Skull base and craniocervical bone pneumatisation: two case reports of differing presentations and a review of the literature

Olivia Francies[1*], Simon Morley[1], Tim Beale[1]

1. *Department of Radiology, University College Hospital, London, UK*

* **Correspondence:** Olivia Francies, Department of Radiology, University College Hospital, 235 Euston Road, London NW1 2BU (*ofrancies@doctors.org.uk*)

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ABSTRACT

We report two patients with increased central skull base and craniocervical junction bone pneumatisation complicated by extra-osseous gas. One patient presented with symptoms of increasing nasal blockage and 'sinus pressure' on a background of extensive nasal polyposis. He was subsequently found to have a history of repeated Valsalva's manoeuvre, the cessation of which resulted in a rapid decrease in the amount of extra-osseous gas on imaging. The second patient presented following a minor head trauma with dysarthria from a hypoglossal nerve palsy and neck pain, with extensive intra- and extra-cranial gas including within the spinal canal (pneumorrhachis). These radiological findings have been reported previously in patients with Eustachian tube dysfunction and/or activities leading to frequently raised middle ear pressures. We review the possible aetiologies, reported risk factors, and the range of associated imaging abnormalities that may be encountered with this rare appearance.

CASE REPORT

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Presentation

A 53-year old man with a long history of nasal congestion due to extensive polyposis with co-existent fungal disease presented acutely with worsening of his sinus pressure symptoms and nasal blockage. There was no history of trauma or recent surgery. He had previously undergone multiple functional endoscopic sinus surgeries (FESS), including polypectomies.

Imaging

A non-contrast computed tomography (CT) scan of the sinuses demonstrated marked pneumatisation of the mastoid processes and petrous apices of the temporal bones as well as complete pneumatisation of the clivus and occipital condyles. Gas extended beyond the pneumatised bones into the cavernous regions, the atlanto-occipital joint and the soft tissues underlying the skull base (Fig 1). The paranasal sinuses were almost completely opacified and all sinus drainage pathways were obstructed. These appearances had prompted the patient to be referred to our institution with a suspected aggressive sinus infection. The skull base pneumatisation was also demonstrated on the patient's subsequent magnetic resonance imaging (MRI) (Fig 2). Interestingly a CT scan performed 2 years previously, to assess the extent of nasal polyposis prior to FESS, demonstrated a normally ossified non-pneumatised clivus (Fig 3).

Management and follow-up

The possibility of repeated Valsalva's manoeuvre (VM) as the underlying aetiology was raised in view of the persistently obstructed nasal cavity demonstrated on previous sinus CT scans. It was confirmed on clinical questioning that the patient performed this multiple times daily to relieve his symptoms. He was counselled on stopping this habitual behaviour and a repeat CT scan 2 months later demonstrated regression in the amount of gas within the soft tissues around the clivus, temporal bone and cavernous regions (Fig 4). The follow-up scan also showed improved patency of the nasal cavity.

CASE REPORT 2

Presentation

A 71-year old man presented acutely following a minor head injury with left-sided neck pain and fluctuating dysarthria. There was no past medical, social or family history of note. On physical examination he had left-sided tongue deviation with ipsilateral tongue weakness.

Imaging

A CT scan of the head and cervical spine did not reveal any traumatic intracranial injury but did demonstrate extensive skull base and craniocervical junction bone pneumatisation, including the squamous portions of both temporal bones, the clivus, occipital bone and atlas of the cervical spine. The middle ear clefts and petromastoid air cells were clear but there was opacification of the abnormal air cells on the left side within the occipital condyle and atlas with presumed micro-fractures at these sites resulting in extensive intra- and extra-cranial gas (Fig 5). Extradural gas was present within the cavernous regions bilaterally, the left hypoglossal nerve canal, the epidural space of the cervical spine, including within the neural exit foramen, and there was extensive subcutaneous emphysema in the extra-cranial soft tissues of the neck (Figs 5 and 6). Of note the paranasal sinuses were clear. Subsequent MRI demonstrated gas and haemorrhage layering within these osseous cavities but no acute brain or cervical spinal cord injury (Fig 7). The left hypoglossal nerve palsy was concluded to be secondary to foraminal gas causing nerve entrapment.

Management and follow-up

Following Neurology and Neuro-surgical reviews, he was fitted with an immobilising cervical spine collar in view of the ‘paper-thin’ cortices, with likely micro-fractures, of the pneumatised craniocervical region and the potential risk of further neurological decline. No potentially reversible risk factors for the development of skull base hyperpneumatisation were identified on clinical review. A CT scan performed several months’ later revealed resolution of most of the extra-osseous gas (Fig 8). Although his tongue regained some compensatory function, with the aid of specialised speech and language therapy, there was progressive atrophy of the left side resulting in difficulties talking and chewing on clinical follow-up 8 months later.

DISCUSSION

The development of normal temporal bone pneumatisation is complex but can be divided broadly into 3 stages: the infantile stage (birth - 2 years) when air cells begin to appear; the transitional stage (from 2 – 5 years) when the squamo-mastoid portions gradually enlarge and air cells migrate to the periphery; and thereafter into adulthood when pneumatisation ceases [1]. The primary regions of pneumatisation are the mastoid and petrous portions however accessory regions of pneumatisation are well recognised to occur within the squamous and styloid portions [1,2]. Accessory areas of pneumatisation have been shown to be related to higher degrees of temporal bone pneumatisation elsewhere, although it is more usual to observe an under-pneumatised pattern of air cells [1,3]. The ‘air reservoir’ function of the temporal bone air cells is theorised to prevent severe negative pressure from developing within the middle ear when there is Eustachian tube dysfunction (ETD) [1].

