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Case Report

Intraspinal Multiple Hereditary Exostosis with Thoracic Myelopathy: A Case Report

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ABSTRACT

Osteochondromas are the most common benign bone tumor and spinal involvement is not uncommon. We report a case of osteochondroma in a 14-year-old girl with underlying multiple hereditary exostosis who presented with thoracic myelopathy symptoms and signs. Magnetic resonance imaging and computed tomography scan revealed intraspinal exostosis causing significant spinal cord compression at the thoracic vertebra. Our patient was treated surgically for exostoses and showed excellent functional and neurological outcomes. The purpose of the report is to share our experience in managing intraspinal exostosis and to emphasize the importance of spinal imaging evaluation in multiple hereditary exostosis patients.

Introduction

Multiple hereditary exostosis (MHE) is an autosomal dominant inherited benign bony growth. Although osteochondromas are commonly found at juxta epiphyseal regions of long bones, the axial skeleton and spinal column can also be involved. Recent studies reported that intraspinal osteochondromas are not uncommon in patients with MHE, of which up to 27% of the patients can have lesions encroaching into the spinal canal [1, 2]. Unidentified intraspinal osteochondroma can lead to serious neurologic injury. Therefore, magnetic resonance imaging (MRI) or computed tomography (CT) evaluation is recommended for patients with MHE at least once during their growing years [1, 3].

Case Report

A 14-year-old girl with multiple hereditary exostosis was presented with bilateral lower limb weakness and numbness for 2 months duration. She had difficulty ambulating due to lower limb weakness and a sense of instability. Her underlying MHE caused her to have exostoses over her upper and lower limbs, which led to mild deformities with no major complications.

Neurological examination showed hypertonia, hyperreflexia, sustained ankle clonus and upgoing Babinski sign of both lower limbs. Clinically her right lower limb power grade was 4/5 and her left lower limb was 2/5. Her pinprick sensation showed reduced sensation from T5 downward, with sacral sparing. All clinical signs pointed toward upper motor neuron disease, which is thoracic myelopathy. Blood investigation did not show any signs of infections or malignancy. There was no obvious abnormality in the conventional spine radiograph. MRI spine showed significant spinal cord compression at the level of T7 with cord oedema (Figure 1). CT scan (Figure 2) revealed a lesion arising from the inferior aspect of the right T7 pedicle involving the T7/8 right facet joint. Axial CT scan of T7 showed the extent of the intraspinal exostosis obliterating the spinal canal.

The patient underwent posterior instrumentation T6-T8 and laminectomy of T7, followed by excision of the right T7 pedicle and tumor with the aid of neurophysiological monitoring. On operation, an extradural mass arising from the right pedicle of T7 compressing the spinal cord was found. Her post-operative histopathology examination confirmed the lesion is benign osteochondroma. Her lower limb neurology improved at one-month post-operation and she returned to her normal function at 3 months post-operation. Her post-operative CT scan at three months (Figure 3) showed no recurrence. No symptomatic recurrence was noted during her one-year post-operative follow-up.

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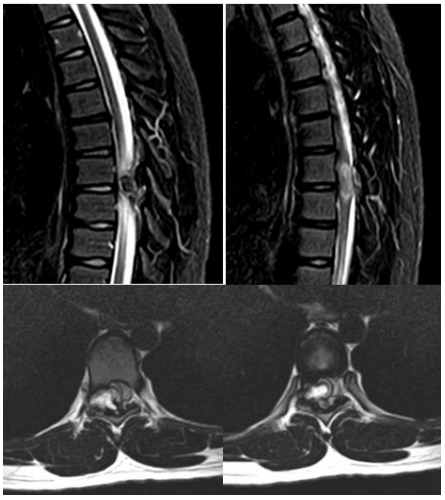

Figure 1: MRI images showing intraspinal lesion causing thoracic cord compression and oedema. The cord was shifted to the left by the right side intraspinal lesion.

