World Journal of Surgery and Surgical Research

Case Report
Published: 20 Feb, 2026

Solitary Median Maxillary Central Incisor Syndrome: A Case Report

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Abstract

Solitary Median Maxillary Central Incisor (SMMCI) syndrome is an uncommon developmental anomaly in which a single central tooth develops only in one-half of the maxilla and is located along the median line. They are commonly associated with other midline developmental malformations and systemic anomalies.

We report a case of an 8-year-old female patient with carious primary dentition and esthetic concern, in whom the isolated SMMCI was diagnosed. We discuss the embryological, clinical, and genetic aspects of SMMCI and highlight the significance of early diagnosis and a multidisciplinary approach for better prognosis.

Introduction

Solitary median maxillary central incisor (SMMCI) is a rare syndrome and an unusual dental anomaly characterized by the presence of a single central incisor located in the midline of the maxillary segment of the oral cavity in either primary or permanent dentition [1].

Originally described as a separate entity with other craniofacial development defects, SMMCI can also exist in isolation or be associated with systemic conditions, such as holoprosencephaly. It has been reported in 1 in 50,000 live births. The dental anomaly can have marked esthetic, functional, and psychological consequences [2].

Etiology is thought to involve the constitutive activation of the SHH (Sonic Hedgehog) pathway with genetic mutations, particularly SHH pathway genes, at a critical niche of embryogenesis, i.e., 35-38 days of gestation. SMMCI can be an early identifying feature of a systemic process, which may warrant further evaluation, so correct diagnosis and management early on are crucial [3].

Case Presentation

An 8-year-old girl presented to the dental department with posterior dental caries and a request for cosmetic treatment. Her medical history, including dental history, was normal. The patient was born to healthy parents, and her pregnancy was uneventful. At birth, her father was 33 years old and her mother was 27. The pregnancy and delivery were uneventful, and no developmental delays were observed during the child's early years.

A solitary right maxillary central incisor was found exactly midline, with no bilateral central incisor.

There were no other serious craniofacial anomalies, and the patient's general and neurological examination was unremarkable. Radiographic examination revealed the absence of a second maxillary central incisor and the formation of a symmetrical alveolar process (Figures 1 and 2). No signs of cleft or skeletal defects were observed. The diagnosis of isolated SMMCI was made because there was only one median maxillary central incisor, in the absence of syndromic signs.

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

Discussion

SMMCI is characterized by a mid-line positioned single central incisor in the maxillary dental arch. This anomaly is of interest because of its presumed relation to a host of important midline

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Received Date: *01 Feb 2026*
Accepted Date: *17 Feb 2026*
Published Date: *20 Feb 2026*

Citation:
Zbir R, Rachid B, Sakout M. Solitary Median Maxillary Central Incisor Syndrome: A Case Report. World J Surg Surgical Res. 2026; 9: 1616.

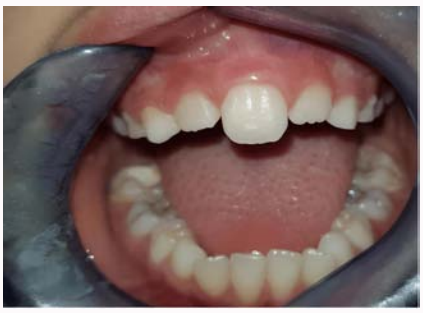

Figure 1: Intraoral photograph showing the presence of a solitary median maxillary central incisor.

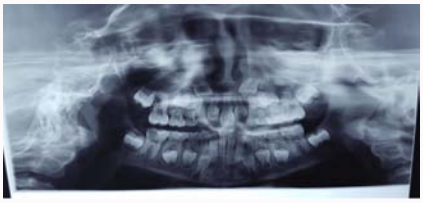

Figure 2: Radiographic evidence of solitary median maxillary central incisor.

developmental anomalies of the brain and face. Even if it is isolated, like in this patient, it deserves to be further investigated for associated anomalies that can have a subtle or late presentation.

This condition is of great clinical interest because it is rare, with an incidence of 1 in 50,000 live births, and it is frequently associated with various systemic anomalies, such as holoprosencephaly and a spectrum of brain and facial malformations [3].

The development of facial and dental anatomy results from the complex embryological phenomena that are generated between the 4th and 12th week of gestation. Days 35-38 also seem to be the key period for central incisor development. Interruption intra-utero, particularly impeding the SHH gene pathway, may lead to midline defects such as SMMCI. This gene is known to play a role in controlling cell growth, differentiation, and tissue patterning. Again, either severe or mild features may develop as a result of the mutations in SHH, and a neurological examination should be performed even if the disease has no manifestation [4-6].

Although such involvement was not found in this patient, systemic associations reported in the literature include endocrinopathy (e.g., panhypopituitarism), craniofacial malformations, and neurological dysfunction. Strabismus, hypotelorism, and even corpus callosum agenesis have been described. Due to the wide spectrum of the variability of the phenotype, a multidisciplinary approach involving genetic counseling, as well as neurology and endocrinology, is recommended [7].

SMMCI may be inherited as an autosomal dominant trait, with variable expressivity, and incomplete penetrance. Familial cases have been described, and molecular genetic testing, in particular searching for SHH mutations, can help to establish diagnosis and counseling. In other words, the lack of a family history and syndromic features is compatible with isolated presentation, though genetic testing might still be indicated to exclude mutations [8].

Diagnosis is dependent on clinical evaluation with the help of radiographs. In some questionable cases, CBCT or 3D imaging can help to demonstrate any associated skeletal abnormalities.

Management techniques are centered on functional and aesthetic rehabilitation, malocclusion correction, and long-term orthodontic and restorative treatment planning [9,10].

The treatment should be comprehensive and sequenced, including pediatric dentists, orthodontists, prosthodontists, and perhaps maxillofacial surgeons [11].

Orthodontic evaluation to evaluate arch development and space considerations is also part of long-term treatment planning. If the patient develops esthetic problems during adolescence, composite restorations, veneers, or even prosthetic replacement after orthodontic preparation may be an option [12].

Conclusion

This 8-year-old girl with isolated SMMCI emphasizes the need for early identification of rare dental anomalies that may have systemic implications. No syndromic features were observed in such a patient, but the association between this condition and severe developmental disorder was reported, which requires interdisciplinary attention.

Dentists are the key to recognition not only of these anomalies but also in coordination with medical professionals for systemic evaluation and genetic counseling.

In the present case, the care plan included restorative management of decayed teeth, maintenance of oral hygiene, and long-term orthodontic monitoring to guide occlusal development and anticipate future esthetic corrections, if necessary.

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