Cases of abnormal extension of pneumatisation from the temporal bone into the surrounding osseous structures of the skull base and cervical spine is rarely reported in the published literature and therefore incompletely understood. As such there are no available figures on annual incidence or patient demographics, however the potential aetiology, risk factors, imaging findings and management of this unusual entity are reviewed and summarised based on case reports and small case series that exist in the current literature.

Etiology & Demographics:

There are several proposed hypotheses for the development of skull base and craniocervical junction hyperpneumatisation. Initially, the observation of atlanto-occipital assimilation, a well-recognised anatomical variant, was proposed to explain extension of air cells into the craniocervical junction [4-6], however this is an inconsistent finding, including in our own cases. Pneumatisation is noted to cross cranial sutures in all of the available case reports, the postulated mechanism for which has been described as the theory of ‘expanding communicating spaces’ [6] or the ‘pneumomechanic theory’ [7]; high middle ear pressures (middle ear hypertension) force air out of the temporal bone and into adjacent bones. This is thought to occur in patients with histories of ETD who repeatedly perform Valsalva’s manoeuvre (VM), whereby a possible ‘ball-valve’ mechanism forces air into the middle ear cavity faster than it can escape [8-10]. Prolonged elevation of intra-osseous pressure could therefore lead to thinning of cortical bone and subsequent fracturing, either spontaneously or related to minor trauma, leading to soft tissue emphysema.

ETD occurs most commonly from inadequate tube opening, due to obstructive causes on the tubal lumen or from muscular failure, or much less commonly from patulous ETD, which occurs when the lumen remains permanently open [11]. VM is performed by exhaling against a closed mouth and pinched nose, thus forcing the Eustachian tube to open and equalising pressures between the middle ear and atmosphere (barometric pressure). This process occurs briefly and involuntarily during swallowing, coughing and sneezing but repeated prolonged VMs have been described in patients with cranial hyperpneumatisation who are frequent air travellers [4,8], those with chronic cough [12], patients who play wind or brass instruments [10], and those with hobbies such as weightlifting and deep sea diving [13,14]. There are however case reports of patients with skull base hyperpneumatisation who had no identifiable relevant habits or hobbies (as in our Case 2 described above) [5,8].

More recently, it has been suggested that these theories could be explained in greater detail by middle ear hypertension causing bone ischemia, leading to micro-fractures within the temporal bone and subsequent bone loss and pneumatisation [15,16].

Transmission of high pressure/gas through the venous network has been proposed to account for the pneumatisation observed within the cervical vertebrae [14]. This process may be facilitated by accessory regions of pneumatisation within the temporal bone, incomplete fusion of the basisphenoid synchondrosis, and atlanto-occipital assimilation.

Of the reported cases available in the literature, the youngest to be affected was a 17-year old male and the oldest was the 71-year old man presented in our second case [10].

Clinical & Imaging findings:

Most patients are imaged initially with CT, either following trauma or performed in the investigation of headache/neck pain or neurological symptoms such as tinnitus and vertigo, or upper limb sensory or motor disturbances. Cranial nerve palsies at presentation are rare but have been described in some cases, the commonest being those exiting the foramen magnum via the jugular foramen or hypoglossal nerve canal (i.e. CNIX – XII) [5,17-19].

Extensive pneumatisation of the temporal bone, including involvement of the petrous apices, is a normal variant. Pneumatisation extending into the occipital bone and clivus differentiates the physiological variant from the pathological entity, with the more extensive cases of pneumatisation more likely to result in symptoms. Most cases, including ours, also describe extension of gas across the atlanto-occipital joint with varying degrees of involvement of the upper cervical spine [4-6,8,9,13-16,20]. The extent of gas within the epidural space varies. More commonly it is seen within the foramen magnum [4,8], but it has also been described within the posterior cranial fossa where accumulation of gas has in several cases led to collections causing mass effect on the adjacent brain parenchyma [9,10]. The presence of gas within the spinal canal (pneumorrhachis) is a rare phenomenon with only one other case associated with skull base pneumatisation identified in the literature; this resulted in moderate thecal compression [14].

Only one other case documenting the appearances of skull base pneumatisation on MRI was identified [6]. Our cases demonstrate loss of normal bone marrow signal within the skull base, replaced with expected low T1 and T2-weighted (w) signal reflecting pneumatisation. In the setting of trauma we also demonstrate the presence of acute haemorrhage within the osseous cavitation of the atlas and within the extra-cranial gas collection.

Treatment & Prognosis:

In the setting of habitual VM the simple management strategy of cessation of this behaviour correlates with prompt symptom improvement as well as a reduction in soft tissue emphysema and extradural gas on imaging follow-up [4,8,9,15,16]. In case 1 the pneumatisation developed in less than 2 years, corresponding with nasal obstruction due to extensive nasal polyposis, but settled within several months following cessation of VM. Other cases have been managed with tympanic drainage via ventilation tubes [6,10,20] and immobilisation with external fixation in a halo jacket [5,8]. The decision to manage case 2 with a cervical collar was based on the presenting cranial nerve palsy and concerns regarding craniocervical instability from presumed micro-fractures.

