

REVIEW ARTICLE

Dysphagia as a primary manifestation of basilar impression: A case report

Andrea Ruiz Hernández^{a,*}, Elena González Arnáiz^b^a Servicio de Neurología del Complejo Asistencial Universitario de León, Spain^b Servicio de Endocrinología y Nutrición del Complejo Asistencial Universitario de León, Spain

Received 26 December 2023; accepted 8 January 2024

Available online 16 February 2024

KEYWORDS

Basilar impression;
Bone diseases;
Dysphagia;
Malnutrition

Abstract Secondary basilar invagination or basilar impression is an anomaly at the craniovertebral junction where the odontoid process prolapses into the foramen magnum with the risk of compressing adjacent structures and obstructing the proper flow of cerebrospinal fluid (CSF). The incidence is less than 1% in the general population and occurs mainly in the first three decades of life when it is associated with malformations of the neuroaxis. In older age, the main aetiologies are diseases that alter bone mineral density. The clinical course is usually progressive and the most common symptoms are asthenia, cervical pain and restricted movement, but also dysphonia, dyspnoea and dysphagia. It is a progressive disease which, if left untreated, can cause severe neurological damage and death. We report the case of a 79-year-old woman with osteoporosis and progressive dysphagia leading to severe malnutrition, which conditioned the decision not to intervene due to the high perioperative risk.

© 2024 SEEN y SED. Published by Elsevier España, S.L.U. All rights reserved.

PALABRAS CLAVE

Impresión basilar;
Enfermedades óseas;
Disfagia;
Malnutrición

Disfagia como manifestación inicial de impresión basilar: a propósito de un caso

Resumen La invaginación basilar secundaria o impresión basilar es una anomalía de la unión craneovertebral en la que la apófisis odontoides prolapsa en el foramen magnum con el riesgo de comprimir estructuras adyacentes y obstruir el correcto flujo del líquido cefalorraquídeo. Presenta una incidencia inferior al 1% en la población general y se produce principalmente en las 3 primeras décadas de la vida, cuando se asocia a malformaciones del neuroeje. En edades avanzadas, las principales etiologías son enfermedades que alteran la densidad mineral ósea. La evolución clínica suele ser progresiva y los síntomas más frecuentes son: astenia, dolor

* Corresponding author.

E-mail address: aruizh@saludcastillayleon.es (A. Ruiz Hernández).

cervical y restricción de movimientos, si bien también disfonía, disnea y disfagia. Se trata de una enfermedad progresiva que, si no se trata, puede causar daños neurológicos graves y la muerte. Presentamos el caso de una mujer de 79 años con osteoporosis y disfagia progresiva que derivó en desnutrición grave, lo que condicionó la decisión de no intervenirla por el alto riesgo perioperatorio.

© 2024 SEEN y SED. Publicado por Elsevier España, S.L.U. Todos los derechos reservados.

Case description

Basilar impression is an alteration of the craniovertebral junction where the odontoid process prolapses into the foramen magnum with the associated neurological and life-threatening risks. The terms basilar invagination and basilar impression are often incorrectly used as synonyms in the craniovertebral junction literature, but basilar impression refers to the acquired form of basilar invagination resulting from softening of the bone at the base of the skull. It is a rare condition in the elderly, caused by diseases that alter bone mineral density, mainly rheumatoid arthritis, Paget's disease, osteomalacia, bone tumours and infections, hyperparathyroidism and osteogenesis imperfecta.¹⁻³

We report the case of a patient whose main clinical symptom was dysphagia and who was diagnosed with a basilar impression.

The patient was a 79-year-old woman whose reason for consulting was progressive asthenia and dysphagia. Her medical personal history included euthyroid multinodular disease since 2021 and osteoporosis since 2018 treated with denosumab, calcium and vitamin D. She had no known cardiovascular risk factors, harmful lifestyle habits or surgical procedures. She had no cognitive impairment and lived at home, but needed help with basic activities and used a walking stick. Her usual treatment consisted of denosumab 60 mg subcutaneously every 6 months, chondroitin sulphate 800 mg daily, cholecalciferol/calcium pidolate 400/600 mg daily, esomeprazole 20 mg daily, lorazepam 1 mg daily and paracetamol 1 g/8 h.

