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CASE REPORT

Atlanto-axial rotatory fixation complicating ventriculo-peritoneal shunt surgery: a case report and literature review

Costanza Zattra[1] · Marco Paolo Schiariti[1] · Emanuele La Corte[1,2] · Morgan Broggi[1] · Francesco Acerbi[1] · Paolo Ferroli[1]

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Abstract

Introduction Atlanto-axial rotatory fixation (AARF) is a rare complication of ventriculo-peritoneal shunt (VPS) surgery.
Case presentation The authors present a unique case of AARF developing early after VP shunting, with persistent torticollis, a "cock-robin" head position, and a thick fibrous band along the catheter path. Due to refractoriness to conservative treatments, AARF, which can be an early-onset complication of VPS surgery, was resolved by removing the distal catheter along with the fibrous band encasing it.
Conclusion Surgical removal of the fibrous band might be enough to solve such complication with no need of further surgical fusion procedures.

Keywords Pediatrics · Atlanto-axial rotatory fixation · Hydrocephalus · Ventriculo-peritoneal shunt

Introduction

Atlanto-axial rotatory fixation (AARF) is a well-described disorder characterized by a fixed sub-luxation of the C1–C2 complex [5–7]. Children with AARF present with painful torticollis, limited range of movements at the cranio-vertebral junction (CVJ), and the so-called cock-robin position—where the head is rotated to one side and laterally flexed to the contralateral one [4, 5]. We present the rare case of a 6-year-old boy with torticollis and computed tomography (CT)-based diagnosis of AARF occurring early after ventriculo-peritoneal shunt (VPS) placement for the resolution of obstructive hydrocephalus caused by a pilocytic astrocytoma of the chiasmatic-hypothalamic region, along with a brief review of the literature.

Costanza Zattra and Marco Paolo Schiariti contributed equally to this work.

✉ Costanza Zattra
costanzamaria.zattra01@universitadipavia.it

[1] Department of Neurosurgery, Fondazione IRCCS Istituto Neurologico Carlo Besta, via Giovanni Celoria 11, 20133 Milan, Italy

[2] Department of Health Sciences, University of Milan, Milan, Italy

Case report

A 6-year-old boy was referred to our institution for a 2-month history of headache and vomiting, poor appetite, weight loss, global weakness, and decreased left visual acuity. CT and magnetic resonance (MR) scans showed a large tumor of the chiasmatic-hypothalamic region ($45 \times 32 \times 39$ mm, AP × LL × CC), occupying the third ventricle and extending into the left carotid cistern, causing obstructive hydrocephalus (Fig. 1a), which prompted for VPS positioning, with resolution of the presenting symptoms. After few months, sub-total tumor resection (STR) via a sub-frontal inter-hemispheric approach, in a neutral supine position (histological diagnosis: pilocytic astrocytoma), was performed (Fig. 1b). The postoperative course was uneventful and intracranial hypertension symptoms resolved; the visual deficit remained stable. However, in the time between VPS and STR, the patient developed a cervical dystonic muscular contraction with the head flexed to the right and rotated toward the left side ("cock-robin position"), accompanied by neck pain, which was only partially relieved upon administration of NSAIDs (non-steroidal anti-inflammatory drugs) (Fig. 2a). His past medical history (PMH) had already been characterized by two episodes of post-traumatic torticollis, both treated and