All patients with extensive skull base pneumatisation should be counselled on avoiding hobbies and habits that result in repeatedly raised middle ear pressures, including VMs, as well as activities that are associated with an increased fracture risk, such as contact sports. Even minor head or neck trauma in these patients can result in fractures of the craniocervical region with potentially serious consequences [19,21].

Other cases followed up radiologically demonstrate that the air-spaces are replaced by fluid [9,13,14,16], with one case also showing new bony trabeculae [12], but the exact consistency and mechanism of this is unknown.

Differential Diagnoses:

There are no known pathological processes that mimic the described imaging findings of skull base and craniocervical junction bone hyperpneumatisation. This phenomenon is an important observation that may only be detected on imaging, but is unlikely to cause symptoms unless there are secondary fractures. Knowledge of the normal pattern and variants of temporal bone pneumatisation is crucial in order to differentiate from the abnormal extension of air cells into the occipital bone and cervical vertebrae.

There are varied causes of pneumocephalus and subcutaneous emphysema around the skull base but these do not lead to intra-osseous gas. Traumatic temporal bone fractures frequently lead to both subcutaneous emphysema and pneumocephalus. Subcutaneous emphysema around the skull base may result from necrotising fasciitis, which can occur secondary to dental, tonsillar or salivary infection, or following trauma to the face and neck [22]. These causes should be suspected from the clinical history and examination and should be evident on CT e.g. dental caries, sialoadenitis, and MRI e.g. high T2w oedema, abscess. The retrograde IV injection of air can lead to idiopathic pneumocephalus, which should be suspected when air follows the cranial venous anatomy e.g. within the cavernous sinuses, and other causes of pneumocephalus have been excluded [23]. Pneumorrhachis is usually secondary to migration of gas from adjacent spaces as a result of trauma e.g. pneumocephalus, pneumothorax, subcutaneous emphysema [14].

Conclusion:

We present two rare cases of extensive skull base pneumatisation and its complications, including pneumorrhachis and a hypoglossal nerve palsy, which resulted in a long term neurological deficit. From our own experience and on reviewing the available literature this phenomenon is associated with repetitive positive pressure activities, such as Valsalva's manoeuvre in patients with longstanding Eustachian tube dysfunction. The simple conservative management strategy of cessation of any pre-disposing habits or hobbies can result in a prompt improvement in symptoms as well as the acute radiological findings. The serious risk of fracture in this sensitive area, even following minor trauma, should be highlighted to these patients, particularly if they partake in activities that may be associated with an increased risk.

TEACHING POINT

Skull base and craniocervical bone pneumatisation is a rare phenomenon, best demonstrated on CT imaging, and when identified should prompt investigation into possible causative factors, such as repetitive Valsalva's manoeuvre. The potential neurological complications, which include cranial nerve palsies, must be highlighted to the referring clinician as well as the patient so that management options and lifestyle changes can be considered.

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FIGURES

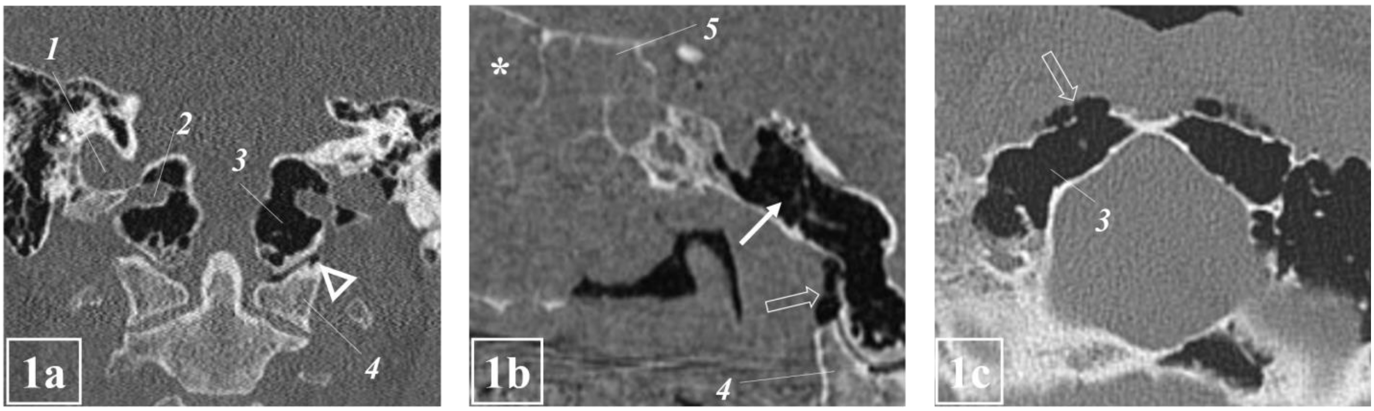

Figure 1: Case 1 - 53-year old male with skull base pneumatisation secondary to repetitive Valsalva's manoeuvre.
FINDINGS: CT images in the coronal (a), reformatted sagittal (b), and axial (c) planes demonstrating extensive pneumatisation of the petromastoid temporal bones, clivus (arrow), the posterior occipital bone as well as both occipital condyles. Gas extends into the extra-osseous pre-clival space (open arrows) and into the atlanto-occipital joint (open arrowhead). Note the complete opacification of the visualised sphenoid sinus and ethmoid air cells (asterisk) secondary to extensive sinonasal polyposis. Anatomical landmarks: 1. Jugular foramen. 2. Hypoglossal nerve canal. 3. Occipital condyle. 4. Lateral mass of C1. 5. Sphenoid sinus.
TECHNIQUE: Non-contrast CT (Siemen's Definition AS+ 128-slice scanner), 625mAs, 125kVp, 1mm slice thickness, DLP 158.42mGycm.

