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Symptomatic intraspinal exostoses – Case series of five children

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Introduction: Until today only few studies systematically investigated spinal involvement in patients with multiple hereditary exostoses (MHE). Spinal involvement was reported to be present in 70% of MHE patients. In 25% of patients exostoses protrude into the spinal canal. 75% of these patients were previously reported to be asymptomatic and neurological intact. We report here our experience with MHE patients harboring intraspinal lesions.

Materials and Methods: We retrospectively reviewed our clinical charts of the last 6 years and identified 6 children with intraspinal lesions with a median age of 14.4 years (range: 8 - 17 years). In all cases imaging studies with MRI were performed preoperatively.

Results: In our center 5 out of 6 patients with intraspinal lesions displayed neurological symptoms and were therefore operated. In 4 cases the intraspinal exostoses were located in the cervical spine (C3, C4, 2x C5), in one case in the thoracic spine (T10). Two lesions originated from the lamina, one from the facet joint, one from the spinous process and one ventrally from the vertebral body. All of them were either encroaching nerve roots (2 cases) or directly compressing the myelon (3 cases). Consequently two children presented with radiculopathy (C5, C6) and three with signs of myelopathy. We carried out 6 operations in these 5 patients, in one case the removal of the lesion was insufficient and had to be completed in a second intervention. In all cases a microsurgical approach was utilized to minimize surgical trauma. In all patients preoperative neurological symptoms resolved shortly after surgery.

Discussion: This is the largest series of neurological symptomatic surgical cases of intraspinal exostoses in children. In our experience neurological impairment is present in more than 80% of MHE patients with intraspinal lesions in this age group, which is higher than previously reported. Due to the possibly irreversible nature of neurological sequelae timely detection of intraspinal exostoses is desirable to enable early surgical resection. Therefore, all patients with MHE should receive MRI imaging at least once during their growth period.

Keywords: intraspinal exostoses; multiple hereditary exostoses.