

Case report

POSTERIOR FOSSA TUMORS: A CASE REPORT OF ACQUIRED TORTICOLLIS IN CHILDREN

Salvatore Accomando ¹, Claudia Sciarrotta ¹, Claudio Montante ¹, Veronica Notarbartolo ¹, Francesca D'Aiuto ², Giovanni Corsello ¹, Mario Giuffrè ¹

1. Department of Health Promotion, Mother and Child Care, Internal Medicine and Medical Specialities, University of Palermo, Italy.

2. ARNAS Civico Hospital, G. Di Cristina & Benfratelli, Palermo. Italy.

ARTICLE INFO

Article history:

Received 28 Sep 2022

Accepted 13 Dec 2022

Published 29 Dec 2022

Keywords:

headache, cancer, early diagnosis, torticollis.

ABSTRACT

Torticollis is a pathological condition characterized by involuntary flexion of the neck to the affected side and contextually a rotation of the chin in the opposite direction. We present the case of a 7-year-old girl, admitted to the paediatric emergency department complaining of headache, laterocollis rotation, left torticollis and deviation of the head to the right, without history of trauma or systemic illness. Computed Tomography (CT) and Magnetic Resonance Imaging (MRI) brain scans were performed, showing a subtentorial lesion, expanded into the left median and paravermian region, associated with hypertensive hydrocephalus. Histopathological analysis revealed a first-degree astrocytoma. The aim of this case report is to highlight the importance of tumors as the differential diagnosis of acquired torticollis in children.

© EuroMediterranean Biomedical Journal 2022

1. Introduction

Torticollis is a pathological condition characterized by involuntary flexion of the neck to the affected side and contextually a rotation of the chin in the opposite direction. Torticollis can be classified into two types: *congenital* which is more frequent and *acquired*. Congenital torticollis is usually due to an injury of sternocleidomastoid muscle (birth trauma or intrauterine malposition) [1]. This mass regresses and leaves a fibrous lump in the muscle, causing contracture of the neck. Acquired torticollis could have the same presentation but be secondary to a variety of causes such as ligamentous, muscular, osseous, ocular, psychiatric, and neurologic disorders (abnormalities of the spinal cord and brain, like spinal syrinx or - as in our case - central nervous system, CNS, neoplasia) [1]. As torticollis can be secondary to a wide spectrum of underlying problems, it is advisable not to underestimate this symptom. Literature points out that torticollis can be the first presenting symptom of CNS tumors in almost 20% of children, aged 2-8 years old [2,3].

2. Case presentation

A 7-year-old girl, with a previous history of correct divergent strabismus, presented at the emergency department complaining of nocturnal headache, nausea, vomiting and hyporexia associated with laterocollis rotation, left torticollis and deviation of the head to the right. These symptoms had occurred about a month previously, in the week prior to admission, they had worsened despite treatment with analgesics. An eye examination was performed by a private ophthalmologist who found "drusen". Our physical examination of the patient revealed pain with passive and active head movements, weakness and functional limitation. Blood tests showed a normal blood cell count and normal indices of inflammation. A CT and MRI brain scan were performed: a subtentorial lesion was detected with expansion into the left median and paravermian region (46x52x40 mm), with marked and patchy contrast enhancement (cystic and solid components) [Figures 1-2]. Also, hypertensive hydrocephalus and a modest distension of the perioptic sheath were described. The patient underwent external ventricular shunt followed by craniotomy, with a first subtotal excision of the tumor. After 20 days, a

* Corresponding author: Claudia Sciarrotta claudiasciarrotta@gmail.com

DOI: 10.3269/1970-5492.2022.17.44

All rights reserved. ISSN: 2279-7165 - Available on-line at www.embj.org

second neurosurgical intervention was performed and no residual tumor was found on radiological control. Histopathological analysis revealed a first-degree astrocytoma. One month after surgery, clinical examination revealed no symptoms or signs of neurological dysfunction, except for a transient *claudicatio*. Neither chemotherapy nor radiotherapy have been performed.