In the anamnesis the patient reported that her symptoms had started 2–3 years ago and had worsened in the last 6 months. She complained of dysphonia, a sensation of instability with some falls without cranioencephalic trauma and progressive fine motor clumsiness. She reported a weight loss of 20 kg in the last 2–3 years due to hyporexia and mixed dysphagia with foreign body sensation in the oropharynx, without infectious processes suggesting bronchoaspiration. Due to the patient's clinical condition and in order to complete the study, it was decided to admit the patient to hospital.

Physical examination revealed severe cachexia with generalised muscle wasting. There were no abnormalities on cardiorespiratory auscultation or abdominal examination, no lower limb oedema or signs of venous pathology. Neurological examination showed dysphonia with nasal voice and generalised symmetrical vivid osteotendinous reflexes, preserved muscle tone and strength without fasciculations

with significant generalised atrophy, no changes in speech, eye movements, pupils, campimetry, facial musculature or other lower cranial nerve pairs with symmetric elevation of the palatine pillars and normal position of the lingula, no altered sensitivity, no dysmetria of the limbs and no gait ataxia, but she required two supports to walk.

Morpho-functional assessment was performed by the Endocrinology and Nutrition Department at the time of admission: height 143 cm, usual weight 50 kg, current weight 30 kg, weight loss 40% in 3 years, ideal weight 47 kg, corrected weight 41.4 kg and BMI 14.6 kg/m², hand grip strength 8 kg, calf circumference (CC) adjusted to BMI of 30 cm, corrected basal energy expenditure of 951 kcal, total energy expenditure (corrected with a stress factor of 1.2) of 1142 kcal, protein requirement of 56–70 g. The 24-h intake record of the shredded diet was 30% (900 kcal and 40 g of protein).

With the diagnosis of severe malnutrition based on the GLIM criteria, mixed dysphagia and high risk of refeeding syndrome, additional tests were performed to aid in the aetiological diagnosis. Blood tests showed prealbumin of 17 mg/dl, retinol binding protein (RBP) of 2.5 mg/dl and normocytic normochromic anaemia with Hb 10.1 g/dl. Other parameters, including renal, hepatic and lipid profiles, vitamins B12, B1 and D, coagulation, thyroid hormones, autoimmune profile and tumour markers were in the normal range; and an endoscopy and a cervico-thoracic-abdominal computed tomography (CT) scan ruled out structural and tumour pathologies.

Finally, cranio-cervical magnetic resonance imaging (MRI) was performed, which showed craniometric changes at the cranio-cervical junction, with marked basilar invagination (Fig. 1), with impression of the medulla oblongata and bulbo-medullary junction, without signal intensity changes.

The patient was assessed by the Neurosurgery Department who, in agreement with the patient's family and the patient herself, declined surgery due to the high perioperative risk.

Management and evidence

Basilar impression is an acquired condition resulting from a change in bone tissue at the craniovertebral junction. Diagnosis is based on clinical and imaging studies demonstrating protrusion of the odontoid process into the foramen magnum beyond normal limits established by radiographic reference lines (Fig. 2). The most common signs and symptoms are asthenia, cervical pain and limited movement,

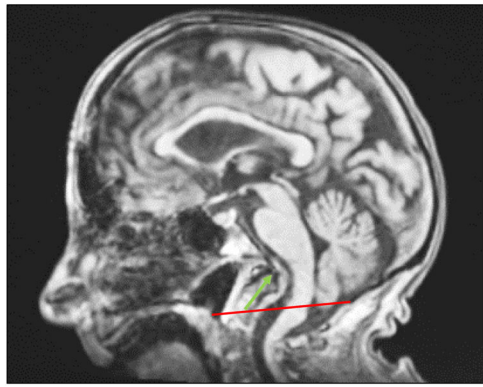


Figure 1 Sagittal cranio-cervical MRI. Chamberlain's line (red mark) is exceeded by the odontoid process (green mark) by >5 mm, defining a marked basilar invagination, imprinting the medulla oblongata and the medullary-bulbar junction.

