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# Non-Congenital Cystic Teratoma of the Maxillary Sinus - A Rare Occurrence

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## Abstract

**Background:** Teratomas of the head and neck due to their extremely controversial origin, bizarre microscopic appearance, unpredictable behaviour of development, and often dramatic clinical presentation are a clinical surprise when encountered. Teratomas are germ cell tumours that contain tissues of variable histological maturity and have a known malignant potential, which is unpredictable from their histological features or stage of development. They are composed of various tissues of ectodermal, mesodermal and endodermal origin.

**Case report:** In our current case report, we have acknowledged the presence of teratoma which is a rare entity in the maxillofacial region in an 18-year-old male who was successfully treated by complete excision and showed no signs of recurrence.

**Conclusion:** Since something as bizarre as a teratoma is a rare occurrence in the head and neck region along with its potential to mimic a lot of other lessons which are more common and typical in the maxillofacial region A correct diagnosis of a teratoma can only be made with a proper histopathological study preceded by clinical and radiographic correlation.

**Keywords:** Teratoma, Dermoid tumour, AFP (Alpha Fetoprotein), Teratoids, Epignathus.

## Introduction

Teratomas are defined as true neoplasms composed of multiple tissues foreign to the site from which they originate.[1] To put in simpler terms, they are congenital germ cell tumours that contain tissues of variable origin (viz ectoderm, mesoderm, and/or endoderm), maturity and possess a known malignant potential, which is often

difficult to predict based on their marked histological features and/or the stage of their development.

Teratomas occur in approximately 1 in every 4000 live births, showing a more female predilection, and have an 18% risk of other congenital malformations, some of which can be incompatible with life.[2] The most common sites of incidence of such lesions are usually located in a midline or paraxial location from the brain to the sacral area with sacrococcygeal being the most common site.[1]

However, Teratomas rarely originate from other superficial facial structures of the head and neck. The actual rate of occurrence in the head and neck region is reported to be only 10%. [3,4] Of these head and neck teratomas, a pure oral presentation is especially rare and only 13 cases of pure oral teratomas (seven from the tongue and six from the hard palate) have been reported in the literature, to our knowledge.[1]

Here we report a case with a teratoma in an adult which originated from the maxillary sinus and was presented as a swelling on the hard palate which was deceptively suspected to be an infected dentigerous cyst before its histopathological analysis.

**Case report**

An 18-year-old male patient reported to the Department of Oral and Maxillofacial Surgery, Jaipur Dental College, with a chief complaint of swelling and fluid discharge from his right maxillary premolar region for the past 25 days. The patient presented with a history of first observing a noticeable growth like swelling 6 months ago but chose not to seek any medical attention for the same. However, in the past 25 days, the patient experienced a significant increase in the size of swelling along with a continuous straw-coloured discharge from the site. The patient also gave a history of congenitally missing tooth at the site of swelling which was examined to be a missing right maxillary premolar (According to Universal Tooth Numbering System #13). Apart from his current complaint, the patient did not give any contributing medical history and also claimed to be without any other symptoms apart from the swelling for the past 6 months. (Fig 1)

On inspection of the extraoral features of the swelling, it was found to be extending superiorly from the epicanthal fold below the lower eyelid up till the commissures of the mouth inferiorly and from the septum of the nose medially to the prominence of cheeks laterally. It was measured to be 5x6 cm in dimension roughly. The skin over swelling appeared normal and showed no signs of inflammation. The nasal septum showed deviation towards the left side of the patient. On Intraoral inspection, swelling in the right maxillary vestibular area measuring about 2x3 cm extending from the mesial aspect of the maxillary right central incisor to the second premolar while at the same time obliterating the vestibule. It was accompanied by the presence of a peculiar foul odour.

On further clinical evaluation, the swelling was firm and hard to touch and was palpable extra-orally with its extent superiorly from the infraorbital ridge till the commissures of the mouth inferiorly and from the septum of the nose medially to the malar prominence laterally.

To ascertain a clinical diagnosis, the swelling was then aspirated using a narrow needle (#25 Gauge) and a straw-coloured fluid was yielded as its result which on further evaluation revealed the presence of protein content of 3000 mg/dl in the aspirate.

Radiographically, an orthopantomogram revealed a well-defined unilocular radiolucency extending from the mesial aspect of the right maxillary central incisor to the distal aspect of the right maxillary second premolar with

a well-defined sclerotic border. The right maxillary sinus revealed a cloudy picture with a concave border, making it appear as a fluid-filled sinus. Further Computed tomographic evaluation on axial and coronal sections revealed a cystic pathology present in the right maxillary sinus and deviating the nasal septum towards the left side. (Fig 2 & Fig 3)

Based on the clinical, radiological, and pathological lab findings, a provisional diagnosis of infected dentigerous cyst associated with impacted right maxillary 1st premolar, involving the maxillary sinus was formulated.

After obtaining informed consent, the cystic lesion along with the impacted premolar was enucleated under general anaesthesia using an intraoral approach, followed by intranasal antrostomy for drainage. The excised specimen was sent for histopathological examination for confirmation of provisional diagnosis. (Fig 4)

The H and E staining revealed a soft tissue section that showed a cystic lesion lined by non-keratinized stratified squamous epithelium. The underlying tissue was made up of dense bundles of collagen fibers with numerous fibroblasts. The connective tissue was deeply infiltrated by chronic inflammatory cells predominantly lymphocytes, a few macrophages, giant cells, and numerous reactive plasma cells. The connective tissue also showed the presence of small islands of osteoid tissue with groups of mucous glands dispersed throughout. Numerous endothelial lined blood vessels with extravasated RBCs were seen as well as the presence of some pseudostratified ciliated columnar epithelium typical to the epithelium common to the respiratory tract were also evident in the specimen. Smooth muscle fibers were also found to be scattered throughout. All these histopathological features resembling an array of various types of tissues convey that the tissue yielded from the maxillary sinus was indeed a Teratoma of Immature Variety with Cystic degeneration hence a final diagnosis of Non-Congenital Cystic Teratoma of Maxillary Sinus of Immature Variety was made. (Fig 5)

The patient was observed and followed up for 1 year and he did not report any post-op complications along with the absence of any symptoms of reoccurrence or any malignant transformation.