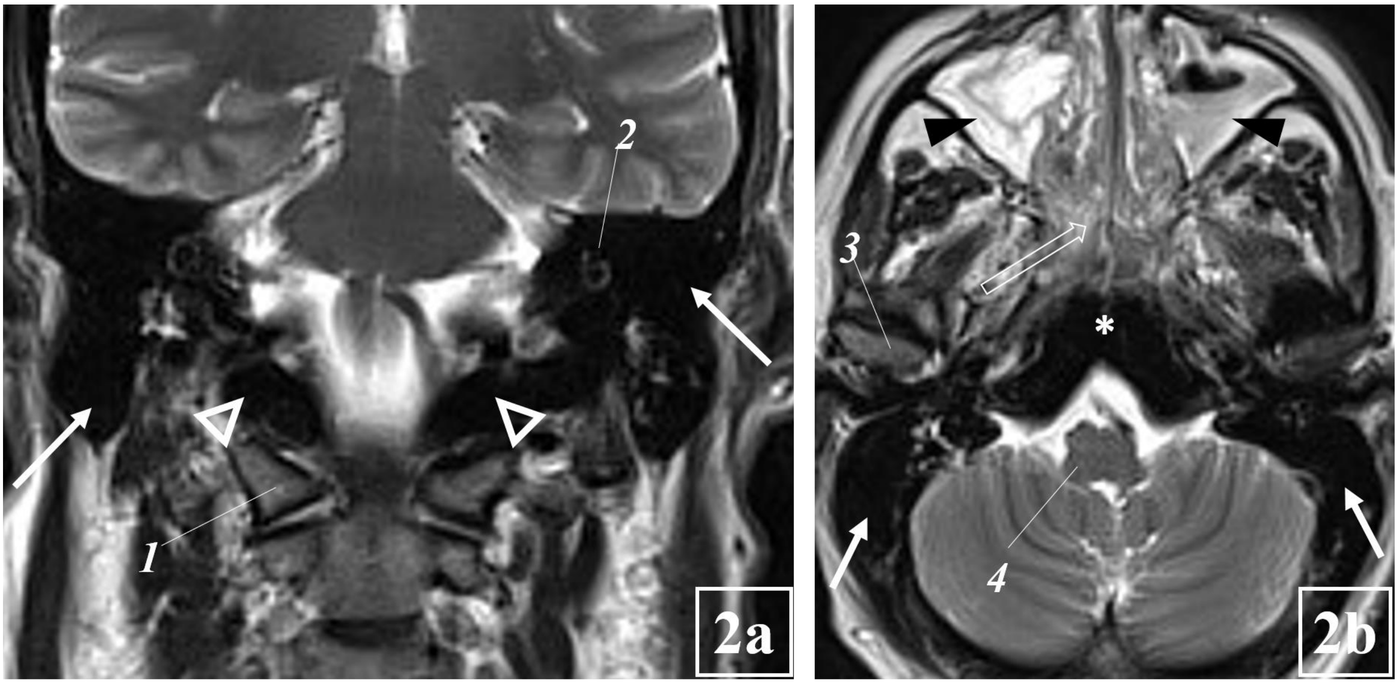

Figure 2: Case 1 - 53-year old male with skull base pneumatisation secondary to repetitive Valsalva's manoeuvre.
FINDINGS: Coronal (a) and axial (b) MR T2-weighted (w) images demonstrating loss of normal bone marrow signal within the skull base, secondary to extensive pneumatisation, including within the temporal bones (solid arrows), occipital condyles (open arrowheads) and the clivus (asterisk). Extensive nasal polyposis (open arrow) and maxillary sinus mucosal inflammation with obstructed secretions (black arrowheads) are also seen. Anatomical landmarks: 1. Lateral mass of C1. 2. Membranous labyrinth. 3. Mandibular condylar head. 4. Medulla oblongata.
TECHNIQUE: Siemens Magnetom Avanto 1.5 Tesla. T2 BLADE: TE 107ms, TR 8520ms, 3mm slice thickness, non-contrast.

