J Korean Child Neurol Soc 2015;23(3):135-137
pISSN 1226-6884•eISSN 2383-8973

이차적 사경을 증상으로 한 척추 종양

연세대학교 의과대학 세브란스병원 소아과학교실[1], 신경외과학교실[2]
손여주[1] · 이현주[1] · 이하늘[1] · 강훈철[1] · 김흥동[1] · 이영목[1] · 김동석[2] · 이준수[1]

Spinal Cord Tumor Presenting as Secondary Torticollis: A Case Report

We report a case of 6-year-old boy who presented with a 1-month history of neck pain, right-sided torticollis, with no neurological deficit on physical examination. Computed tomography (CT) and magnetic resonance imaging (MRI) of the brain and spine revealed an expansive intramedullary tumor in the cervical spine. Histopathologic examination of the biopsy specimen revealed primary low-grade glioma. This highlights the importance of physician's vigilance when diagnosing patients with common chief complaints such as torticollis and neck pain in order to avoid missing possible severe causes and to ensure patient management and prognosis.

Key Words: Cervical dystonia, Torticollis, Spinal cord neoplasms

Yeo Ju Son, MD[1], Hyun Joo Lee, MD[1], Ha Neul Lee, MD[1], Hoon Chul Kang, MD, PhD[1], Heumg Dong Kim, MD, PhD[1], Young Mock Lee, MD, PhD[1], Dong Suk Kim, MD, PhD[2], Joon Soo Lee, MD, PhD[1]

[1]Division of Pediatric Neurology, Department of Pediatrics, Severance Children's Hospital, Yonsei University College of Medicine, Seoul, Korea, [2]Division of Pediatric Neurosurgery, Department of Neurosurgery, Severance Children's Hospital, Yonsei University College of Medicine, Seoul, Korea

Introduction

Cervical dystonia is a clinical syndrome characterized by involuntary sustained muscle contractions in the neck that usually produce twisting and repetitive movements or abnormal postures[1)]. Previously known as spasmodic torticollis, its severity can vary, but the disorder can cause significant pain and discomfort. Secondary cervical dystonia is usually due to craniocerebral trauma, stroke, encephalitis, and brain tumor[2)]. Torticollis is rarely seen in childhood, with an estimated incidence of 1.3%, but more than 80 different causes of torticollis have been described[3)]. The differential diagnosis of torticollis in children is wide and extensive. Torticollis can appear as the first sign of central nerve system pathology with relatively high occurrence, but often ignored[4)]. The natural history of cervical dystonia is unknown, and secondary causes should be investigated in cases that manifest in infancy or early childhood. We report a case of 6-year-old boy presented with cervical dystonia who was eventually diagnosed with a spinal cord tumor.

Case report

A 6-year-old boy presented to our pediatric neurology clinic with sudden onset cervical dystonia. His ante- and perinatal histories were unremarkable, and his development was age-appropriate. He started to have symptoms about

Submitted: 28 July, 2015
Revised: 20 September, 2015
Accepted: 28 September, 2015

Correspondence to Joon Soo Lee, MD, PhD
Division of Pediatric Neurology, Department of Pediatrics, Severance Hospital, Yonsei University College of Medicine, 50-1, Yonsei-ro, Seodaemun-gu, Seoul, 03722, Korea
Tel: +82-2-2228-2050, Fax: +82-2-393-3080
E-mail: JOONSL96@yuhs.ac

1 month prior to presentation with neck tilting to the right and had intermittent pain precipitated by movement. A neurological examination was unrevealing. Plain radiographs of the cervical spine and neck sonography were also unremarkable, but because he continued to have symptoms with no improvement, we performed brain magnetic resonance imaging (MRI), which showed swelling of the cervical cord with a syrinx, and subsequent spinal MRI revealed an intramedullary tumor at the C4-T6 level with cystic and enhancing solid components (Fig. 1). He underwent partial tumor removal due to the lesion location and received postoperative radiotherapy. Tumor was pathologically confirmed with low grade glioma. His symptoms such as cervical dystonia resolved immediately after surgery. Serial MRI follow-up confirmed tumor stabilization (Fig. 2). He did not experience significant side effects from radiotherapy or surgery and is currently attending school with no further issues.

