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Case report

Pseudo-Grisel's syndrome as a complication of adenotonsillectomy

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Abstract

Introduction: Grisel's syndrome is non-traumatic subluxation of the atlanto--axial joint unrelated to any disorder of the skeletal system. It is a rare complication of infections in the ENT region and surgeries in the head and neck region. Symptoms of Grisel's syndrome include: pain in the neck, nuchal rigidity or torticollis.

Aim: The aim of the study is to present Grisel's syndrome as one of possible complications following surgeries in the ENT region.

Case study: In our article we present a case of a child, who was suspected to suffer from Grisel's syndrome after adenotonsillotomy.

Results and discussion: Grisel's syndrome can be a consequence of nasal, throat and ear infections; and it can result from head and neck surgeries, such as tonsillectomy, adenoidectomy, mastoidectomy. Symptoms of Grisel's syndrome are: nuchal rigidity or torticollis, and pain in the neck which occurs a few days after the infection or surgical procedure. When diagnosing Grisel's syndrome, it is recommended to perform X-ray images in AP and lateral projection on the atlas, as well as a CT scan and MRI scan of the neck. The treatment can be conservative or operative and it is is based on the degree of subluxation and the severity of clinical symptoms.

Conclusions: Medical imaging and comprehensive treatment as soon as possible is of paramount importance in order to avoid dangerous consequences, such as permanent deformation of the neck or neurological losses which would require extensive surgical procedures.

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1. INTRODUCTION

Grisel's syndrom is a non-traumatic subluxation of the atlanto-axial joint unrelated to any disorder of the skeletal system.[1] It is a rare complication of infections in the ENT region and surgeries in the head and neck region.[1] The set of symptoms comprising Grisel's syndrome was first presented by Charles Bell in 1830.[1] He described a patient with a parapharyngeal abscess in the course of syphilis. The patient died due to medulla oblongata compression which occurred as a result of atlanto-axial subluxation.[1,2] In the post-mortem examination an ulceration of the transverse ligament of the axis was revealed.[3] In 1930, a French physician Pierre Grisel, who gave his name to the disease, described two cases of subluxation of the atlanto-axial joint in children with history of nasopharyngeal infection.[1–5]

Grisel's syndrome can result from an infection in the area of nose, throat, and middle ear; it can also occur subsequent to head and neck surgeries, such as adenoidectomy, tonsillectomy, mastoidectomy, and others. Symptoms of Grisel's syndrome include: pain in the neck, nuchal rigidity or torticollis which occur a few days after the infection or surgery. The diagnosis of Grisel's syndrome consists in X-ray examination of the cervical spine, as well as a CT scan or an MRI scan of the neck. The recommended treatment depends on the degree of subluxation and the severity of clinical symptoms.[3]

2. AIM

The aim of the study is to present Grisel's syndrome as one of the possible complications following surgeries in the ENT region. The presented case concerns a 6-year-old girl after adenotonsillectomy who reported pain accompanied by nuchal rigidity in the 2nd day after the surgery. The girl was suspected to suffer from Grisel's syndrome.

3. CASE STUDY

A 6-year-old girl was admitted to the Department of Laryngology of the Regional Specialised Children's Hospital in Olsztyn with enlarged tonsils – pharyngeal and palatine – in order to undergo surgical treatment. The surgery was performed under general anaesthesia, the head was not tilted backwards. The pharyngeal tonsil was removed with the use of Beckmann adenoid currete. After excising it, quite profuse bleeding from the nasopharynx occurred. Compression to achieve hemostasis was not effective, so bipolar electrocoagulation was used to stop the bleeding. Next, palatine tonsils were trimmed with a loop. The bleeding was moderate, and it stopped spontaneously. The patient was woken up in the operating room with no complications. During the stay in the postoperative ward and later in the Department of Laryngology, no bleeding was observed. In the 2nd day after the procedure in the afternoon, the temperature increased to 37.8°C, and pain in the neck and nape occurred. An abnormal position of the head was observed – a slight rotation to the right and tilt towards the right shoulder, with limited movement of the head when turning it sideways and tilting forwards. The wound healing in the pharynx did not show any symptoms of infection. In the postoperative period, paracetamol and ibuprofen were used to alleviate the pain, Etamsylate was administered preventively. The laboratory tests were normal, with an elevated level of CRP. Because of this and subfebrile temperature, amoxicillin with clavulanic acid were administered intravenously. The temperature decreased, pain in the neck and limitation of its movement subsided. On the third postoperative day the child was discharged on the parents' demand. The recommendations included continuing antibiotic, ibuprofen and paracetamol treatment.