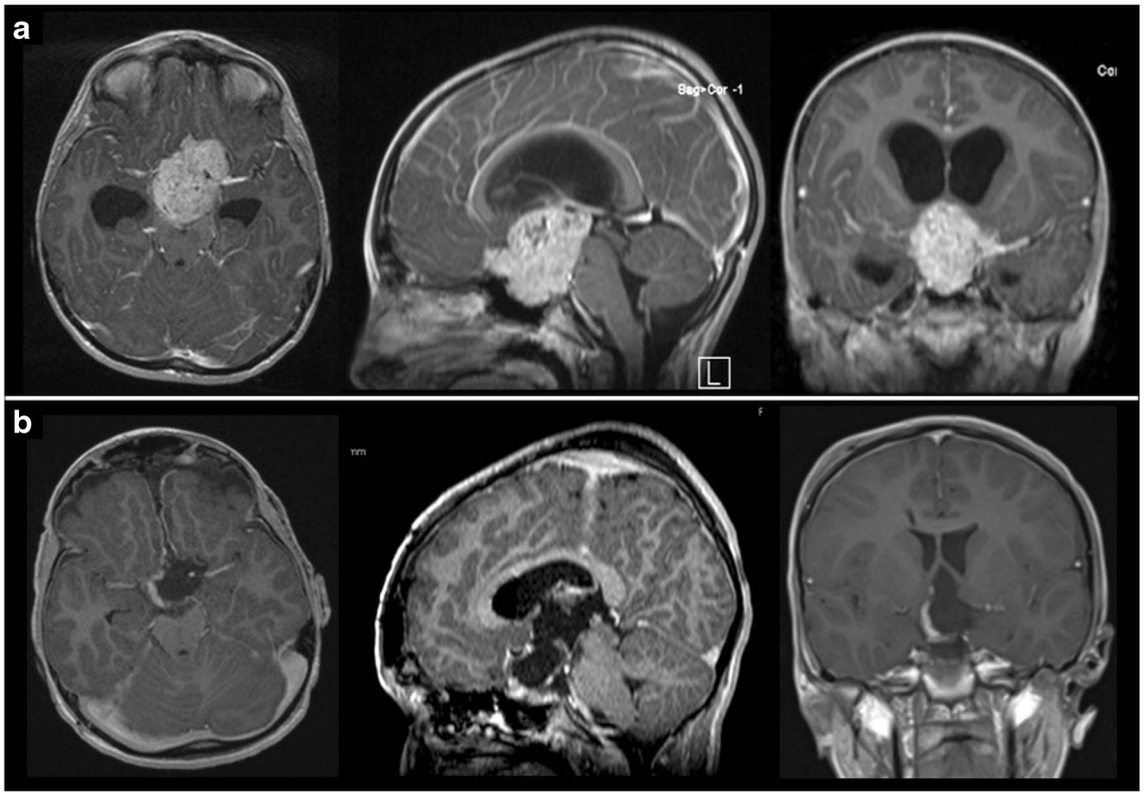

Fig. 1 **a** Pre-operative axial, sagittal, and coronal T1-weighted post-contrast MR images showing a large third ventricular lesion. **b** Post-operative axial, sagittal, and coronal T1-weighted post-contrast MR images showing a sub-total removal with a residual tumor firmly attached to the right medial surface of the diencephalon

solved with a conservative approach (rest and NSAIDs). The torticollis persisted also after STR, warranting the execution of CVJ volumetric CT and MR scans, which showed the presence of a rotated C1 over C2 sub-luxation, a condition known as atlanto-axial rotatory fixation (AARF) (Fig. 2b–e). There was an attempt to reduce the sub-luxation with the use of manual traction under sedation and neurophysiological monitoring. During the traction maneuvers, the cervical part of the VPS was particularly tense and a thick fibrous-like band was visible on the right latero-cervical aspect of the patient's neck, consistent with the subcutaneous pathway of the shunt (Fig. 3a). After the procedure, the patient was discharged with myo-relaxant medications and cervical neck brace positioning for 2 months, but after a few weeks, his parents noted a relapse of the torticollis, despite the cervical neck brace. Different treatment approaches were discussed, including CVJ surgical stabilization, but based on the previous findings concerning the cervical portion of the VPS catheter, we decided to revise the whole VPS system. Intra-operatively, a thick fibrous, scar-like band around the cervical portion of the catheter was noted and surgically lysed and the encased tube removed and replaced by a new catheter positioned on the left side. The post-operative course was uneventful with progressive improvement of the torticollis until complete resolution at 12 months follow-up (Fig. 3b, c).

Discussion

AARF is a rare pathological condition consisting of a C1–C2 rotatory sub-luxation that manifests itself as a painful torticollis with the head in the so-called cock-robin position (Fig. 2a). The main known predisposing factors are post-traumatic and post-infectious conditions (Grisel's syndrome). In particular, Grisel's syndrome was excluded in this case since no infective processes in the head and neck region were described in the patient PMH. There are also rare case reports of AARF developing after oto-laryngological surgeries, especially when the head was rotated and extended to extreme positions [5].

