

CASE REPORT

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# Total aplasia of the paranasal sinuses and nasal cavity: the first case report

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## Abstract

**Background:** The paranasal sinuses (PNSs) are small empty spaces around the nasal cavity at the entrance to the respiratory tract, and they have various functions. The development and anatomy of the PNSs are exposed to many variations, often leading to patients' complaints upon presentation.

**Case presentation:** Here, the first case of total aplasia of the paranasal sinuses and nasal cavity and its contents is presented. Several cases of varying degrees of combined aplasia and hypoplasia, even total sinus aplasia, have been reported in the literature, but to the author's knowledge, no cases of total sinus aplasia and associated nasal aplasia have been described thus far.

**Conclusion:** This case report describes a rare, and probably the first, case of severe sinonasal developmental defects elegantly identified with bone window computed tomography (CT).

**Keywords:** Paranasal sinuses, Computed tomography, Aplasia, Nose

## Background

The PNSs are four sets of empty spaces in the facial and cranial bones located at the center point of the face surrounding the nose. The PNSs function to warm inspired air, decrease the weight of the skull, support facial growth/architecture, and help protect the airway against offending pathogens, with many other functions that are still unclear. The nose is the essential organ of smell and the upper respiratory system [1]. PNS development and anatomy are subject to many peculiarities and anatomical variations [2]. Computed tomography (CT) is an excellent method to determine the detailed anatomy of the PNSs [3].

## Case presentation

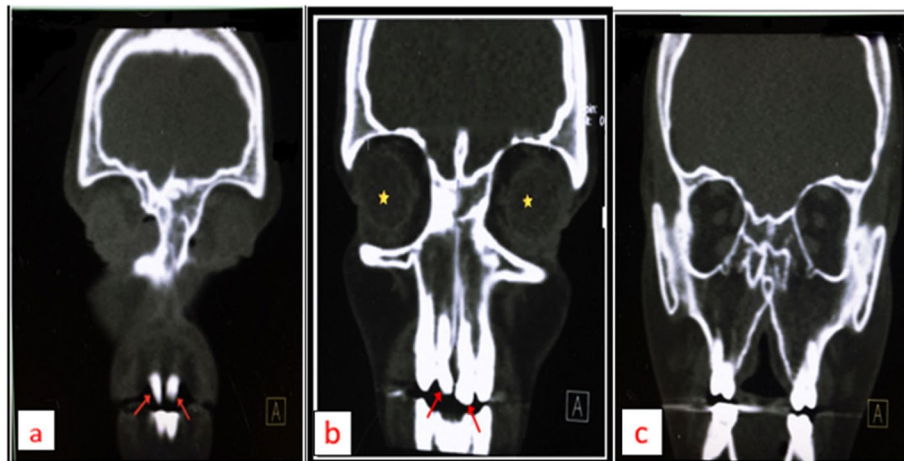
The patient was a young 26-year-old male from a rural area who visited the ENT outpatient clinic of our hospital complaining of nasal obstruction and mouth breathing since birth. The patient was referred to the radiology

department for nonenhanced bone window multidetector computed tomography (CT) for paranasal sinus evaluation. Clinically, the patient was mentally intact. His face was dysmorphic due to very small, rudimentary nonpatent nostrils; a short, flat midface; underdeveloped cheeks; and the absence of a nasal bridge. He denied any previous nasal surgery, trauma, radiotherapy, or granulomatous or chronic sinonasal infections.

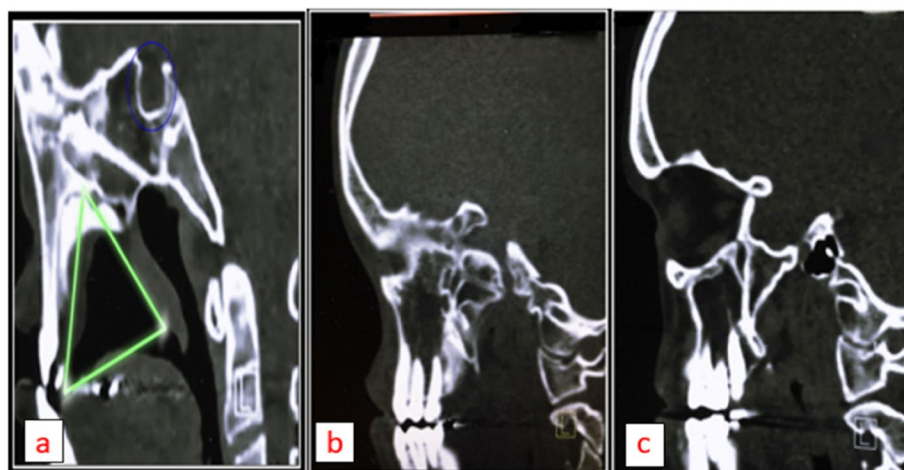
## Imaging findings

Multiphase reconstructed CT images were obtained in soft tissue and bone windows, and in coronal (Fig. 1), sagittal (Fig. 2), and axial (Fig. 3) planes with thin sections (0.6 mm), using the Siemens SOMATON Definition AS 2012B scrollable multitask workstation. These images revealed the following: bilateral, symmetrical, and complete absence of the pneumatization of almost all paranasal sinuses; the absence of a nasal cavity and associated internal structures (nasal bone, turbinates, bony and cartilaginous septum, vomer bone, uncinate process, and nasal process of the maxillary bone); a high riding/convex arched hard palate, resulting in a spacious oral cavity; the absence of nasolacrimal ducts; a thick cribriform plate of the ethmoid bone;