Discussion

Neck pain is a very common chief complaint that physicians face in clinics and emergency departments[5]. Torticollis is also quite common, but these two conditions are not necessarily related. Acquired torticollis combined with neck pain is sometimes idiopathic and benign; however, this condition can be caused by trauma, infection, or even malignancy[3,6,7]. Thorough history taking and physical and neurological examinations are extremely important; however, these tests cannot screen for all severe causes, making imaging studies necessary[8]. In the present case, even though his history, physical and neurologic exams, and plain radiographs were unrevealing, brain MRI showed a spinal cord tumor extending from the lower cervical cord, and spinal MRI revealed its full extent. The final diagnosis of a spinal cord tumor was made at the third visit, which led to immediate surgery and radiotherapy with favorable prognosis and good quality of life[9,10]. Seven percent of pediatric patients with central nervous system tumor show a symptom of head tilt[11]. Low-grade gliomas are

Fig. 2. Postoperative spinal MRI. At 18 months after surgery, we confirmed tumor size reduction and stable status by follow up spinal MRI.

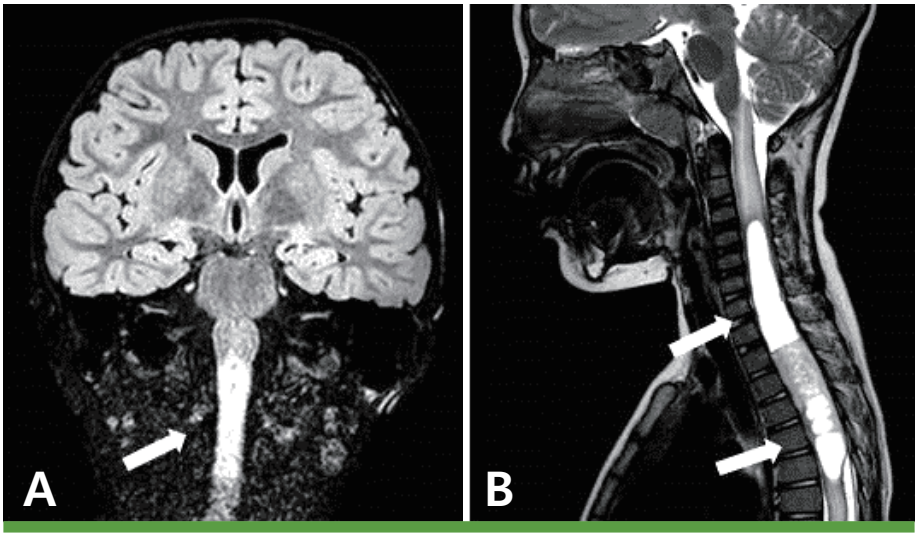

Fig. 1. Brain (A) and spine MRI (B) showing an intramedullary tumor at the C4-T6 level with cystic and enhancing solid components (white arrows indicate intramedullary tumor lesions).

the most common brain tumor of childhood accounting for 35% of all pediatric central nervous system tumors. The symptoms caused by low-grade gliomas are dependent on the location of the tumor[12]. Bandopadhayay et al. reported in a study with four thousand and forty patients with either WHO grade I or II pediatric low-grade gliomas, 5% of the total had spinal cord as the primary site. Children with pediatric low-grade gliomas are known to have excellent 10-year survival rates[13]. This highlights the importance of physician's vigilance in diagnosing patients with common chief complaints, such as torticollis and neck pain, to identify those with severe causes and ensure patient management and prognosis.