As the subfebrile temperature reaching 37.5°C persisted, accompanied by limitation of head movement (yet not as severe as before), abnormal position of the head tilted to the side and pain the neck when moving it, on the 11th postoperative day the child was readmitted to the department. The laboratory tests were normal. The physical examination showed: the healed wound in the pharynx, no symptoms of infection, the nose unobstructed, no discharge in the nose. Because of suspected Grisel's syndrome, an MRI scan of the neck was performed under general anaesthesia (the parents did not agree for the child to have a CT scan performed). Description of the findings: in the pharyngeal tonsil fossa upwards from the skull base and downwards to the border of the body and the base of the axial dens, a layer of fluid – evolving blood AP 8 mm, CC 19 mm – postoperative lesions. Slight linear post-contrast enhancement of the mucous membrane of the posterior wall of the nasopharynx – most probably reactionary. No pathological post-contrast enhancement of the tissues and no abscess in the retropharyngeal/prespinal space. No visible pathological lesions in the paraspinal muscles. No symptoms of subluxation or luxation in the atlanto-occipital joint and the atlanto-axial joint. The distance between the anterior arch of C1 and the anterior surface of the axial dens – 2 mm (normal). Pterygoid muscles, masticators, sternocleidomastoid muscles bilaterally symmetrical, with no focal lesions. Parotid and maxillary glands and structures of the throat and larynx normal. Upper cervical lymph nodes: right 15 × 8 mm, left 17 × 8 mm; lower cervical lymph nodes not enlarged. The lateral retropharyngeal lymph node: right 9 × 7 mm, left 8 × 4 mm. Supracalvicular fossae was clear (Figure 1).

After the MRI examination a consultation of a physician specialising in rehabilitation was requested and he did not find it necessary to stabilise the spine or administer physiotherapy. The child was discharged with a recommendation to return for a follow-up in the Rehabilitation Centre after 2 weeks. In this period pain in the neck disappeared and its mobility came back to normal. The child had a follow-up appointment in the laryngological centre after 6 weeks – the pharynx was healed, the nose unobstructed, mobility of the neck normal without pain.

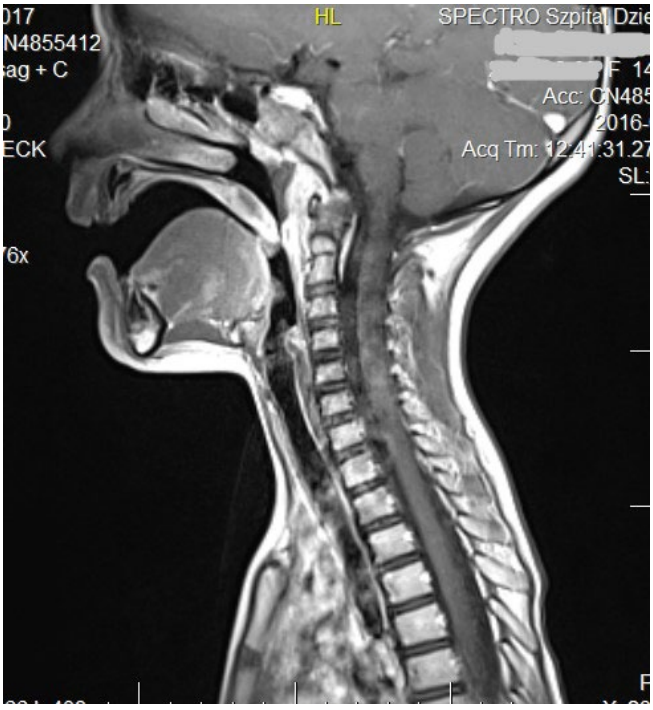

Figure 1. Inflammatory swelling of the mucous membrane of the nasopharyngeal posterior wall and the normal position of vertebrae in the cervical spine (MRI scan).