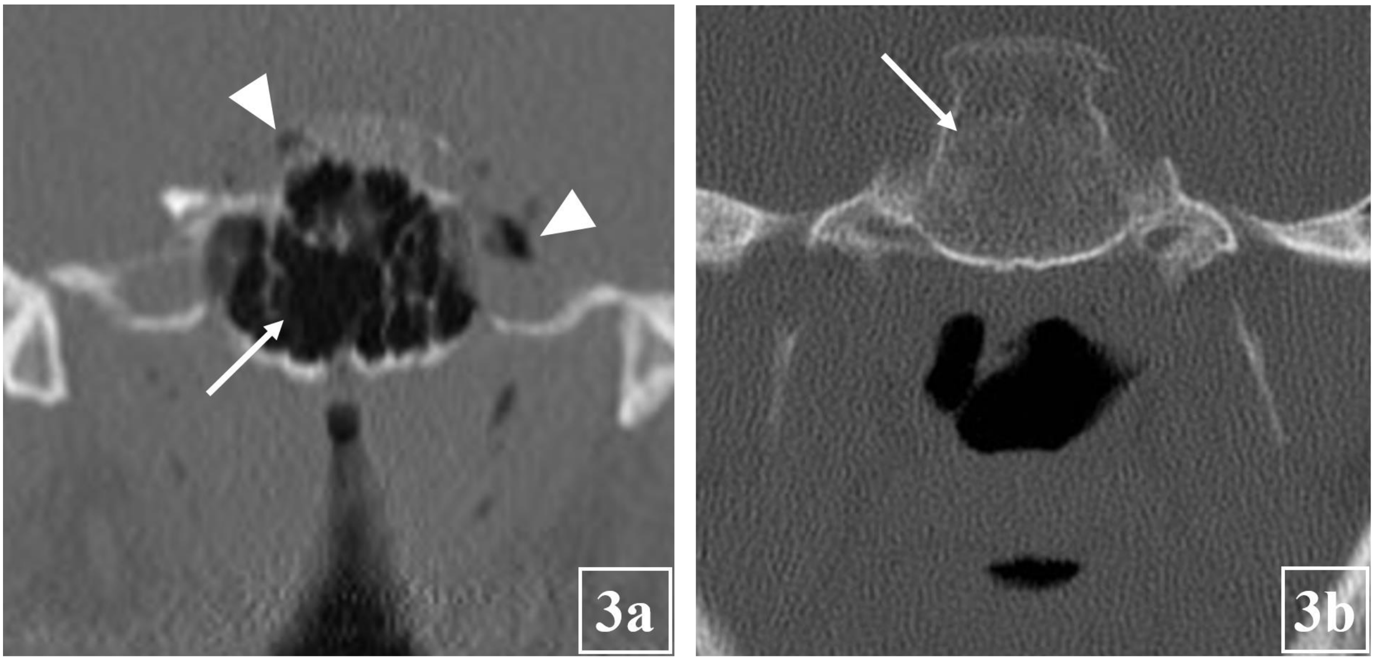

Figure 3: Case 1 - 53-year old male with skull base pneumatisation secondary to repetitive Valsalva's manoeuvre.
FINDINGS: Coronal CT images comparing the clival appearances at presentation (a) and 2 years previously (b). The clivus has completely pneumatised (arrows) in the interim. Extradural gas within the adjacent cavernous regions (arrowheads) on the presentation CT scan is most likely secondary to micro-fractures within the adjacent pneumatised clivus as the patient had not had a preceding IV injection and there was no other identifiable cause for pneumocephalus.
TECHNIQUE: (a) non-contrast axial CT with coronal reformat (Siemen's Definition AS+ 128-slice scanner), 625mAs, 120kVp, 1mm slice thickness, DLP 158.42mGycm; (b) non-contrast coronal CT (GE 32-slice HiSpeed scanner), 150mAs 120kVp, 5mm slice thickness. Estimated effective (whole body) dose 0.49mSv.

