Retropharyngeal pseudomeningocele formation as a traumatic atlanto-occipital dislocation complication: case report and review

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Received: 5 April 2007 / Revised: 6 August 2007 / Accepted: 13 October 2007 / Published online: 31 October 2007
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Abstract Retropharyngeal pseudomeningocele after atlanto-occipital dislocation is a rare complication, with only five cases described in the literature. It develops when a traumatic dural tear occurs allowing cerebrospinal fluid outflow, and it often appears associated with hydrocephalus. We present a case of a 29-year-old female who suffered a motor vehicle accident causing severe brain trauma and spinal cord injury. At hospital arrival the patient scored three points in the Glasgow Coma Scale. Admission computed tomography of the head and neck demonstrated subarachnoid hemorrhage and atlanto-occipital dislocation. Three weeks later, when impossibility to disconnect her from mechanical ventilation was noticed, a magnetic resonance imaging of the neck showed a large retropharyngeal pseudomeningocele. No radiological evidence of hydrocephalus was documented. Given the poor neurological status of the patient, with spastic quadriplegia and disability to breathe spontaneously due to bulbar-medullary injury, no invasive measure was performed to treat the pseudomeningocele. Retropharyngeal pseudomeningocele after atlanto-occipital dislocation should be managed by means of radiological brain study in order to assess for the presence of hydrocephalus, since these two pathologies often appear associated. If allowed by neurological condition of the patient, shunting procedures such as ventriculo-peritoneal or lumbo-peritoneal shunt placement may be helpful for the treatment of the

pseudomeningocele, regardless of craniocervical junction management.

Keywords Atlanto-occipital dislocation · Pseudomeningocele · Retropharyngeal · Spinal cord injury

Introduction

Atlanto-occipital dislocation (AOD) is a rare cervical injury that entails poor prognosis [4, 6]. It can be occasionally complicated by the development of a retropharyngeal pseudomeningocele, which means an even rarer pathology according to the literature. In fact, only five cases have been documented [2, 8, 10, 11]. A pseudomeningocele develops as the result of cerebrospinal fluid (CSF) outflow and extradural collection formation after a traumatic dural tear. If present, concomitant hydrocephalus can also force CSF diversion [8, 10, 11].

We present a case of severe brain and cervical injury with AOD, complicated by the development of a large retropharyngeal pseudomeningocele as a delayed sequela of this specific craniocervical disease. We also review the literature and briefly describe clinical and radiological features as well as management alternatives concerning this pathology.

Case report

A 29-year-old female was admitted at our hospital after suffering a multi-traumatism as a result of a high-speed motor vehicle collision. When attended at the accident scene she scored five points in the Glasgow Coma Scale (GCS), requiring endotracheal intubation. At hospital

None of the authors have received any financial support for this report.

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arrival, she scored three points in the GCS, her pupils were miotic and sluggishly reactive to light stimulus and no corneal reflex was documented. A computed tomography (CT) of the head demonstrated subarachnoid hemorrhage in basal cisterns and bilateral fracture of sphenoid wings. The CT scan of the neck demonstrated a left occipital condyle fracture with a displaced fragment into the spinal canal as well as AOD (Fig. 1). An intracranial pressure monitor was immediately implanted, but no intracranial hypertension was registered during the whole posttraumatic period. Since patient's condition was poor, AOD was managed by means of a rigid collar.

Twenty-one days after admission, sedation withdrawal was achieved and the patient presented absence of spontaneous respiratory activity, requiring tracheostomy and connection to mechanical ventilation. She also presented spastic quadriplegia. In view of these findings suggesting bulbar-cervical dissociation, magnetic resonance imaging (MRI) of the neck was performed showing high signal cord intensity in T₂-weighted images from bulbar region to C3 level, consistent with bulbar-medullar edema and/or ischemia. A cystic collection in the retropharyngeal space, 77 × 30 mm in size, was also identified. It extended from

occipito-cervical junction to C4 level and it was isointense compared with the CSF signal on all sequences (Fig. 2a–c). All these findings were consistent with retropharyngeal pseudomeningocele. Computed tomography of the head revealed no evidence of hydrocephalus. Conservative management of the pseudomeningocele was decided given the dreadful neurological condition of the patient.