4. RESULTS AND DISCUSSION

The most common complications of otolaryngological surgeries in children include: reactionary and secondary postoperative haemorrhages, infections of the operative wound, earache, otorrhoea subsequent to drainage, obstruction in the nose. Other more severe complications occur more rarely, for instance open nasality, leaking of food (mainly liquids) into the nose, or a trauma of prevertebral fascia which causes nuchal rigidity and irritation of the spine, which in turn may lead to Grisel's syndrome.[6]

Grisel's syndrome is a non-traumatic subluxation of the atlanto-axial joint unrelated to any disorder of the skeletal system. Grisel's syndrome pathogenesis is not fully known. It is certain that this syndrome occurs more often in children aged 5–12 than in other age groups, these children constitute 68% of Grisle's syndrome patients.[3,6]

Grisel's syndrome can be a consequence of nasopharynx infection, throat inflammation, tonsillitis, acute otitis media, pharyngeal tonsil inflammation, acute parotid gland inflammation, peritonsillar abscess, neck abscess; and it can result from head and neck surgeries, such as tonsillectomy, adenoidectomy, mastoidectomy, a surgery unblocking posterior nasal apertures or veloplasty.[1–3,7,8] Symptoms of Grisel's syndrome can follow an oligosymptomatic upper respiratory tract infection or even an infection which parents do not notice in their children. This syndrome is observed in people with Down syndrome in whom an increased distance between C1 and C2 vertebrae appears, as well as in Marfan syndrome patients who show excessive ligament instability, and in Klippel–Feil syndrome patients, and those with brittle bone disease and neurofibromatosis.[1,3,4]

Symptoms of Grisel's syndrome are: nuchal rigidity or torticollis, and pain in the neck which occurs a few days after the infection or surgical procedure. Symptoms result from hyperaemia of the tissues in the nasopharynx caused by an infection or inflammation subsequent to a surgical procedure performed in the adjacent area. The inflammatory process which spreads through the blood circulation via pharyngeal-vertebral veins, which go through prevertebral fascia, leads to decalcification of the anterior arch of the atlas and laxity of the anterior longitudinal ligament, which is the main stabiliser of the atlanto-axial joint between the first and the second cervical vertebrae, which in turn can lead to subluxation in the atlanto-axial joint.[1,2,5–8]

The physical examination in the presented case revealed an abnormal position of the head, the so called 'cock-robin position' – the head was tilted to the side and slightly rotated.[1] The patient reported pain in the neck while tilting the head forward or twisting it to the sides.

Clinically, Gisel's syndrome is diagnosed when three of the below presented symptoms occur simultaneously:[1,3,4]

(1) On palpation, it can be determined that the spinous process of C2 is dislocated towards the side in which the head rotates – Sudeck's sign;[3,6]
(2) The ipsilateral sternocleidomastoid muscle is contracted and as a result the head rotates and is kept in this position in order to minimize the pain;
(3) It is impossible for the patient to rotate the head beyond the midline of the body in the direction opposite to the one where the damage occurs.

When diagnosing Grisel's syndrome, it is recommended to perform X-ray images in AP and lateral projection on the atlas, as well as a CT scan and MRI scan of the neck, which will make it possible to assess the surrounding soft tissues and the spinal cord.

In differential diagnosis of Grisel's syndrome it is necessary to consider the tumours of the posterior cranial fossa and the spinal cord, Arnold-Chiari malformation, spinal gliosis, neoplasms of the spine, fractures in the area of C1–C2.

The classification of subluxation of the atlanto-axial joint proposed by Fielding and Hawkins comprises 4 degrees, out of which 1 and 2 are most common and do not give serious consequences,[1,3] while 3 and 4 occur more rarely, yet they result in more serious consequences such as compression of the spinal cord.

