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## Case Report

# An unusual presentation of empty sella syndrome in oral and maxillofacial surgery: A case report

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### ABSTRACT

Oral and maxillofacial surgeons frequently encounter various anatomical abnormalities and incidental discoveries while reviewing routine radiographs. These serendipitous findings can ultimately benefit the patient by enhancing our diagnostic practices and facilitating timely treatment. This report aims to spotlight a noteworthy radiological finding known as "Empty Sella Syndrome" (ESS) and present a case report from our department. ESS involves the compression of the pituitary gland into a thin rim due to increased cerebrospinal fluid pressure, resulting in the appearance of an almost empty sella turcica. This condition impacts both the neurological and endocrine systems in individuals of all age groups, exhibiting a preference for females. A young patient reported with Buccal space infection and drainage of pus extra-orally along with decayed teeth. The treatment planned was Incision & Drainage, extraction of non-restorable teeth under General Anesthesia as it was a challenge to make the patient cooperate under local anesthesia due to her social anxiety and comprehension difficulties. Patients with empty sella syndrome who present with hormonal deficiencies and delay in developmental milestones may experience intellectual disabilities, they require multidisciplinary collaboration, long term treatment and observation. A thorough knowledge of human anatomy and interpretation of radiographs as well as investigations is critical in recognizing such rare and unusual conditions.

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## 1. Introduction

Clinical and radiologic examinations are indispensable in the diagnostic and treatment processes. They play a vital role in identifying various syndromes and conditions, enabling us to provide appropriate care and anticipate potential complications. Recently, we encountered a unique anatomical anomaly in the field of Oral and Maxillofacial surgery while attending to a 13-year-old patient who has had empty sella syndrome since birth. This syndrome poses challenges in examining, diagnosing, and treating the patient effectively.

Empty sella syndrome is a pathological variant of the radiologically described empty sella, distinguished by the intrasellar extension of the subarachnoid space leading to the flattening of the pituitary gland. It has also been referred to as arachnoidocele due to the underlying mechanism. Typically, the diagnosis is by chance, frequently discovered incidentally during brain imaging studies conducted for various unrelated indications. This syndrome can be linked to a range of neurological and endocrinological conditions, with growth hormone deficiency being a common association, often necessitating specific treatments.

The majority of exodontia treatments are conducted under local anesthesia via nerve blocks, alongside detailed

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patient explanations and their active cooperation. However, patients with mental challenges pose distinct management challenges. In such instances, procedures are often carried out under general anesthesia or conscious sedation to optimize outcomes.

The objective of this case report is to enhance our understanding of empty sella syndrome through clinical observations of a child with the syndrome, who presented with social anxiety and comprehension difficulties. The patient exhibited challenges in following even simple instructions, necessitating the decision to perform the procedure under general anesthesia.

## 2. Case Report

A 13-year old female patient was referred from Paediatric department to the Department of Oral and Maxillofacial surgery.

The patient presented with short stature, speech and intellectual disabilities with great sense of social anxiety. (Figure 1) Patient also has delayed developmental milestones such as pubertal signs, which were believed to be deficiency of growth hormone owing to Empty Sella syndrome (ESS) that was diagnosed post MRI scan. (Figure 2) Patient also had a history of epilepsy for which she was on medication which was discontinued 5 years ago following medical advice.



Figure 2: The MRI scan showing empty sella



Figure 1: Profile photo of the patient with swelling seen on the left lower part of face

Patient reported to have swelling on the left side of face since 3 days which was gradually increasing along with pus discharge with history of pain in the left lower back tooth region and inability to chew. (Figure 3) On extraoral examination swelling appeared to be localized, firm in consistency approximately 3x3 cms size, circular in shape and tender on palpation which was febrile and visible punctum with active pus discharge.



Figure 3: Swelling on left side of face with active pus discharge

As she was suffering with ESS, patient was bound to be uncooperative for intraoral examination or any further investigations which may include radiographs or blood investigations.

Tell-show-do method was used to persuade the patient for detailed intraoral examination and grossly decayed teeth were identified as left lower first molar and radiograph was procured. Other decayed teeth were recorded i.e. 55, 74, 75. Based on these findings it was provisionally diagnosed as buccal space infection on the left side of face secondary to carious 36. Treatment planned was incision and drainage and extraction of decayed teeth under general anesthesia.