The patient remained in a vegetative state and ventilator dependent, dying 2 years after the accident.

Discussion

Pseudomeningocele is an extradural collection of CSF that diverts through a dural tear and becomes encapsulated in a fibrous sac. The most common etiology is iatrogenic, especially as a consequence of lumbar spine surgery and, less frequently, following cervical spine surgery, posterior fossa surgery or lumbar puncture. In this context, factors such as incomplete dural closure, traumatic dural insult during the intervention or coexistence of hydrocephalus have been involved in its genesis [3, 5, 7]. Besides, less frequent are traumatic and congenital causes. Traumatic pseudomeningoceles are rare events usually organized in the posterior spinal region in connection with the perineural space, resulting from lumbar or cervical nerve root avulsions [3, 7]. Retropharyngeal pseudomeningoceles are considered particular phenomena, since only seven reports have been described in the literature, five of them as a sequela of AOD [2, 8, 10, 11] and the remaining two, derived from atlanto-axial dislocation [1, 9].

Atlanto-occipital dislocation is an unstable injury in the upper cervical spine, traditionally produced by high-energy traumatism. It affects the occipito-cervical junction ligaments more than the vertebral joints, and its true incidence is unknown, although may be low. It entails increased morbi-mortality, presumably because of the close anatomic relationship between this region and vital brainstem-medullar structures or the vasculature which supplies them [4, 6].

It is known that the traumatism can cause a nerve root avulsion, a joint dislocation or a vertebral fracture that, at the same time, originates a dural tear which offers low resistance and helps CSF outflow to surrounding soft tissues developing in this way, a cyst [3, 7]. Provided that the patient presents hydrocephalus, as observed in four of the five cases previously reported of retropharyngeal pseudomeningocele secondary to AOD, increased CSF pressure may force its diversion [8, 10, 11].

Retropharyngeal pseudomeningocele usually appears in a delayed fashion, some weeks after the initial trauma (Table 1). Symptoms often derive from the mass effect when the cyst reaches significant size. The most common

Fig. 1 **a** Coronal reformat of CT demonstrates left occipital condyle fragment and dislocation of the left occipital condyle from the left lateral mass of C1. **b** Sagittal reformat of CT demonstrates the usual distance between the right occipital condyle and the right lateral mass of the C1 compared to the significant displacement of the left occipital condyle relative to the left lateral mass of C1; Both images, **a** and **b**, are consistent with atlanto-occipital dislocation diagnosis. **c** Axial CT scan of the neck demonstrates a left occipital condyle fragment displaced into the spinal canal

Fig. 2 MRI study of the neck. Sagittal (a) T₁-weighted, (b) T₂-weighted and (c) proton density weighted images show a retropharyngeal mass, with the same signal intensity as CSF, 77 × 30 mm in size, consistent with retropharyngeal pseudomeningocele. a High signal intensity in T₁-weighted image in the bulbar-medullar joint

suggests hemorrhage. b Sagittal T₂-weighted image shows high signal intensity from bulb to C3 level consistent with edema-ischemia. Axial T₂-weighted images (d) and (e) suggest that the primarily dural tear source of the collection was presumably due to C1 root avulsion

initial symptoms are respiratory failure and dysphagia, although sometimes the cyst is an incidental finding in a radiological study performed with a different purpose. In these patients, respiratory distress can be attributed, on one hand, to neurological damage due to the cervical injury and, on the other hand, to the retropharyngeal space-occupying mass that may obstruct the airway [11]. In our patient, respiratory debut was noticed when disconnection from assisted ventilation was impossible. Considering the radiological findings, it seems reasonable to assume that bulbar-medullar injury could be considered the main cause of respiratory failure in our case. However, we can not define the true importance of pseudomeningocele in the aforementioned failure.

