adherent to the S-1 nerve roots bilaterally, with tumor resected via sharp dissection off the nerves to the foramen anteriorly. The intact thecal sac severely limited visualization anteriorly below the S-2 nerve roots. Despite the visual limitation, all visible tumor was resected. Postoperative imaging revealed resection of the lesion without evidence of obvious residual tumor (Fig. 3).

Postoperative Course. Postoperatively the patient’s pain improved, and she was subsequently discharged without issues. The patient was followed with serial MRI and was stable both on examination and imaging at 9 months postoperatively. At that time, despite our recommendation, the patient became pregnant and requested no additional imaging.

Final pathological studies revealed fascicles of spindle cells, along with round-to-ovoid cells with vacuolated cytoplasm distributed in solid aggregates, which were consistent with spindle cell hemangioendothelioma (Fig. 4).20

Discussion

Vascular tumors are uncommon entities that seldom occur in bone. Moreover, less than 10% of hemangioendotheliomas are seen in the vertebral column.2 Spindle
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cell hemangioendothelioma typically affect the distal extremities, especially the tibia and fibula. The diagnosis of hemangioendothelioma is suggestive of an intermediate, uncertain tumor grade. A hemangioendothelioma falls between a hemangioma—a benign abnormal buildup of blood vessels, and an angiosarcoma—a malignant neoplasm with a higher incidence of metastasis. Although there is controversy regarding the biological behavior of the spindle cell hemangioendotheliomas, they are typically considered to be reactive processes rather than malignant tumors. However, in comparison with other regions of the spinal cord, sacral tumors are more likely to be malignant, even in younger patients. Posterior sacral tumors are more likely to be benign than are anterior lesions.

Spindle cell hemangioendotheliomas occur within a wide age distribution in both sexes. The lesions are common in young adults. Symptoms include back pain, soft-tissue swelling, paresthesia, and neurological deficit. Ofentimes a patient with this type of lesion is asymptomatic for many years, and only begins presenting with symptoms after the tumor has grown to a considerably larger size.

The histological markers for spindle cell hemangioendothelioma are “ribbons or nests of rounded and slightly spindled endothelial cells.” The cells stain deep pink in H & E preparation. The nuclei are plump, with well-defined nucleoli. A lack of mitotic figures and necrosis confirms that the tumors have a low-grade character. Unlike the other subgroups of hemangioendothelioma, the nuclei are spindle shaped with ill-defined cytoplasmic borders, the vascular lumina are visible, and no myxohyaline matrix is present.

Due to their rarity, there is no established standard of care for the treatment of sacral spindle cell hemangioendothelioma tumors. Controversy exists concerning the treatment of more common sacral tumors, such as giant cell tumors and chordomas, due to the complexity of the anatomical location. Damage to the sacral nerve roots is associated with severe morbidity. Symptoms include incontinence, sexual dysfunction, and loss of anal sphincter control. As discussed earlier, hemangioendotheliomas, including other sacral tumors, initially tend to be asymptomatic until they have reached a relatively larger size, making resection more difficult. Treatment options include postponing treatment, serial arterial embolization, radiation therapy, resection, and surgery with adjuvant radiation therapy.

The literature has little information regarding the efficacy of radiation therapy or serial arterial embolization as a treatment option for spindle cell hemangioendotheliomas. For other sacral tumors, such as giant cell tumors, there is concern that radiation can induce sarcoma formation. Radiation was found to cause sarcomas in Grade I and II epithelioid hemangioendotheliomas of the spine. There is anecdotal evidence that serial arterial embolization provides disease control in the case of sacral giant cell tumors.

In reference to both sacral tumors and spindle cell hemangioendotheliomas, the medical literature has a greater focus on surgical treatment. Surgical options include intrallesional curettage excision or en bloc sacrectomy. En bloc sacrectomy decreases the chance of reoccurrence but is associated with great incidence of morbidity. For many tumors the en bloc approach would require complete destruction of the sacral nerve roots. When both the S-1 and one of the S-2 nerve roots are kept intact, 50% of patients experience incontinence. In cases in which both the S-1 and S-2 are preserved and only one S-3 root is sacrificed, patients retain both continence and bowel function. The nature and size of the tumor often dictates the anatomical level of resection (Fig. 2).

Both surgical options of en bloc sacrectomy and intrallesional curettage are associated with an increased risk of intraoperative hemorrhage, CSF leakage, infected wounds, sacral nerve root damage, and loss of sacral stability. Preoperative embolization is recommended to avoid intraoperative blood loss during the resection of this vascular tumor. This precaution is especially warranted for large tumors with a volume greater than 200 cm³, as well as those occurring in the S2–3 region.

Sacral tumors tend to grow around nerve sheets, making it difficult to dissect them from the nerves, rendering en bloc resection an impractical approach. Furthermore, intrallesional curettage is associated with a greater recurrence rate. Nevertheless, the severe morbidity associated with sacral nerve damage, the typical size of the tumors, and the complexity of the anatomical location

Fig. 4. A: Photomicrograph showing spindle-shaped cells in solid areas arranged into intertwining fascicles with vacuolated cytoplasm. H & E, original magnification ×200. B and C: Photomicrographs showing immunostaining of CD3(4)+ cells in spindle cell areas. Original magnification ×20.
make piecemeal resection of the tumor the most viable option. Additional research on the rate of recurrence and metastasis of spindle cell hemangioendotheliomas would allow for a more informative comparison of the risks and benefits associated with the two surgical approaches. The rate of recurrence of hemangioendotheliomas has been estimated to be 13%. Moreover, it has been noted that there is a 50% recurrence rate for patients with spindle cell hemangioendotheliomas.\(^4\)

**Conclusions**

Spindle cell hemangioendotheliomas are rare lesions that display an intermediate rate of growth. This manuscript represents the first report of this variant of hemangioendothelioma presenting in the sacrum. Treatment options include either en bloc sacrectomy or intralesion curettage. Patients treated with the latter procedure must undergo close serial follow-up to assess for tumor progression and/or recurrence.

**Disclosure**

The authors report no conflict of interest concerning the materials or methods used in this study or the findings specified in this paper.

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