Child's Nervous System (2019) 35:1447–1448
https://doi.org/10.1007/s00381-019-04211-2

LETTER TO THE EDITOR

Spontaneous resolution of syringomyelia secondary to cranio-cervical junction stenosis in a patient with achondroplasia

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Received: 7 May 2019 / Accepted: 15 May 2019 / Published online: 24 May 2019

Dear Editor:

It is well known that children with achondroplasia are exposed to the risk of spinal cord compression, especially in the early months of life, because of abnormal placement and premature fusion of the posterior synchondrosis and a defect in the endochondral ossification in the basiocciput which may lead to anterior extension of the squamous occipital [1–3]. These developmental abnormalities result in hypertrophied posterior occipital rim, short clivus, shallow posterior fossa, undersized transverse diameter, and abnormal craniocervical relationship. Growth of the basiocciput will lead to increase in foramen magnum dimensions with age, after 18–24 months; thus, this could relieve the osseous compression on neurological structures [1, 2].

Management of foramen magnum compression with prophylactic surgery in those asymptomatic subjects is controversial, being the surgical treatment reserved for symptomatic infants [4, 5].

We observed a 3-year-old boy with sporadic-type achondroplasia and age-matching milestones. Parents denied any swallowing difficulty, difficulty walking, snoring, or urinary incontinence. Clinically, his head circumference was at the 50th percentile (head circumference growth chart for male achondroplasia). His neurological examination was clinically unremarkable. However, a MRI cranio-cervical study besides the supratentorial chronic hydrocephalus and the expected craniovertebral stenosis with cervicomedullary compression demonstrated edema and syringohydromyelia of the upper spinal cord (Fig. 1). Due to the presence of the syringomyelia, we suggested decompressive surgery that parents refused.

Nine months later, on MRI, there was still reduced effacement of the subarachnoid spaces at the craniovertebral junction, but at the T2 sequences, the hyperintensity of the spinal cord was markedly reduced and the syringomyelia almost resolved. The anterior-posterior (AP) diameter was 20.4 mm and the transverse diameter (TS) 16.5 mm. A further control MRI after 16 months demonstrated the complete disappearance of the syringomyelia. The AP diameter was 20.4 mm and the TS diameter 17.2 mm. There is a clear rim of CSF around the cord (Fig. 2).

In our opinion, this case is relevant because it demonstrates that in achondroplasic children, the characteristic pattern of growth of the foramen magnum which leads to a minor compression of the spinal cord at the cervical junction may account for the spontaneous resolution of the secondary cerebrospinal fluid dynamic anomalies. This observation may further support the indication for a conservative management of asymptomatic children with achondroplasia.

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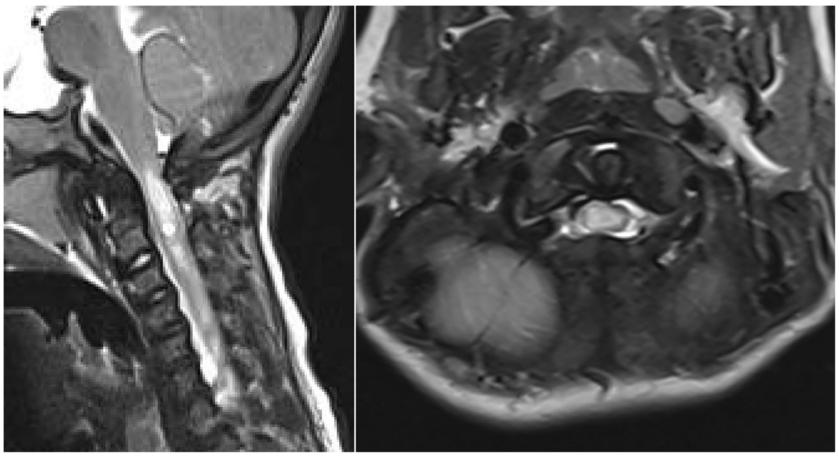

Fig. 1 Sagittal and axial MRI images of the craniovertebral junction showing compression of the cervicomedullary junction and diffuse edema of the spinal cord, effacement of the subarachnoid spaces, and syringomyelia

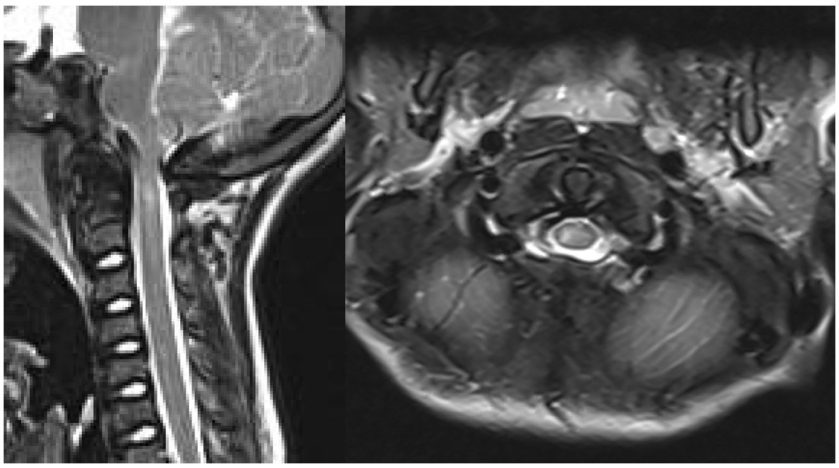

Fig. 2 Sagittal and axial MRI images at the 16-month interval showing the complete disappearance of the syringomyelia with marked reduction of the spinal cord edema

Compliance with ethical standards

Conflict of interest On behalf of all authors, the corresponding author states that there is no conflict of interest.

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