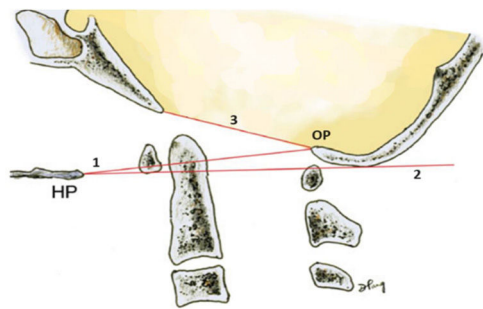


Figure 2 Reference lines. 1 Chamberlain's line between hard palate (HP) and opisthion (OP). Basilar impression is present if the odontoid process protrudes >5 mm above this line. 2 McGregor's line between HP and the lowest point of occiput, positive diagnosis if odontoid process protrudes >2.5 mm above line. 3 McRae's line between basion and opisthion should be above the odontoid process.

Based on: Pang D, Thompson DN. Embryology and bony malformations of the craniovertebral junction. *Childs Nerv Syst.* 2011 Apr;27(4):523–64. doi: 10.1007/s00381-010-1358-9 [Epub 31.12.10; PMID: 21193993; PMCID: PMC3055990].

and symptoms secondary to medulla oblongata and cervical spinal cord compression and CSF flow obstruction. An aetiological study should be performed, including diseases that cause changes in bone mineral density, mainly ruling out rheumatoid arthritis, Paget's disease, osteomalacia and osteoporosis, hyperparathyroidism, osteogenesis imperfecta, Hurler syndrome, and skull base tumours and infections.^{1–3}

This is a progressive disease and treatment is based on correction of the cranio-cervical junction with neurosurgical management and treatment of the underlying aetiology. The type of surgical intervention must be individualised and depends on certain characteristics of the patient, the presence or absence of cranial nerve deficits, C1-C2 mobility, imaging findings, being often complicated by abnormalities such as cranial settling, autofusion of cervical vertebrae, platybasia, and syringomyelia, and the coexistence of other medical conditions that increase perioperative complications. Due to the mechanical and anatomical com-

plexities, surgical treatment involves varying combinations of anterior or posterior decompression with or without traction or fusion. Common approaches to decompression include endonasal and transoral ventral approaches (transoral atlantoaxial reduction plate (TARP) fixation) or occipital cervical posterior decompression and fusion.^{3,4}

In our case, the patient could not be operated on because of her severe malnutrition, which makes the surgical risk too high, and her dependent baseline standard of living. Treatment focused on management of oropharyngeal dysphagia with clinical assessment of safety and efficacy of swallowing and detection of silent aspirations at the bedside. A modified swallowing assessment (MSA)^{5–7} was performed with negative results, and a fiberoptic endoscopic evaluation of swallowing (FEES) was attempted, but it was not possible to perform a complete evaluation due to the protrusion of the posterior pharyngeal wall from the cavum to the hypopharynx, which did not allow the passage of the fibroscope; it was possible to visualise the vocal cords which were mobile without lesions, with an adequate glottic lumen and no salivary retention. A diet of blended solids, free water in small volumes and hypercaloric and hyperproteic oral nutrition supplements were prescribed with good tolerance and no direct or indirect signs of bronchoaspiration. In addition, to avoid refeeding syndrome,⁸ thiamine and a multivitamin supplement were prescribed while she was in the hospital. The patient was discharged from the hospital with nutritional recommendations and treatment, maintaining the previous osteoporosis treatment, and was followed up by the nutrition service.

After six months, the patient underwent a nutritional re-evaluation with significant weight recovery (weight 51.8 kg) and improvement in analytical and functional parameters (prealbumin 22.9 mg/dL, RBP 4.7 mg/dL, hand grip strength 12 kg, CC 32 cm). Symptoms had improved, with less asthenia and good oral tolerance, enabling a better quality of life, but the rest of the symptoms persisted.