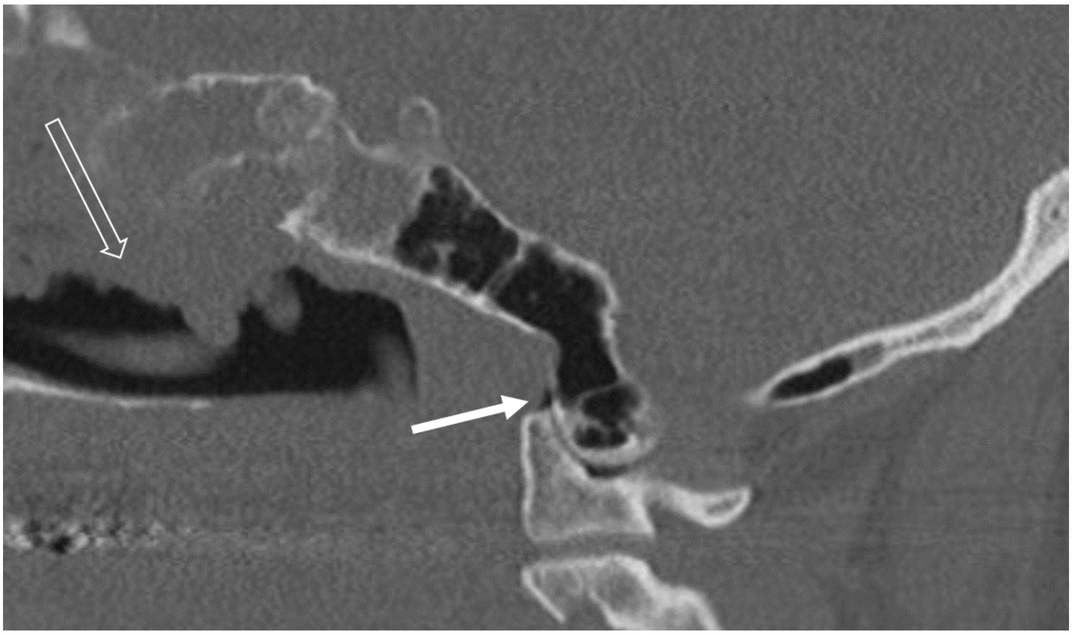

Figure 4: Case 1 - 53-year old male with skull base pneumatisation secondary to repetitive Valsalva's manoeuvre.
FINDINGS: Sagittal CT image demonstrating regression of extra-osseous gas in the pre-clival space 2 months after presentation (arrow). Note the persistent skull base pneumatisation and thin cortices of the clivus and occipital condyle. Polypoidal soft tissue is still present within the nasal cavity (open arrow) however the patency of the airway has improved.
TECHNIQUE: Non-contrast CT (Siemen's SOMATOM Definition AS 128-slice), 292mAs, 100kVp, 1mm slice thickness, DLP 63mGy-cm.

Figure 5: Case 2 - 71-year old man with craniocervical bone pneumatisation and complications following minor trauma.
FINDINGS: Left para-midline sagittal (a), mid-line sagittal (b) and axial (c) CT images demonstrating pneumatisation of the left occipital condyle (white arrowhead), the postero-inferior occipital bone (white arrow), and the left lateral mass and posterior arch of the atlas (open white arrows). There is generalised thinning of the cortices in these regions and partial opacification of the abnormal occipital condyle and atlas air cells (asterisks). The presumed post-traumatic micro-fractures in these areas has resulted in extensive surrounding extra-osseous gas, including within the epidural space of the foramen magnum (black arrow), and within the cervical spinal canal and neural exit foramen (open black arrows). Anatomical landmarks: 1. Exiting neural foramen for C3 nerves. 2. Anterior arch of C1. 3. Basion. 4. Opisthion. 5. Odontoid peg of C2.
TECHNIQUE: Non-contrast axial CT with sagittal reconstructions (Siemens 64 slice scanner), 4754mAs, 120kVp, 2mm sagittal slice thickness, 0.75mm axial slice thickness, DLP 1303mGy-cm.

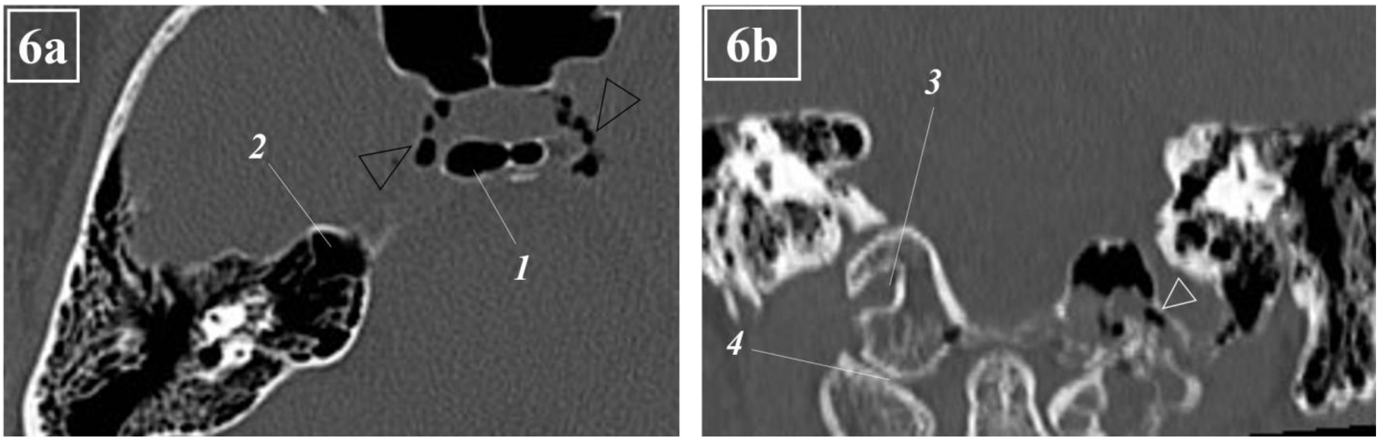

Figure 6: Case 2 - 71-year old man with craniocervical bone pneumatisation and complications following minor trauma.
FINDINGS: Axial (a) and reformatted coronal (b) CT images demonstrating gas within both cavernous regions (open black arrowheads) and within the left hypoglossal nerve canal (open white arrowhead). The left hypoglossal nerve palsy was attributed to the presence of gas within the canal causing nerve entrapment. Anatomical landmarks: 1. Pneumatised dorsum sella (post-sellar pattern of pneumatisation around the sphenoid sinus). 2. Pneumatised petrous apex of the temporal bone. 3. Hypoglossal nerve canal. 4. Atlanto-occipital joint.
TECHNIQUE: Non-contrast axial CT with coronal reformats (Siemens 64-slice scanner), 4754mAs, 120kV, 0.75mm slice thickness, DLP 1303mGy-cm.