요약

경부 긴장 이상은 강직성 사경이라고도 하며 불수의적으로 목의 근육이 수축된 상태를 말한다. 경부 긴장 이상은 원발성 원인과 이차적인 원인에 의해서 발생할 수 있는데, 이차적인 원인에는 외상, 뇌졸중, 뇌염, 종양 등이 있을 수 있어 이러한 증상이 있는 소아를 진찰 시에 면밀히 살펴 보아야 한다. 이 증례는1달전부터 시작된 오른쪽 사경을 가진 6세 남아에 대한 보고이다. 환아는 사경의 반대 쪽으로 경추 회전시 통증을 호소하였으며, 신체진찰 및 신경학적 진찰상 이상 소견이 관찰되지 않았다. 방사선 사진 시행하였으나 이상 소견 없었으며 증상 지속되어 뇌 및 척추 자기공명영상 시행하였으며 검사상 경추 부위에서 수뇌종양이 관찰되었다. 수술시 시행한 생검의 조직학적 검사상 원발성 저급 신경교종으로 진단되었다. 환아는 응급 수술을 통해 증상이 호전되었으며 이후 방사선 치료를 추가적으로 받았으나 후유증 없이 생활하고 있다. 이 증례를 통해 본 보고에서는 환아와 같이 사경이나 목 부위 통증과 같은 흔하게 볼 수 있는 증상을 가진 환자를 진료할 때, 검사자가 주의 깊게 접근하여 중증의 원인을 놓치지 않고 환자에게 좋은 예후를 줄 수 있도록 하는 것이 중요하다는 것을 강조하였다.

References

1) Jankovic J, Leder S, Warner D, Schwartz K. Cervical dystonia: clinical findings and associated movement disorders. Neurology 1991;41:1088-91.
2) Camargo CH, Teive HA, Becker N, Baran MH, Scola RH, Werneck LC. Cervical dystonia: clinical and therapeutic features in 85 patients. Arq Neuropsiquiatr 2008;66:15-21.
3) Per H, Canpolat M, Tumturk A, Gumus H, Gokoglu A, Yikilmaz A, et al. Different etiologies of acquired torticollis in childhood. Childs Nerv Syst 2014;30:431-40.
4) Fafara-Les A, Kwiatkowski S, Marynczak L, Kawecki Z, Adamek D, Herman-Sucharska I, et al. Torticollis as a first sign of posterior fossa and cervical spinal cord tumors in children. Childs Nerv Syst 2014;30:425-30.
5) Natarajan A, Yassa JG, Burke DP, Fernandes JA. Not all cases of neck pain with/without torticollis are benign: unusual presentations in a paediatric accident and emergency department. Emerg Med J 2005;22:646-9.
6) Aydin R, Bilgici MC, Dagcinar A. A very rare cause of neck pain: primary Ewing sarcoma of the axis. Pediatr Emerg Care 2013;29:1197-200.
7) Cafarotti A, Bascietto C, Salvatore R, Breda L, Chiarelli F, Piernicola P. A 6-month-old boy with uncontrollable dystonic posture of the neck. Sandifer syndrome. Pediatr Ann 2014;43:17-9.
8) Haque S, Bilal Shafi BB, Kaleem M. Imaging of torticollis in children. Radiographics 2012;32:557-71.
9) Gerard CS, Straus D, Byrne RW. Surgical management of low-grade gliomas. Semin Oncol 2014;41:458-67.
10) Shields LB, Choucair AK. Management of low-grade gliomas: a review of patient-perceived quality of life and neurocognitive outcome. World Neurosurg 2014;82:e299-309.
11) Wilne S, Collier J, Kennedy C, Koller K, Grundy R, Walker D. Presentation of childhood CNS tumours: a systematic review and meta-analysis. Lancet Oncol 2007;8:685.
12) Bergthold G, Bandopadhayay P, Bi WL, Ramkissoon L, Stiles C, Segal RA et al. Pediatric low-grade gliomas: how modern biology reshapes the clinical field. Biochim Biophys Acta 2014;1845:294-307
13) Bandopadhayay P, Bergthold G, London WB, Goumnerova LC, Morales La Madrid A, Marcus KJ et al. Long-term outcome of 4,040 children diagnosed with pediatric low-grade gliomas: an analysis of the Surveillance Epidemiology and End Results (SEER) database. Pediatr Blood Cancer 2014;61:1173-9