Areas of uncertainty

One of the things we were concerned about was the presence of cranial nerve deficits such as dysphagia and dysphonia due to brainstem compression. The literature reports that compression of the spinal cord and medulla oblongata can lead to severe neurological injury, and persistent compression can lead to respiratory compromise with high mortality rates. When these symptoms are present, neurosurgical decompression is required.^{3,4} A review of the literature shows that patients with oropharyngeal disease or a small oral space, as in our case, were preferentially treated with occipitocervical fixation, although this has a significantly higher intraoperative blood loss, longer operative time and lower rates of bone fusion at 3 and 6 months postoperatively than TARP.⁹ In this case, when the perioperative risks were assessed, they were too high to be taken. Furthermore, it has also been reported that preoperative nutritional support must be provided to patients with compromised nutritional status, and that failure to do so may result in wound dehiscence and even non-fusion.^{1,3,9,10}

Another concern was what to do in cases where intervention was not possible. Sasun et al.¹¹ published the case of a

patient with basilar impression due to degenerative spondylolisthesis, with clinical symptoms of proximal upper limb weakness and torticollis. In this case, rehabilitation was chosen with improvement in symptoms, allowing independent living. There are no published cases of conservative management with clinical manifestations of cranial nerve deficits and their outcomes, neither there are cases published of basilar invagination isolated secondary to osteoporosis.

Guidelines

On the basis of the literature reviewed, there are no well-established studies that clearly delineate a treatment algorithm for this condition. Based on the review of the literature, intervention should be considered in all patients due to its progressive course, but is of particular importance in those with high cervico-medullary and brainstem compression.^{1-4,9,10} Throughout the management triage process, careful preoperative planning and perioperative risk assessments should be undertaken. Treatment of symptoms and the treatment of the underlying aetiology must be carried out. In our case the patient's assessment was in accordance with the GLIM guidelines for malnutrition¹² and guidelines for the risk of refeeding syndrome,¹³ the nutritional treatment was carried out in accordance with the ESPEN guideline,¹⁴ dysphagia was managed following the guidelines of the European Society for Swallowing Disorders,^{15,16} and to treat osteoporosis the European guidance for the diagnosis and management of osteoporosis in postmenopausal women were followed.¹⁷

Conclusions and recommendations

In conclusion, basilar impression is a progressive disease that can lead to severe neurological damage and even death if left untreated.^{1-4,9,10} This case shows that dysphagia can be a leading symptom and its management is essential to ensure proper nutritional status, reduce perioperative risk and allow for better recovery.

Conflict of interest

The authors declare that they have no conflict of interest.