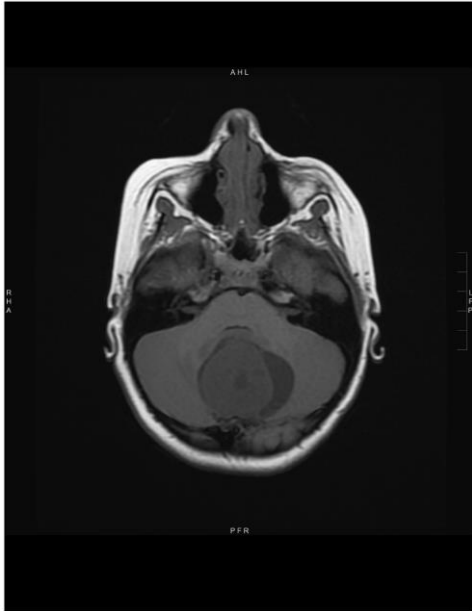


Figure 1. Posterior fossa lesion with expansion in the left median paravermian region (T1 sequency).

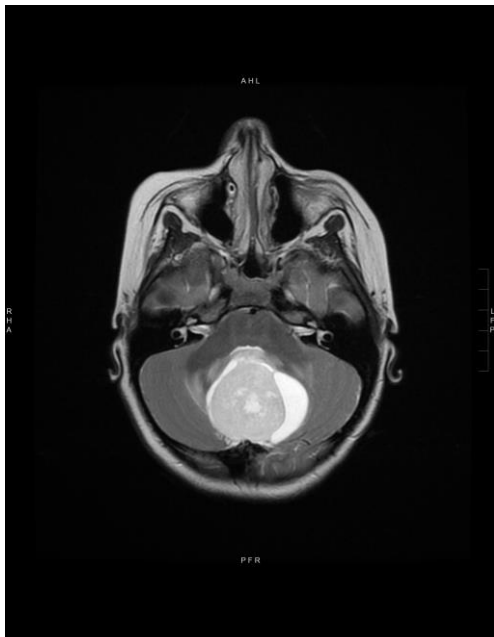
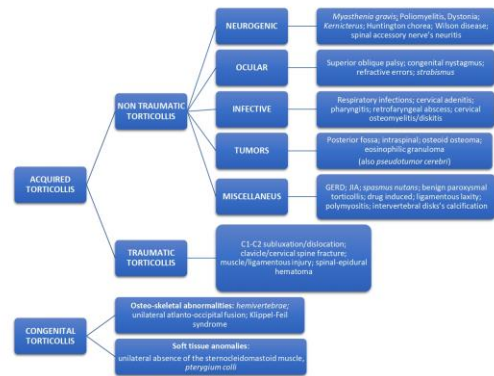


Figure 2. Marked and patchy contrast enhancement due to liquid and solid components (T2 sequency).

3. Discussion

Acquired torticollis is a challenge in the Emergency Department setting. Several diseases arise with torticollis and some of them are life-threatening. The exact mechanism of CNS tumor-associated torticollis is unclear and it might be caused by stretching and irritation of the dura, innervated by ascending meningeal branches from the upper three cervical nerves and the accessory (XI) nerve pressure, causing pain and involuntary position of the head [4]. A wide range of underlying neurological conditions can cause this symptom, such as benign paroxysmal torticollis, CNS abnormalities (syringomyelia, cervical cord, and posterior fossa tumors, cervicomedullary malformations), upper respiratory infections, diplopia, dystonia, juvenile rheumatoid arthritis, Sandifer syndrome and *spasmus nutans* [Table 1] [5].



Acquired Torticollis in Children, 2021. UpToDate (Modified)

Table 1. Differential diagnosis of torticollis in children.

