Cervical Radiculopathy from Spinal Melorheostosis: A Case Report and a Review of the Literature

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INTRODUCTION

Cervical radiculopathy is caused by mechanical compression and subsequent inflammation of a cervical nerve root. Common causes of the condition include soft disc herniation, neural foraminal stenosis by an osteophyte, ossification of the posterior longitudinal ligament, or traumatic fracture, but rare conditions like neoplasia also need to be considered. Melorheostosis is a rare connective tissue disease that causes overgrowth of cortical bone. The condition is also called Léri disease after it was first described by Léri and Joanny in 1922. Although more than 400 cases of melorheostosis have been reported in the literature, isolated spinal involvement requiring surgery is extremely rare.

Melorheostosis is a rare connective tissue disease that causes overgrowth of cortical bone. The condition is also called Léri disease after it was first described by Léri and Joanny in 1922. Although more than 400 cases of melorheostosis have been reported in the literature, isolated spinal involvement requiring surgery is extremely rare. In addition, the only 2 case reports of melorheostosis involving the cervical spine manifested with myelopathy, rather than radiculopathy, due to thecal sac compression. To the best of our knowledge, no case report has been issued on isolated cervical radiculopathy caused by melorheostosis in the absence of myelopathy.

CASE REPORT

1. History

A 59-year-old woman visited our outpatient clinic with persistent neck pain and radiating pain to the left arm of 3 months duration. The patient described a continuous tingling sensation and numbness along the neck, left shoulder, outer part of the left upper arm, dorsal aspect of the forearm, and dorsal area of the middle finger consistent with C7 nerve root involvement. There was no history of previous medical disease. The patient had tried non-steroidal anti-inflammatory drugs, muscle relaxants, and physical therapy but without any improvement in symptoms. Her physician at a local clinic referred her to our institute for surgical treatment under a tentative diagnosis of bone tumor. Neurological examination revealed left C7 dermatome hypoesthesia. Her symptoms did not respond to medication or physical therapy, and thus, biopsy and surgery were performed under suspicion of melorheostosis. Persistent pain was relieved and the histologic examination was consistent with melorheostosis. Spine melorheostosis is rare but symptoms such as pain are responsive to surgery.

Key Words: Melorheostosis; Neoplasms; Radiculopathy

Cervical radiculopathy is caused by mechanical compression and subsequent inflammation of a cervical nerve root and is usually caused by soft disc herniation. Melorheostosis is a rare connective tissue disease causing overgrowth of cortical bone. We describe a rare presentation of melorheostosis and its successful surgical treatment. A 59-year-old woman with persistent neck pain and radiating pain to the left arm of duration 3 months visited our out-patient clinic. On neurologic examination, the patient showed left C7 dermatome hypoesthesia. Her symptoms did not respond to medication or physical therapy, and thus, biopsy and surgery were performed under suspicion of melorheostosis. Persistent pain was relieved and the histologic examination was consistent with melorheostosis. Spine melorheostosis is rare but symptoms such as pain are responsive to surgery.

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on preoperative imaging findings, laminectomy margins were not extended beyond the area of hyperostosis to avoid vertebral artery injury. After laminectomies, no residual compression of the left C7 nerve root was evident. C4, 5, and 6 lateral mass screws and C7 and T1 pedicle screws were inserted. Screws at C6, 7 were inserted only on the right side because left side insertion points were compromised by wide decompression (Fig. 4). Intraoperative monitoring did not show any significant change during the surgical procedure.

Macroscopically, the internal surface of the lamina was diffusely thickened with a globoid mass protruding from the inner surface. The serial section revealed that the thickened lamina was composed of pearly white or ivory-like compact cortical bone. Representative sections were decalcified and embedded in cassettes, but no definite tumor cells were observed in hematoxylin-eosin stained sections. Whereas lamina trabecular bony tissue remained normal, hyperdense mature cortical bone resulted in cortical and subchondral hyperostosis around the lamina, which was consistent with melorheostosis (Fig. 5).