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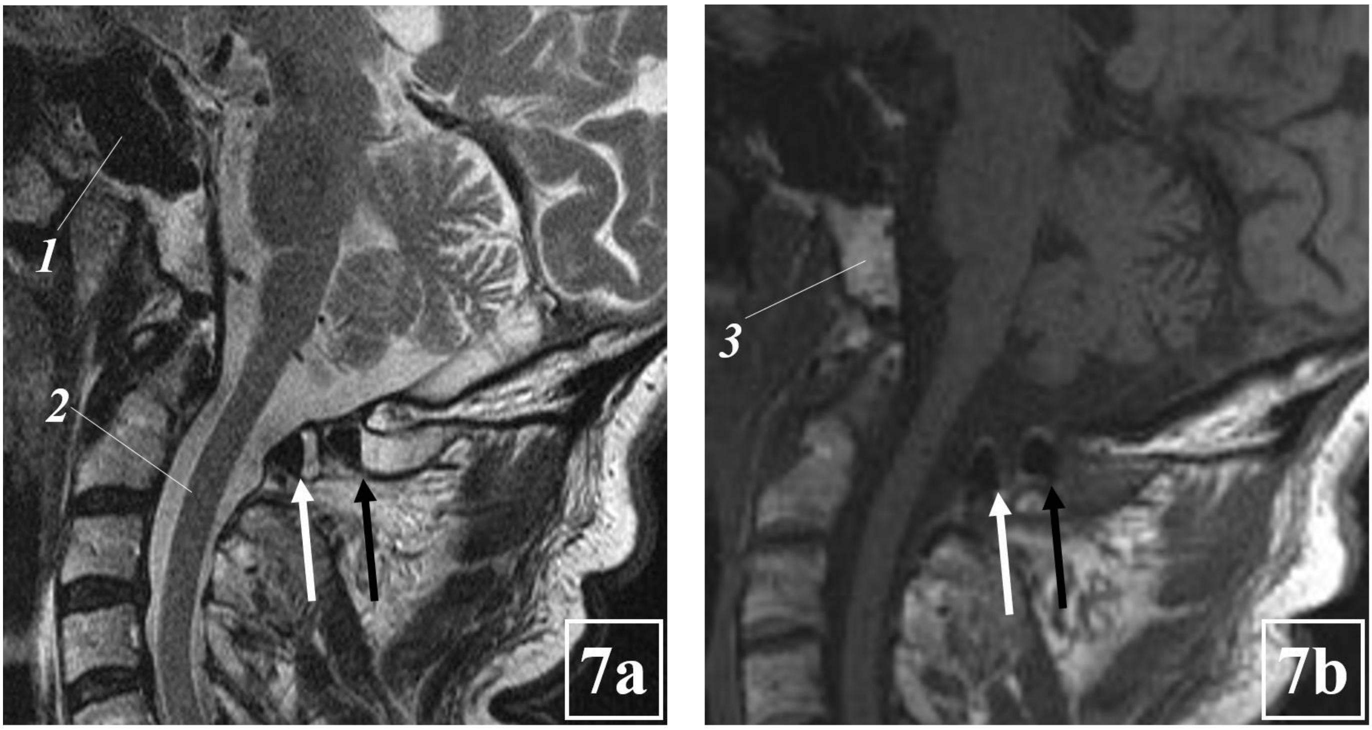

Figure 7: Case 2 - 71-year old man with craniocervical bone pneumatisation and complications following minor trauma.
FINDINGS: Sagittal MR T2 (a) and T1-w (b) images demonstrating air-fluid levels within the pneumatised spinous process of C1 (white arrows) as well as within an encapsulated pocket of gas just posterior to this and inferior to the opisthion of the occipital bone (black arrows). The signal intensity of the fluid is consistent with acute haemorrhage (hyperintense on T2w and isointense on T1w). The MRI confirms the normal appearance of the spinal cord with no extradural haematoma. Anatomical landmarks: 1. Aerated sphenoid sinus 2. Normal T2w signal within the cervical spinal cord. 3. Normal bone marrow signal within the clivus.
TECHNIQUE: Siemens Magnetom Symphony 1.5 Tesla, (a) T2 Turbo Spin Echo: TR 3780, TE 108. (b) T1 Spin Echo (SE): TR 404, TE 14. Non-contrast.