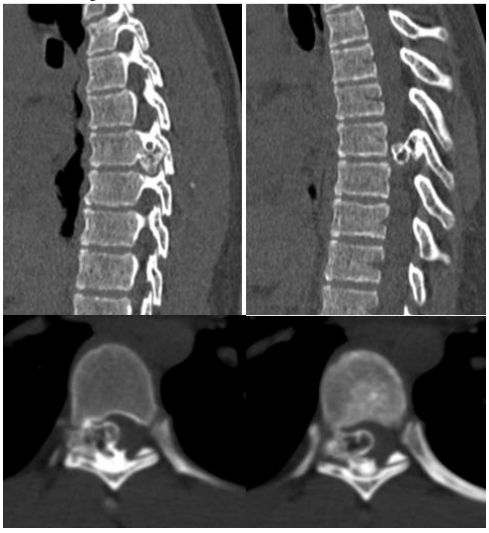

Figure 2: Computerized tomography images showing intraspinal bony exostosis arising from T7 right inferior pedicle involving the right facet joint and compromising the spinal canal.

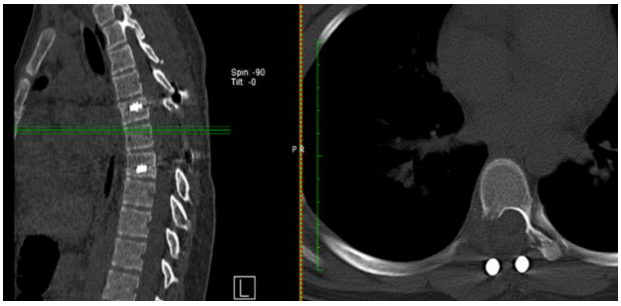

Figure 3: Post-operative 3 months CT scan showed complete excision of the lesion and no recurrence.

Discussion

Osteochondromas are the most common benign bone tumors with a wide spectrum of clinical manifestation and spine involvement is not uncommon. Spinal exostoses can occur as either solitary lesions or in association with multiple hereditary exostoses [2]. The majority of the vertebral column lesions occur in the cervical (50-80%) and thoracic (20-36%) regions and rarely in the lumbar spine [3]. Lesions commonly originate from posterior vertebral elements [2]. These histologically benign expanding intraspinal lesions can cause significant cord compression with neurologic symptoms, including radiculopathy and myelopathy. Our patient's lesion was found from the posterior element, which was from the T7 pedicle projecting into the spinal canal. She presented with thoracic myelopathy as the lesion was encroaching into the spinal canal and causing significant spinal cord compression at the T7 level. The complex anatomy of the spine makes plain radiograph film difficult to identify the spinal osteochondromas lesion. Both CT scan and MRI are good imaging modalities to visualize the spinal exostoses [2, 3]. CT scan can clearly delineate the bony nature of the lesion, whereas MRI is good for visualization of the spinal cord compression and thickness of cartilage cap.

Weinstein-Boriani-Biagini (WBB) classification incorporates the Enneking principles into the staging and surgical approach of primary tumors of the spine [4]. This classification is helpful in surgical planning of en-bloc resection according to the region of the spine and the tumor extent and location within that region [4]. Our patient's lesion was located at the right T7 pedicle. Therefore, according to WBB classification, her lesion is located at deep intraosseous zones 10 and 11. Hence en-bloc resection can be accomplished using a posterior approach. En-bloc resection is recommended as compared to intralesional to prevent recurrence [2]. En-bloc resection is the surgical removal of the tumor as a single whole piece as opposed to piecemeal resection. In our case, we removed the osteochondroma lesion as a single whole piece via osteotomized right pedicle base after the laminectomy. The surgical intervention led to functional and neurological recovery with no evidence of recurrence after 1-year follow-up.

In conclusion, MRI and CT scan are useful in detecting and localizing spinal lesions. Therefore, spinal screening using these imaging modalities is warranted in multiple hereditary exostosis patients for early diagnosis and to prevent a permanent neurological deficit. Surgery is indicated for patients who present with compressive symptoms and the surgical outcome is good with a low recurrence rate.

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