Magnetic resonance imaging has showed superior to CT regarding diagnosis of spinal cord and soft tissue injuries and therefore, is considered the mainstay diagnostic procedure to confirm the presence of retropharyngeal pseudomeningocele. Pseudomeningocele is characteristically identified as a cystic collection with signal intensity

consistent with CSF on all sequences. More complete studies such as CSF flow imaging or CT myelography can be helpful in order to identify the communication between the cyst and subarachnoid space in those cases where conventional MRI yields negative [3, 7]. Once the diagnosis is confirmed, performing a cranial neuroimaging study is recommended in order to assess for the presence of hydrocephalus, as these two pathologies often appear associated. In the case we present, probably a left C1 root avulsion secondary to AOD distraction produced the dural tear which originated the collection, as we can check in the MRI study (Fig. 2d, e). However, CT of the brain ruled out the presence of concomitant hydrocephalus.

Conservative management of retropharyngeal pseudomeningocele, such as bed rest, head of bed elevation, acetazolamide and/or osmotic diuretics may be initially attempted. Nevertheless, this therapeutic option failed in those cases reported by Natale et al. [9] and Cognetti et al. [2]. Surgical alternatives include shunting procedures to reduce CSF pressure gradient or even collection removal

Table 1 Reported cases of posttraumatic retropharyngeal pseudomeningocele

Author	Age	Ethiology	Symptoms	Delay	Hydrocephalus	Treatment	Results
Williams et al. [11]	3.5	AOD	Respiratory + Dysphagia	4 weeks	Yes	VP shunt	Resolution
Naso et al. [8]	26	AOD	Respiratory + Dysphagia	3.5 months	Yes	VP shunt	Resolution
	11	AOD	Respiratory?	5 weeks	Yes	None	Exitus
Reed et al. [10]	9	AOD	Incidental	4 weeks	Yes	None	Exitus
Cognetti et al. [2]	19	AOD	Dysphagia	6 weeks	No	LP shunt	Resolution
Present case	29	AOD	Respiratory	3 weeks	No	None	Exitus
Natale et al. [9]	33	transient C1-C2?	Respiratory + Dysphagia	Few days	No	LP shunt	Resolution
Achawal et al. [1]	38	C1-C2	Plegia	4 weeks	No	LP shunt	Exitus

AOD atlanto-occipital dislocation, VP ventriculo-peritoneal, LP lumbo-peritoneal

and direct closure of the dural tear [3, 5, 7]. According to the literature, patients with concomitant hydrocephalus underwent ventriculo-peritoneal shunt placement [8, 11] while patients in absence of hydrocephalus were treated by means of lumbo-peritoneal shunt [1, 2, 9]. None of the previously published patients underwent direct closure of the dural tear, probably because of the difficulty of this type of surgical approach. In the case we present in this paper, the dreadful neurological condition of the patient prevented that surgical management was considered.

Successful recovery from pseudomeningocele is usually achieved when proper diagnosis and treatment is provided [2, 8, 9, 11]. Outcome in these patients is determined by the original traumatism and other concomitant injuries, not by the pseudomeningocele itself. Atlanto-occipital dislocation entails poor prognosis and 30-day mortality rate among patients who arrive alive at hospital is 35%, figures that reflect the fatal outcome of this injury [6].

Conclusions

Traumatic AOD may be complicated by the development of a retropharyngeal pseudomeningocele. These patients often present hydrocephalus and therefore brain neuroimaging study is recommended in order to exclude this association.

When medically feasible, the pseudomeningocele can be treated, among other procedures, by means of ventriculo or lumbo-peritoneal shunt placement (depending on the presence or absence of hydrocephalus, respectively) or surgical repair of the dural defect, regardless of the AOD management.

Acknowledgments We thank Cristina Ruiz Quevedo for assistance in the translation of this manuscript. None of the authors have received any financial support for this work.

Conflict of interest statement None of the authors has any potential conflict of interest.

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