Physical examination is essential in differential diagnosis, the coexistence of torticollis and other symptoms may suggest causes other than neoplasms. Conversely, if a tumor is suspected, it is mandatory to perform further tests such as CT and MRI. In the event of CNS tumors, underestimating acquired torticollis could lead to delayed diagnosis and poor prognosis. In these cases, symptoms frequently associated with torticollis are vomiting, headache, ataxia, behavioral disturbances, lacrimation and photophobia [Table 2]. In our case, torticollis was present at the clinical onset of the disease, associated with persistent headache and nausea. Pain was atypical and resistant to analgesics. Seizures are not a typical symptom of posterior cranial fossa tumors (PFTs). CNS Tumors connected to this presentation are mainly located at the cerebellum (57%), the fourth ventricle (17%), and the brainstem (13%). The predominant histological type is astrocytoma (51%), followed by medulloblastoma (24%), ependymoma (13%) and glioma (4%) [6]. Prompt investigations allowed for the diagnosis of cerebellar neoplasm, with radiological characteristics suggestive of a pilocytic astrocytoma. As described in literature, complete surgical excision of the mass resulted in complete regression of the torticollis and other symptoms.

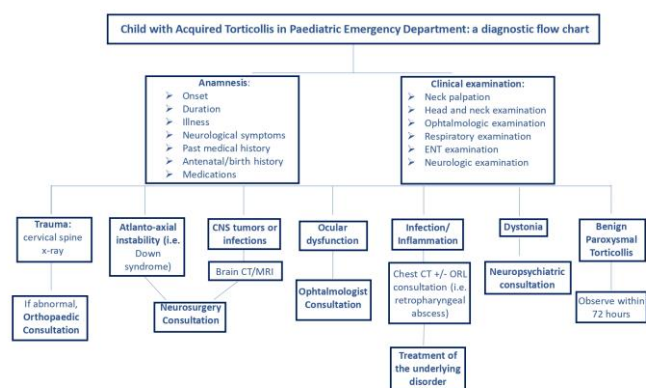


Table 2. Diagnostic flowchart of acquired torticollis in children.

4. Conclusions

PFTs represent 50% of paediatric brain tumors and torticollis is a crucial symptom in early diagnosis and should not be ignored, thus intracranial malignancy should be ruled out in any patient with persistent torticollis with unidentified explanation.

References

1. Kumandaş S, Per H, Gümüş H, Tucer B, Yikilmaz A, Konaş O, Coşkun A, Kurtsoy A. Torticollis secondary to posterior fossa and cervical spinal cord tumors: report of five cases and literature review. *Neurosurg Rev.* 2006, 29(4):333-8. doi: 10.1007/s10143-006-0034-8
2. Tümtürk A, Kaya Ozcora G, Kacar Bayram A, Kabaklioglu M, Doganay S, Canpolat M, Gumus H, Kumandas S, Unal E, Kurtsoy A, Per H. Torticollis in children: an alert symptom not to be turned away. *Childs Nerv Syst.* 2015, 31(9):1461-70. doi: 10.1007/s00381-015-2764-9
3. Fařara-Leš A, Kwiatkowski S, Maryńczak L, Kawecki Z, Adamek D, Herman-Sucharska I, Kobylarz K. Torticollis as a first sign of posterior fossa and cervical spinal cord tumors in children. *Childs Nerv Syst.* 2014, 30(3):425-30. Doi: 10.1007/s00381-013-2255-9
4. Mutsaers P, Fick M, Plötz FB. Acquired torticollis as the only initially presenting symptom in a child with a brainstem glioma. *Eur J Pediatr.* 2007 Oct;166(10):1075-6. doi: 10.1007/s00431-006-0348-0.
5. Per H, Canpolat M, Tümtürk A, Gumuş H, Gokoglu A, Yikilmaz A, Özmen S, Kaçar Bayram A, Poyrazođlu HG, Kumandas S, Kurtsoy A. Different etiologies of acquired torticollis in childhood. *Childs Nerv Syst.* 2014 Mar;30(3):431-40. doi: 10.1007/s00381-013-2302-6.
6. Extremera VC, Alvarez-Coca J, Rodríguez GA, Pérez JM, de Villanueva JL, Díaz CP. Torticollis is a usual symptom in posterior fossa tumors. *Eur J Pediatr.* 2008 Feb;167(2):249-50. doi: 10.1007/s00431-007-0453-8.