References

- Smith JS, Shaffrey CI, Abel MF, Menezes AH. Basilar invagination. *Neurosurgery*. 2010;66 Suppl.:39-47, <http://dx.doi.org/10.1227/01.NEU.0000365770.10690.6F> [PMID: 20173526].
- Pinter NK, McVige J, Mechtler L. Basilar invagination, basilar impression, and platybasia: clinical and imaging aspects. *Curr Pain Headache Rep*. 2016;20:49, <http://dx.doi.org/10.1007/s11916-016-0580-x> [PMID: 27344347.5].
- Menezes AH. Surgical approaches: postoperative care and complications "transoral-transpalatopharyngeal approach to the craniocervical junction". *Childs Nerv Syst*. 2008;24:1187-93, <http://dx.doi.org/10.1007/s00381-008-0599-3> [Epub 04.04.08; PMID: 18389262].
- Wang J, Zhu C, Xia H. Management of unique basilar invagination combined with C1 prolapsing into the foramen magnum in children: report of 2 cases. *World Neurosurg*. 2019;127:92-6, <http://dx.doi.org/10.1016/j.wneu.2019.03.249> [Epub 01.04.19; PMID: 30947013].
- Suiter DM, Leder SB. Clinical utility of the 3-ounce water swallow test. *Dysphagia*. 2008;23:244-50, <http://dx.doi.org/10.1007/s00455-007-9127-y> [Epub 04.12.07; PMID: 18058175].
- Perry L. Screening swallowing function of patients with acute stroke. Part one: Identification, implementation and initial evaluation of a screening tool for use by nurses. *J Clin Nurs*. 2001;10:463-73, <http://dx.doi.org/10.1046/j.1365-2702.2001.00501.x> [PMID: 11822494].
- DePippo KL, Holas MA, Reding MJ. Validation of the 3-oz water swallow test for aspiration following stroke. *Arch Neurol*. 1992;49:1259-61, <http://dx.doi.org/10.1001/archneur.1992.00530360057018> [PMID: 1449405].
- Reber E, Friedli N, Vasiloglou MF, Schuetz P, Stanga Z. Management of refeeding syndrome in medical inpatients. *J Clin Med*. 2019;8:2202, <http://dx.doi.org/10.3390/jcm8122202> [PMID: 31847205; PMCID: PMC6947262].
- Zou X, Ouyang B, Yang H, Wang B, Ge S, Chen Y, et al. Surgical treatment for basilar invagination with irreducible atlantoaxial dislocation: transoral atlantoaxial reduction plate fixation vs occipitocervical fixation. *BMC Musculoskelet Disord*. 2020;21:825, <http://dx.doi.org/10.1186/s12891-020-03838-6> [PMID: 33292209; PMCID: PMC7724810].
- Chaudhry NS, Ozpinar A, Bi WL, Chavakula V, Chi JH, Dunn IF. Basilar invagination: case report and literature review. *World Neurosurg*. 2015;83, <http://dx.doi.org/10.1016/j.wneu.2015.02.007>, 1180.e7-e11 [Epub 18.02.15; PMID: 25701769].
- Sasun AR, Qureshi MI. A pragmatic neuro-rehabilitation approach for basilar invagination: a case report. *Cureus*. 2023;15:e41921, <http://dx.doi.org/10.7759/cureus.41921> [PMID: 37583726; PMCID: PMC10424607].
- Cederholm T, Jensen GL, Correia MITD, Gonzalez MC, Fukushima R, Higashiguchi T, et al. GLIM criteria for the diagnosis of malnutrition - a consensus report from the global clinical nutrition community. *Clin Nutr*. 2019;38:1-9, <http://dx.doi.org/10.1016/j.clnu.2018.08.002> [Epub 03.09.18; PMID: 30181091].
- da Silva JSV, Seres DS, Sabino K, Adams SC, Berdahl GJ, Citty SW, et al. ASPEN consensus recommendations for refeeding syndrome. *Nutr Clin Pract*. 2020;35:178-95, <http://dx.doi.org/10.1002/ncp.10474> [Epub 02.03.20; erratum in: *Nutr Clin Pract* 2020 June;35(3):584-585; PMID: 32115791].
- Wunderle C, Gomes F, Schuetz P, Stumpf F, Austin P, Ballesteros-Pomar MD, et al. ESPEN guideline on nutritional support for polymorbid medical inpatients. *Clin Nutr*. 2023;42:1545-68, <http://dx.doi.org/10.1016/j.clnu.2023.06.023> [Epub 08.07.23; PMID: 37478809].
- Baijens LW, Clavé P, Cras P, Ekberg O, Forster A, Kolb GF, et al. European Society for Swallowing Disorders - European Union Geriatric Medicine Society white paper: oropharyngeal dysphagia as a geriatric syndrome. *Clin Interv Aging*. 2016;11:1403-28, <http://dx.doi.org/10.2147/CIA.S107750> [PMID: 27785002; PMCID: PMC5063605].
- Peyer R, Cordier R, Farneti D, Nascimento W, Pilz W, Verin E, et al. White paper by the European Society for Swallowing Disorders: screening and non-instrumental assessment for dysphagia in adults. *Dysphagia*. 2022;37:333-49, <http://dx.doi.org/10.1007/s00455-021-10283-7> [Epub 31.03.21; PMID: 33787994; PMCID: PMC8009935].
- Kanis JA, Cooper C, Rizzoli R, Reginster JY, Scientific Advisory Board of the European Society for Clinical and

Economic Aspects of Osteoporosis (ESCEO), Committees of Scientific Advisors and National Societies of the International Osteoporosis Foundation (IOF). European guidance for the diagnosis and management of osteoporosis in postmenopausal women. *Osteoporos Int.* 2019;30:3–44,

<http://dx.doi.org/10.1007/s00198-018-4704-5> [Epub 15.10.18; erratum in: *Osteoporos Int* 2020 January;31(1):209; erratum in: *Osteoporos Int* 2020 April;31(4):801; PMID: 30324412; PMCID: PMC7026233].