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**Fig. 1 a–c** The coronal sections of the facial CT scan reveal the absence of frontal, ethmoid, and maxillary sinuses; the absence of the nasal bone and cavity; normal maxillary incisors (red arrows) and orbits (yellow stars); and normal bone density



**Fig. 2 a–c** The sagittal sections of the facial CT scan demonstrate a non-pneumatized, compact maxilla; an aplastic sphenoid bone; aplastic frontal sinuses; and a high-riding, convex palatal arch, resulting in increased space in the oral cavity in the craniocaudal dimension (green triangle). The scan also shows a normal pituitary fossa (blue circle) and the absence of a nasal cavity

and bilateral patent but medially displaced infraorbital canal and foramen.

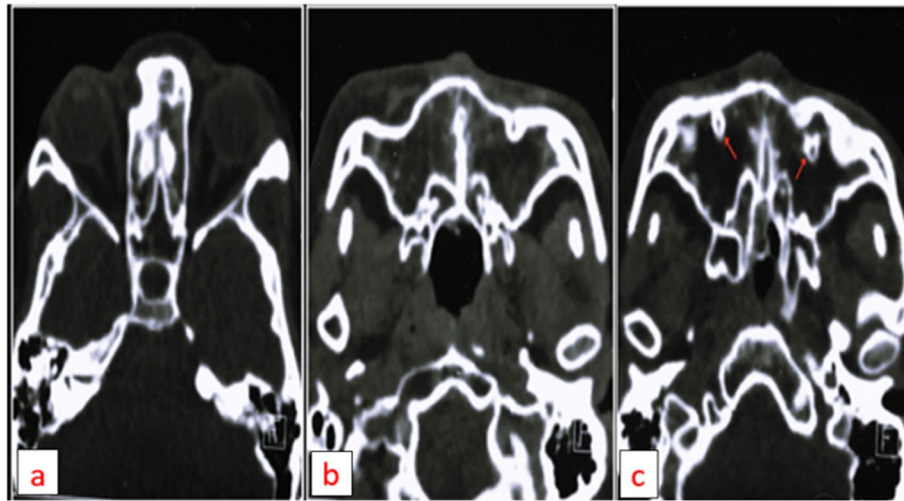
The following structures were normal: both orbit and optic canal/foramen, lamina papyracea, crista Galli, pituitary fossa, other skull base foramina, petrous bone, mandible and temporomandibular joints, pterygoid plates, zygomatic processes, maxillary incisor teeth, brain, and other intracranial structures.

No evidence of prior surgical changes was observed, and the overall bone density was normal.

## Discussion

Understanding the embryological development of the nose and paranasal sinuses (PNSs) is crucial for the interpretation of morphological abnormalities. The development of the PNSs occurs via a series of complex coordinated events that start as early as the 3rd week of gestation and progress thereafter.

By the seventh week of intrauterine life, the partitioning of the oronasal membrane brings about the arrangement of the oral and nasal cavities. The nasal pits migrate



**Fig. 3** a–c The axial sections of the facial CT scan demonstrate a non-canalized nasal cavity, a non-pneumatized maxilla, non-pneumatized ethmoid and sphenoid sinuses, normal orbits, and normal infraorbital nerve canals (red arrows)

anteriorly to the outside by means of the nostrils and posteriorly via the primitive choana to the nasopharynx. Congenital choanal atresia results from the failure of the oronasal membrane to rupture. By the eighth week, the lateral nasal wall and turbinate are highly developed. The cartilaginous nasal capsule gives rise to the uncinate process by 10 weeks of gestation, and by 13–14 weeks of gestation, the primitive maxillary sinuses start to develop [1].

*The maxillary sinuses* are the first sinuses to develop. At birth, the maxillary sinuses are rudimentary and have a volume of 6–8 cm. In infancy, the growth of the maxillary sinuses continues, and the final phase of pneumatization is completed after the permanent teeth erupt. Unevenness in the size and shape of the maxillary sinuses is common. Unilateral maxillary sinus hypoplasia has been reported to be present in 7% of adults, whereas bilateral maxillary sinus hypoplasia has been reported to be present in 2% of adults.

*Ethmoidal air cells* are pneumatized at birth, even though they achieve maturation in adulthood.

*The sphenoid bone* is compact at birth and harbors hematopoietic marrow; pneumatization of the sphenoidal sinuses starts as early as 2 years of age. Sinus maturation is completed by the age of 14 years. Sphenoidal sinus aplasia is very uncommon. Therefore, the lack of the pneumatization of the sphenoidal sinuses in patients less than 10 years old indicates underlying pathology [2]. Sphenoid sinus pneumatization arrest is recognized radiologically by nonexpansile lesions with osteosclerotic margins and internal fat content (marrow fat) within the skull base [3, 4].