Patient was admitted in the paediatric ward, preanesthetic clearance taken, and pre-emptive antibiotics, analgesics course started. Surgery was planned under general anesthesia with anti-epileptic drugs for emergency. Incision and drainage of swelling and extraction of carious teeth 55, 74, 75, 34,36 was done with thorough curettage under strict asepsis. Throughout, the procedure was uneventful. Patient was discharged after 1 week. The patient had also been referred to the pediatric department for further treatment of syndromic manifestations.

### 3. Discussion

The term “Empty Sella” refers to an anatomical & radiological condition in which subarachnoid space herniates into the sella pushing the pituitary gland either to the bottom or to one side of the sella.<sup>1</sup> It is partially or completely filled with cerebrospinal fluid (CSF). When this radiographic picture is accompanied by symptoms it is referred to as Empty Sella Syndrome (ESS).<sup>2</sup>

First described by Bush in 1951, Kauffman et al noted the relationship between increased Intracranial Pressure [ICP] & empty sella syndrome are emphasized that a prolonged increase in CSF pressure is a direct cause of ESS.<sup>1</sup>

It is referred to as partial ES when the sella turcica is filled with CSF to a level of 50% of the sella and pituitary gland thickness ranges from 3 to 7 mm, with 7 mm being the lower limit of normal thickness. A diagnosis of complete empty sella is made when more than 50% of sella is filled with CSF and the pituitary gland thickness is less than or equal to 2 mm.<sup>3</sup>

It is classified into Primary & Secondary. Primary empty sella is postulated to be the consequence of congenital deficiency of sellar diaphragm and intermittent pressure changes in CSF resulting in herniation of subarachnoid space into the sella from above with compression of the pituitary tissue.<sup>2</sup> Other possible etiologies are perinatal trauma, ischemia with subsequent interruption of blood supply to the hypophysis and rupture of intrasellar cysts.

Normally, the sella turcica is a small concavity in the sphenoid bone completely filled by the pituitary gland. A sellar diaphragm is attached anteriorly to the tuberculum sellae and posteriorly to the clinoid processes. The only

opening is that through which the pituitary stalk passes. The diaphragm usually prevents cerebral spinal fluid (CSF) from entering the sella. However, in the ESS a defect in the diaphragm occurs, allowing herniation of the CSF-containing cistern into the sella. There is gradual expansion of the sella due to CSF pressure and eventual compression of the pituitary tissue.<sup>2</sup>

Chronically transmitted CSF pulsations from the herniated subarachnoid space lead to bony expansion and remodelling of the sella turcica.<sup>4</sup> The role of increased ICP in the pathogenesis and perpetuation of primary ESS has been confirmed in many studies.<sup>5</sup>

Secondary empty sella is due to previously expansile pituitary tumours, trauma, surgery, radiation therapy or pituitary hypertension<sup>6</sup> it results as a sequela of pituitary injury such as spontaneous necrosis of a adenoma, infective or autoimmune process or a previous surgical, pharmacological or radiologic treatment.<sup>7</sup>

The prevalence of empty sella ranges from 5.5% to 12% in autopsy studies, 12% of patients undergoing neuroimaging and in clinical practice it is reported around 8 to 35%.<sup>6</sup> ESS has been identified in approximately 40% of individuals with hormonal abnormalities and in up to 94% of patients with intracranial hypertension.

In adults it is common and often asymptomatic, in children it is most often associated with endocrine abnormalities, growth hormone (GH) deficiency, hyperprolactinemia, sexual precocity, diabetes insipidus.<sup>8</sup> Since it is a radiologically significant lesion, diagnosis requires CT and MRI scanning to be performed. Although PES is often an incidental finding in asymptomatic patients, a variable degree of neurological, visual, and/or endocrine disorders have been described. The pooled prevalence of hypopituitarism is estimated between 19 and 68%, so that hormone evaluation is always recommended at diagnosis. Multiple axis dysfunctions are more commonly described than isolated pituitary insufficiency. According to current evidence, the somatotrophic axis is the most frequently affected, followed by the gonadotropic axis.