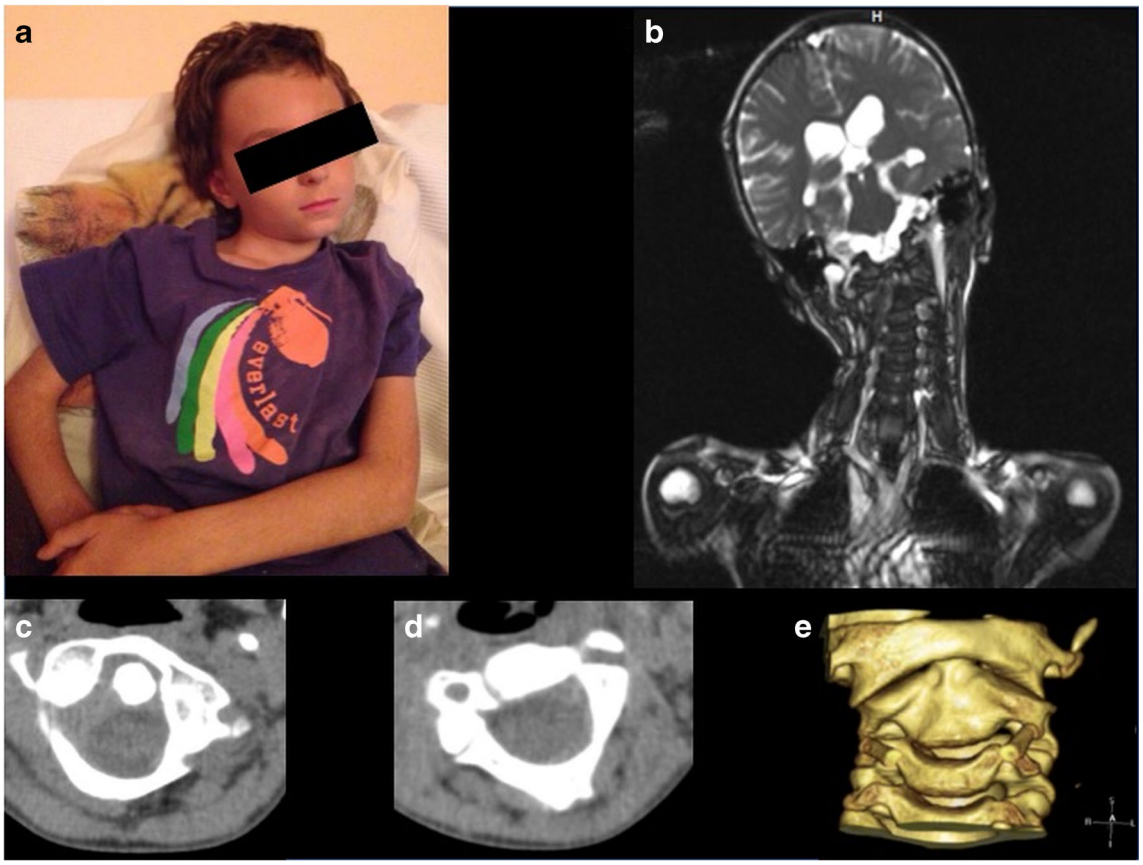

Fig. 2 **a** Pre-operative photograph showing the patient with torticollis and the typical cock-robin head position. **b** Pre-operative coronal MR scout image of the cervical spine showing the torticollis. **c**–**d** Pre-operative axial CT images showing atlanto-axial rotatory fixation (AARF). **e** Pre-operative 3D volumetric CT reconstruction showing C1–C2 sub-luxation with significant facet deformity and lateral inclination of the articular complex

The management algorithm depends upon the type, severity, and time of presentation. The treatment mainly involves reduction and stabilization of the C1–C2 articular complex with a neck brace or a halo-vest or ultimately surgical C1–C2 fusion. We describe the case of a patient developing AARF early after the positioning of a VPS with the presence of a fibro-adherent reaction of the sternocleidomastoid muscle fascia to the catheter. AARF seems to have been produced by the rigid nature of the distal catheter which became encased by fibrous tissue and calcium. Surgical lysis of the fibrous band surrounding the tube solved the torticollis and C1–C2 regained the normal articular anatomy. There are only two similar published cases where torticollis developed after VPS surgery (Table 1). In one case [2], the authors hypothesized that AARF developed because of a combination of multiple surgeries to the CVJ, macrocephaly, and a weak cervical musculature. The sub-luxation was resolved conservatively with cervical traction and halo-vest positioning. Singh et al., instead, presented the case of a boy with torticollis after a VPS performed 12 years earlier, with no radiological evidence of AARF. This child harbored a fibro-calcific band around the shunt tube that led to the displacement of the ventricular catheter and torticollis. Removal of the whole VPS system solved the pathological torticollis [8]. This fibrous reaction on the catheter which leaves it brittle and subject to breakage is common in patients who have had shunt for decades and, in particular, shunts which have been repaired previously with connectors. On the other hand, our presented case highlights the rarity of such early complication after VPS surgery. Based on the presence of a thick and palpable scar-like fibrous band around the cervical portion of the catheter, we concluded that such finding could be the cause of the presenting symptoms.