The first degree is rotation of C1 without displacement of the C1 vertebral body in relation to C2 on a lateral scan (displacement lower than 3 mm). The second degree is rotation of C1 with a 3–5 mm anterior displacement on a lateral scan. The third degree is rotation of C1 with an anterior displacement greater than 5 mm on a lateral scan. The fourth degree is a posterior displacement of C1 onto C2 on a lateral scan.

Having diagnosed Grisel's syndrome, it is necessary to implement treatment as soon as possible in order to avoid neurological complications – from less serious ones, such as parasthesia, through tetraplegia and bladder function

disorders, to paralysis of the respiratory centre. Conservative treatment methods include rest in a supine position, 4–6-week-long antibiotic therapy against aerobic and anaerobic bacteria in the nasopharynx, muscle relaxants, anti-inflammatory medicines. Other treatment methods depend on the degree of subluxation, its duration and severity of symptoms.[3,6] The treatment should be conducted by an orthopaedist or neurosurgeon. Patients of 1 and 2 Fielding and Hawkins degree should be stabilised with a cervical. Patients of type 3 and 4 should be treated with skeletal traction applied in the cervical spine region. Operative treatment consisting in atlanto-axial stabilisation is applied when conservative treatment is ineffective and when subluxation symptoms recur. Operative treatment is also necessary if neurological symptoms occur.[1,7]

The atlanto-axial joint is a typical presentation area of subluxation in Grisel's syndrome. However, the literature of the subjects presents cases in which symptoms typical of Grisel's syndrome occurred but subluxation was located in a different segment of the cervical spine. Martinez-Lange et al.[8] described a case in which subluxation in the C2–C3 joint occurred after sinusitis.[1,8] Lopes and Li[8] presented two patients with Grisel's syndrome symptoms in whom subluxation occurred in the C3–C4 join.[1,8] Hettiaratchy et al.[8] described a case of coexisting subluxation in the C1–C2 and C2–C3 joints in a patient after upper respiratory tract infection, and they believe that pathogenesis of subluxation is similar to that in Grisel's syndrome – an inflammatory state, hyperaemia of tissues and excessive laxity of the ligaments. Jay Ching Chieh Wang et al.[5] described a case of unilateral hypoplasia of the facial skeleton and microtia in which the set of symptoms typical of Grisel's symptom appeared subsequent to a surgical procedure of auricular reconstruction. In this case, pathogenesis took into account not the inflammation but the coexistence of irregularities in the spinal structure or the traumatic mechanism which appeared when the patient's head was rotated during the surgery.

Literature of the subject reports that Grisel's syndrome follows laryngological procedures, while its second cause is history of upper respiratory tract infections.[7,9]

Deichmueller and Welkoborsky[9] analysed all Grisel's syndrome cases in their centres in Hanover in 2002–2009. Out of 12 cases, 8 occurred in patients after surgical procedures in the nasopharynx and pharynx, 5 of whom were patients after adenoidectomy and tympanostomy). Four remaining patients were found to have upper respiratory tract infections, in 3 of them it was tonsillitis.

Bocciolini et al. believe that applying electrocoagulation to stop bleeding following adenoidectomy can be a risk factor for Grisel's syndrome.[2] They recommend preventive administration of an antibiotic in such patients.

Grisel's syndrome is a very rare complication of upper respiratory tract infections and surgeries in the ENT region. Yet in every case when a child with history of infection of laryngological procedure suffers from a sudden pain in the neck with its limited mobility, one has to consider the occurrence of this syndrome.

5. CONCLUSIONS

(1) Profuse bleeding and use of electrocoagulation hemostasis could cause inflammation in the nasopharyngeal tissues which spread into the prevertebral space and caused pain in the neck.
(2) In the presented case, the MRI scan did not confirm subluxation in the atlanto-axial joint, and the administered treatment (antibiotic and anti-inflammatory medicines) was effective and led to removal of pain symptoms.
(3) Implementing medical imaging and comprehensive treatment as soon as possible is of paramount importance in order to avoid dangerous consequences, such as permanent deformation of the neck or neurological losses which would require extensive surgical procedures.

Conflict of interest

None declared.

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