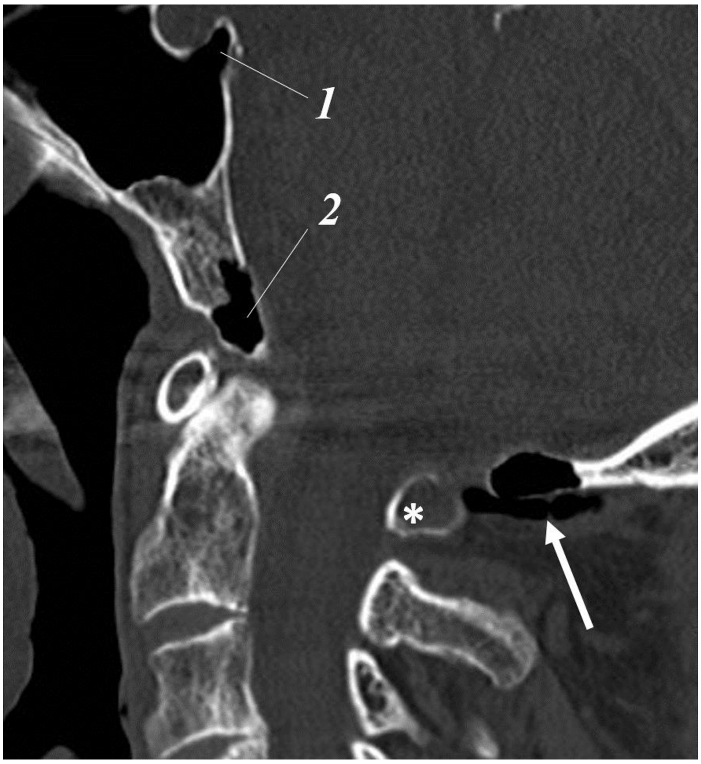

Figure 8 (left): Case 2 - 71-year old man with craniocervical bone pneumatisation and complications following minor trauma.
FINDINGS: Sagittal CT performed 5 months later demonstrates that most of the abnormal intra- and extra-cranial gas has resolved with only a small pocket of gas inferior to the opisthion (arrow) remaining. The abnormal skull base and craniocervical pneumatisation persists and some of the air spaces are filled with fluid (asterisk). No conspicuous callus formation was present to assist in localization of a focal fracture site. Anatomical landmarks: 1. Pneumatised dorsum sella. 2. Partly pneumatised clivus.
TECHNIQUE: Non-contrast CT (Siemens 64-slice scanner), 275mAs, 120 kV, 2mm slice thickness, DLP 189mGy-cm.

Etiology (proposed hypotheses)	• Repeated Valsalva's manoeuvres (VMs) in patients with Eustachian tube dysfunction (ETD) creates a 'ball-valve' mechanism leading to middle ear hypertension ○ This forces air out of the temporal bone, across sutures, and into adjacent bones • Middle ear hypertension causes bone ischemia, leading to micro-fractures and subsequent bone loss • Gas transmitted through the craniocervical venous network accounts for cervical vertebrae pneumatisation • Atlanto-occipital assimilation may play a role in some cases
Incidence; gender ratio; ethnicity; age predilection	• Unknown due to the rarity of the entity, which is currently limited to case reports and small case series • Only documented to occur in adults
Risk factors	Patients who repeatedly perform VMs, including for the following reasons: • ETD • Chronic cough • Frequent air travel • Wind and brass instrument players • Free-divers and scuba-divers • Weightlifters
Treatment	• Cessation of VMs • Tympanic drainage with ventilation tube • Cervical spine immobilisation should be considered if there are concerns about craniocervical junction/cervical spine instability • Counselling on avoidance of activities associated with a higher cranial/cervical fracture risk e.g. contact sports
Prognosis	• Regression of hyperpneumatisation is observed if habitual VMs are stopped
Computed tomography (CT) imaging	• First-line • Extensive temporal bone pneumatisation <u>with</u> extension of pneumatisation into the occipital bone, clivus and upper cervical spine • Spontaneous micro-fractures or post-traumatic fractures of the affected bones can lead to extensive intra and extra-cranial gas ○ Rarely this extends into the spinal canal (pneumorrhachis) • Comment on features of rhinosinusitis, nasal polyposis and middle ear opacification • Performed in follow-up/monitoring
Magnetic resonance (MR) imaging	• Not necessary for diagnosis; recommended to investigate any potential post-traumatic complications or complications secondary to rhinosinusitis • Normal T1 and T2 hyperintense bone marrow of the skull base is replaced by low signal pneumatisation (low signal on all sequences) • Intra-osseous haemorrhage may be observed following trauma

Table 1: Summary table for extensive skull base and craniocervical bone pneumatization.

Differential Diagnoses	Computed tomography (CT) findings	Magnetic resonance (MR) findings
Hyperpneumatisation of the skull base and craniocervical junction	Extensive pneumatisation of the temporal bones *and* pneumatisation extending into the occipital bone and clivus. The upper cervical spine is involved in most cases. Gas may be seen within the foramen magnum and intra-cranially, and rarely gas may also extend into the cervical spine.	Normal bone marrow signal within the skull base is replaced with low T1 and T2-weighted (w) signal reflecting pneumatisation.
Trauma	Visible skull and/or skull base fracture with pneumocephalus/pneumorrhachis/ subcutaneous gas/pneumothorax. Physiological pattern of temporal bone pneumatisation.	Varying T1/T2w signal intensities of fluid i.e. cerebrospinal fluid (CSF) or haemorrhage within the temporal bone. Intra-cerebral and spinal cord complications of trauma.
Necrotising fasciitis	Subcutaneous gas within the head and neck soft tissues with evidence of the underlying source of infection e.g. dental caries, tonsillar or salivary gland abscess, peri-orbital cellulitis. Physiological pattern of temporal bone pneumatisation.	High T2w signal oedema within soft tissues. Heterogeneous T2w signal and restricted diffusion within an abscess cavity.
Retrograde intravenous (IV) injection of air	Gas locules follow extra and intra-cranial venous anatomy. No other cause of pneumocephalus identified. Physiological pattern of temporal bone pneumatisation.	No specific features.

Table 2: Differential diagnosis table for hyperpneumatisation of the skull base and craniocervical junction.

ABBREVIATIONS

CT = Computed Tomography
ETD = Eustachian Tube Dysfunction
FESS = Functional Endoscopic Sinus Surgery
MRI = Magnetic Resonance Imaging
VM = Valsalva's manoeuvre

KEYWORDS

skull base pneumatization; craniocervical pneumatization; hyperpneumatization; skull base; atlanto-occipital joint; computed tomography; magnetic resonance imaging; Valsalva's manoeuvre; pneumorrhachis; hypoglossal nerve palsy; Eustachian tube dysfunction