Among the major risk factors for the development of AARF, we identified ligamentous laxity, a weak cervical

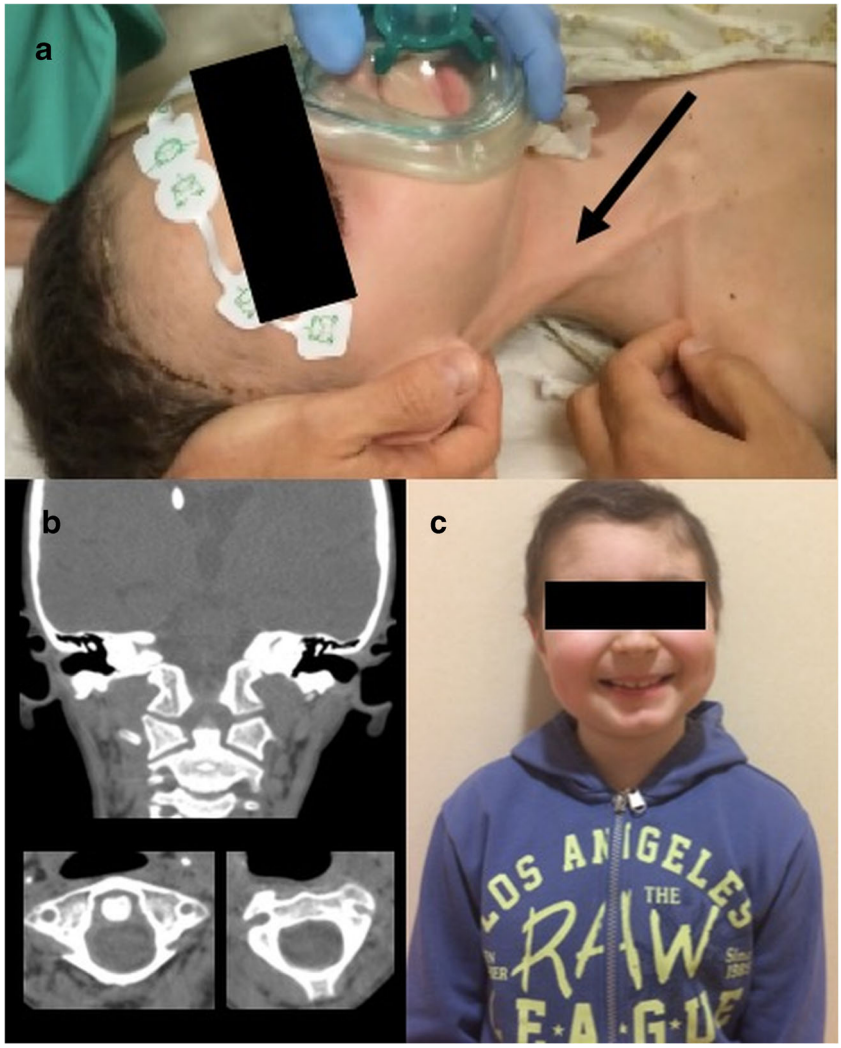

Fig. 3 **a** Photograph showing the subcutaneous fibrous band under tension along the cervical portion of VPS catheter determining torticollis. **b** Post-operative coronal and axial CT image of the cranio-vertebral junction showing the correct realignment of articular relationships of the C1–C2 complex and the resolution of AARF. **c** Last follow-up photograph of the patient with complete resolution of torticollis

musculature, and a history of multiple surgeries entailing extreme head positioning. Allergic reactions to shunt hardware were also considered, but allergy to silicone is quite rare and very few cases are reported in the literature [1, 3]. Therefore, this element was not considered among the main risk factors.

Table 1 Cases published in the literature describing torticollis/AARF as complications after VPS surgery [2, 8]

References	Sex	Age (year)	Primary pathology	Previous VPS surgeries	AARF	Onset of CVJ abnormality after VPS	Treatment
Heary RF et al. [2]	M	10	Congenital hydrocephalus and temporal arachnoidal cyst	Y (3)	Y	Immediate (1st POD)	Traction and reduction, halo-vest
Singh G et al. [8]	M	14	Congenital hydrocephalus Acqueductal stenosis	Y (1)	Unknown (torticollis)	Late (12 years after VPS)	Surgical release of fibrous band
Present case	M	6	Obstructive hydrocephalus Hypothalamic/chiasmatic pylocytic astrocytomas	N	Y	Early (2 months after VPS)	Surgical release of fibrous band

Conclusions

Torticollis and AARF can be early-onset complications after VPS surgery and a thorough assessment of the course of the distal VPS catheter should be conducted before considering CVJ surgical stabilization. In our case report, surgical removal of such a band led to the reduction of C1–C2 sub-luxation with no need of further surgical fusion procedures.

Compliance with ethical standards

Conflict of interest The authors declare that they have no conflict of interest.

Informed consent Informed consent was obtained from the patient and his parents.

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