The common symptoms of empty sella can be divided into neurological symptoms such as headache, mental retardation, ophthalmologic abnormalities, tinnitus, vertigo, papilloedema, optic atrophy, blurred vision, diplopia, visual field defects, rhinorrhea. Endocrine abnormalities such as hyperprolactinemia, growth hormone deficiency, short stature, delayed puberty, abnormal growth rate, obesity, precocious puberty, diabetes insipidus, and multiple pituitary hormone deficiencies, pituitary microadenomas causing acromegaly and Cushing's disease may be associated.<sup>2,8</sup> The most prevalent pituitary deficiency was growth hormone deficiency. In the majority of cases, the reason for presenting to the medical services was the short stature according to a study by Jurca MC et al.<sup>9</sup>

In a study by Durodoye et al, 5 patients diagnosed with empty sella syndrome presented with endocrine manifestations. Three of the 5 patients required glucocorticoid replacement, a rare finding in the literature, and 2 of these patients had life-threatening symptoms of glucocorticoid deficiency. This report shows the varied presentations of the hormonal disturbances and highlights the need for prompt diagnosis and appropriate treatment.<sup>10</sup>

Krytsi et al have reported a rare case of a 9 year old with empty sella syndrome associated with Neurofibromatosis type 1, Hypopituitarism and Growth hormone deficiency, short stature, treated with hormone replacement therapy and lifelong endocrine follow-up.<sup>11</sup>

In the present case diagnosis of the syndrome was made incidentally when the patient reported for the dental abscess. Short stature, mental retardation, speech disability and delay of puberty were noted clinically and radiological imaging also confirmed the diagnosis of empty sella syndrome.

Therapy is generally focused on hormone replacement. Replacement hormone therapy in PES syndrome must be assessed for every single hormone and administered according to the appropriate temporal sequence. In the presence of multiple pituitary hormone deficiencies, it is recommended that hormonal replacement treatment starts with hydrocortisone, followed by levotiroxine.<sup>12</sup> However, treatment of the ES itself has been attempted. Repairing the sella or performing chiasmectomy,<sup>1</sup> extradural transsphenoidal packing of the ES with lyophilized dura, fat, or acrylic has been performed, but postoperative complications are common. An extradural filling of the sella with a detachable silicone balloon filled with liquid silicone reversed the headaches and visual symptoms, but all the balloons deflated at longterm follow-up.<sup>10</sup> In cases of ES associated with idiopathic intracranial hypertension, medical treatment of this predisposing condition has led to reversal of the ES in a few reported cases.

There is no clear treatment paradigm for children. The management strategy for children should be tailored to each patient and be based on the cause and pathophysiology of their ESS.

Three guidelines should be considered: 1) early detection; 2) control and reduction of high ICP; and 3) hormone replacement therapy when it is needed.

Accordingly, we advocate treating these patients as soon as possible, by endoscopic third ventriculostomy and fenestration of the suprasellar arachnoid cyst, by shunt placement, or by early tumor removal. It is possible to ameliorate symptoms related to high ICP or to tumor. Early detection and treatment may prevent the development of this phenomenon.

Patients with primary empty sella need to be carefully evaluated at the time of diagnosis, even if PES is incidentally discovered. We suggest completing the assessment with dynamic tests (screening for GHD and

adrenal insufficiency). Hypopituitarism is frequent (40%) but a deterioration in pituitary function seems uncommon (3%).<sup>1</sup>

PESS is often considered an incidental and unimportant finding and has not got much attention by clinicians. But as we are starting to know more about developmental biology of psychiatric disorders, we are finding more and more evidence that findings like ESS may actually indicate some possible underlying developmental deficits. While empty sella requires no treatment, ESS may need pharmacologic or surgical interventions.<sup>13</sup>

#### 4. Conclusion

In our departments, we frequently encounter patients with various physical and mental challenges, and the ability to effectively manage their care is a hallmark of a skilled surgeon. It's crucial to recognize each patient's individual limitations and their level of cooperation, which then informs our surgical planning. Previously, empty sella syndrome was primarily regarded as a radiological finding with no clinical significance. However, recent studies have revealed the far-reaching effects of a shrunken pituitary gland on multiple organ systems and the variety of potential presentations. Therefore, having knowledge of this syndrome positions us one step ahead in terms of diagnosis and therapeutic approaches.

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None.

#### 6. Conflict of Interest

The authors declare that they have no conflict of interest.

#### 7. Informed Consent


Informed consent for publication of clinical details and clinical images was obtained from the legal guardian of the patient.

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