*The frontal sinuses* are the last sinuses to develop, as their growth begins after 2 years of age. Unilateral frontal sinus aplasia occurs in 15% of adults, and bilateral frontal sinus aplasia occurs in 5% of adults, as documented in the literature [2].

Regarding the development of the paranasal sinuses, hypoplasia is considered a delay in the pneumatization of the sinuses after 8 years of age, while aplasia is considered a total lack of pneumatization in individuals aged 10 years and older [5].

A wide spectrum of variations in PNS structures have been mentioned in the literature; for instance, bilateral maxillary sinus severe hypoplasia/aplasia [6], combined aplasia of the sphenoid, frontal, and maxillary sinuses along with ethmoid sinus hypoplasia have been reported in a 47-year-old woman with complaints of headache, nasal obstruction, and postnasal drip [7]. Bilateral aplasia of a single sinus [8] and even combined aplasia of the sphenoid and frontal sinuses accompanied by bilateral maxillary and ethmoidal sinus hypoplasia were reported in a female patient aged 54 years who presented with nasal obstruction and persistent headache; bone window CT of the paranasal sinuses confirmed the diagnosis [9]. Furthermore, total aplasia of the PNSs with normal nasal cavities has been described in a 57-year-old woman [10].

CT is the gold standard for the radiological workup for paranasal sinus abnormalities (developmental and acquired pathological) in pre- and postsurgical assessments, as bone window CT has the best accuracy in outlining the sinuses and provides the maximum detail of bony structural, anatomical, and architectural variations [11].

Numerous syndromes related to sinonasal anomalies are generally divided into those due to defects of midfacial skeletal growth and those attributed to bone dysplasia (osteosclerosis) that prevent sinus pneumatization [10].

Defects of midfacial skeletal growth include more than 100 forms of craniofacial dysostoses. Among the most known midfacial skeletal growth defects are Treacher-Collins syndrome (also known as mandibular dysostosis), Binder's syndrome/maxillomandibular dysplasia, Williams syndrome, elfin facies syndrome, Apert's syndrome, and Crouzon's syndrome, all of which are related to bilateral paranasal sinus effects. Moreover, in Goldenhar's syndrome (hemifacial microsomia), a unilaterally diminutive maxillary sinus is accompanied by the hyperplasia of the ipsilateral bony zygoma [10, 12]. However, none of the abovementioned conditions, alone or in combination, is associated with nasal cavity aplasia, which is the major aspect of this case report.

Among anomalies attributed to bone dysplasia (osteosclerosis) that prevent sinus pneumatization, many hereditary bony dysplasias produce changes within the skull and facial skeleton through abnormal hardening of the bone and increased bone density, leading to sinus obliteration. Examples of these anomalies include polyostotic craniofacial fibrous dysplasia, craniometaphyseal dysplasia, craniodiaphyseal dysplasia, and other skeletal hyperostosis [12]. However, none of these features applied to the case presented here, as normal bone density was preserved in this patient.

Furthermore, Bosma syndrome is an extremely uncommon hereditary condition characterized by nasal aplasia, orbital abnormality, and sexual maturation defects. Although sinus hypoplasia can occur in Bosma syndrome, it is not generalized and commonly affects only the maxillary sinuses [13]. In the case presented here, both eyeballs, orbits, and their contents were unremarkable, and sinus aplasia was global.

Congenital nasal cavity abnormality is a component of solitary median maxillary central incisor syndrome (SMMCI), which is also a rare, complex genetic disorder consisting of midline craniofacial structural defects, including the nose (choanal atresia and nasal pyriform aperture stenosis) and developing brain (holoprosencephaly), along with pituitary gland malfunction [14]. Again, none of these components were observed in the case presented here; the maxillary incisors were normal, and no brain malformations were detected.

Hence, the case presented here is rare, in that it is of an individual with unique structural abnormalities that represent nasal cavity and paranasal sinus congenital/developmental defects.

## Conclusions

Among the wide range of anomalous structural defects described in the region of the nose and paranasal sinuses, total aplasia of the paranasal sinuses and nasal cavity and its components, identified radiologically by multidetector bone window CT, is reported here for the first time in a Middle Eastern patient.

## Abbreviations

PNSs: Paranasal sinuses; CT: Computed tomography; ENT: Ear, nose, and throat; SMMCI: Solitary median maxillary central incisor.

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## Author's contributions

The author conceived of the study, analyzed, and interpreted the radiological findings, established manuscript design and organization, designed the illustrations, and drafted the manuscript. The author read and approved the final manuscript.

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Not applicable

## Availability of data and materials

The data and materials supporting the findings of this study are available on request from the corresponding author.

## Declarations

### Ethics approval and consent to participate

This study was approved by the ethical committee of Hawler Medical University. Written informed consent was obtained from the patient for the publication of this case report and accompanying images.

### Consent for publication

The patient included in this case report gave written informed consent for the publication of the data and materials contained within this study.

### Competing interests

The author declares no competing interests.

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