Postoperatively, the patient recovered well without any neurological deficit. Motor function remained intact, and the arm pain improved immediately after surgery. Actually, she only complained of pain around the surgical site. The patient was discharged from the hospital one week after surgery. At a follow-up examination 2 years after surgery, the patient was doing well without recurrent radicular pain (Fig. 6).
**DISCUSSION**

Melorheostosis is a benign, non-hereditary disease characterized by dense, sporadic sclerosis of skeletal and adjacent soft tissue. The incidence of melorheostosis is 0.9 cases per million\(^\text{18}\)\). Spinal involvement of melorheostosis is extremely uncommon because melorheostosis typically affects appen-

Table 1. Nine cases of spinal melorheostosis requiring surgical treatment since the case report of melorheostosis involving the spinal column in 1982\(^\text{3}\).

| References          | Year | Sex | Age (year) | Location    | Symptom/Sign | Image          | Treatment                          |
|---------------------|------|-----|------------|-------------|--------------|----------------|------------------------------------|
| Garver et al.\(^\text{6}\) | 1982 | M   | 21         | Upper thoracic | Myelopathy  | X-ray              | Biopsy                             |
| Garver et al.\(^\text{6}\) | 1982 | M   | 35         | Sacrum       | -            | X-ray              | Biopsy                             |
| Robertson et al.\(^\text{13}\) | 2003 | F   | 54         | L5-S1        | Axial pain   | X-ray, CT, MRI   | Decompression/Instrumented fusion   |
| McCarthy et al.\(^\text{10}\) | 2004 | F   | 40         | T10          | Axial pain   | X-ray, CT, MRI, WBBS | Biopsy                             |
| Reznik and Fried\(^\text{12}\) | 2005 | M   | Mid 30     | CS-T4        | Myelopathy   | CT, MRI         | Decompression                      |
| Zeiller et al.\(^\text{21}\) | 2005 | M   | 35         | Whole spine  | Myelopathy   | X-ray, CT, MRI   | Decompression/Instrumented fusion   |
| Yoon et al.\(^\text{20}\) | 2011 | M   | 57         | L5-S2        | Radiculopathy| X-ray, CT, MRI, WBBS | Decompression                      |
| Saxena et al.\(^\text{16}\) | 2013 | F   | 33         | L4-S         | Radiculopathy| X-ray, CT         | Decompression                      |

M: male; F: female; CT: computed tomography; MRI: magnetic resonance imaging; WBBS: whole body bone scan.
dihydrate deposition disease, ivory osteomas, heterotopic ossification (myositis ossificans), and parosteal and periosteal osteosarcoma.\(^{11}\)

Indications for surgical decompression are the same as those for advanced spondylosis, and the aim of surgery is to relieve pressure on affected neural elements.\(^{20}\) As shown in Table 1, spinal melorheostosis may be treated by biopsy, decompression, or decompression and instrumented fusion as determined by considerations of patient symptoms and radiologic test results.

Basic follow-up requires periodic plain radiographs to enable the detection of lesion size changes and to check for spinal fusion following surgery. More detailed imaging in the form of CT or MRI may also be required and close clinical follow-up of neurological status is important.

No case of melorheostosis involving the spine has recurred. However, given the limited number of reported cases, it is not possible to determine whether total removal of the affected area should be attempted. In the described case, there was no adhesion with dura, but as melorheostosis is characterized by hyperostosis and hardening of soft tissues, this possibility should be borne in mind during surgery.

**CONCLUSION**

After reaching a diagnosis of melorheostosis, the patient was treated surgically for radiculopathy without complication. Two years later, a follow-up CT scan disclosed no recurrence of melorheostosis, and clinically the radiculopathy and hypoesthesia had fully improved. Spinal melorheostosis is a rare condition and needs to be differentiated from other bone tumors and vertebral lesions, and thus, biopsy through surgery is sometimes required. Although no case of recurrence has been reported, the possibility must be addressed by continuous follow-up. Importantly, a patient’s symptoms should be carefully considered when deciding whether to observe after biopsy or perform additional decompression.

**CONFLICTS OF INTEREST**

No potential conflict of interest relevant to this article was reported.

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