**Discussion**

Teratomas are seldom referred to as monstrous lesions that are composed of tissues foreign to the part in which they arise. They are tumours that are characterized by the presence of various tissues of ectodermal, mesodermal, and endodermal origin. These tissues exhibit various degrees of maturation. They can arise in new-borns, children, or adults alike.

Based on the type of tissue present in the teratoma, it can be of the following types

- Mature teratoma is the principal benign germ cell tumour and shows histologic features similar to its counterparts in the gonads and other extragonadal locations.
- Immature teratomas are composed of variable quantities of immature tissue elements mostly neuroepithelial that are interspersed with mature and immature tissues derived from the three embryonic germ layers

Based on histological classifications which are widely used (Arnold's system) they are classified as [5]: -

- Dermoid tumours: These makeup for most common types to be encountered and are composed of only ectoderm and mesodermal tissues.
- Teratoids: These are tumours that are poorly differentiated and usually contain cells/tissues from all three germ layers.

- Teratomas: Similar to Teratoids these contain tissue from all three germ layers. However, these are histologically more identifiable than teratoids. They may be of solid or cystic variety.[1]
- Epignathus: These are tumours that contain fully developed organs and appendages. Other terms used for these include ‘‘fetus in fetu’’ or ‘‘parasitic fetus,’’ containing fetal organs. Epignathus is a misnomer, and its etymological meaning is ‘‘upon the jaw.’’ However, it has been used for almost every ter0atoma of the oral cavity and pharynx tissues.[1]

The histogenesis of teratoma formation remains debatable. The most popular theory suggests that, presumably due to alterations in cellular membrane chemistry, teratomas arise from totipotential embryonic tissues that are somehow displaced during ontogeny. This leads to a collection of various tissues which are often alien to the site in which they arise.[6]

Ninety percent of head and neck teratomas present during the neonatal and infantile period, predominantly involving the neck and nasopharynx and occurring only once in every 20,000–40,000 births.[7] Any other presentations of teratomas extracranially are very rare. Only 13 cases of pure oral teratomas (seven from the tongue and six from the hard palate) have been reported this far in the literature.[1] Teratomas arising from the paranasal sinuses and tonsil are even rarer. Only Two cases of malignant teratomas arising in the ethmoid sinus have been reported in adults.[8]

Calcification and cysts within a mass are more typical of teratomas of the head and neck than of other site.[2] Cystic lumen containing caseous, sebaceous, purulent material with hair, nail, and fat globules have also been reported.[9] Teratomas of these types on palpation are cystic, but this area alternates with areas that are solid.[2]

A precise patho-radiological correlation is required to confirm the diagnosis as it presents with such a bizarre presentation. Plain radiographs of the head demonstrate calcifications within the lesion.[5] Ultrasonography establishes the presence of solid and cystic components and can differentiate a cyst from surrounding tissue. Of all the available modalities, MRI has been proven to be better than other imaging modalities, as it can locate the precise position, extension, and demarcations of the lesion.[10] Teratomas are not enhanced when viewed by administration of contrast material because of their avascularity and thus can cause diagnostic confusion with choriostoma, endodermal sinus tumours, and granular cell tumours. The important blood investigation is alpha-fetoprotein (AFP).[4] AFP is generally elevated and it returns to normal after total excision.[11] Alpha-fetoprotein (AFP) is a reliable indicator of disease activity and some authors advocate serial serum AFP levels as a marker of teratoma recurrence. They have been shown to increase in cases of malignant transformations or so-called teratocarcinoma.[12]

Clinical differential diagnoses of oropharyngeal teratoma include cystic hygroma, lymphangioma, duplications, and neuroblastoma.[5] This lesion should also be differentiated from encephalocele, glioma, haemangioma, congenital rhabdomyosarcoma, and neurofibromatosis.[8] Histopathologic ally differential diagnosis include hamartoma, dermoid cyst, and a heterotopic gastrointestinal cyst. It is seen with growth, oral teratomas extend outside the mouth rather than toward the oropharynx. That is why the incidences of respiratory distress may be less especially in adults [5].

Early surgical intervention is necessary. Surgical management aims to remove the diseased mass as well as to provide a good airway and Esthetic in long-term follow-up. Furthermore, the extirpation of mass should

be done as soon as possible, although there is no initial upper airway obstruction, as the problem develops eventually the surgical [8] The final goal is to remove the complete lesion, as any residual tumour may be the cause of recurrence. If the histological analysis shows any malignant components, adjuvant chemotherapy and radiotherapy are indicated [5].

In this case, the lesion was presumed to be a cyst probably a dentigerous cyst based on the aspirate and some deceiving clinical features, however on eventual excisional biopsy, it was revealed the cystic cavities were lined by stratified squamous epithelium along with some pseudostratified respiratory epithelium, and underlying the epithelium were groups of mucous glands, small islands of osteoid tissue and transitional epithelium. Smooth muscle fibers were also found to be scattered throughout. Which gave it a diagnosis of Immature Teratoma and was successfully treated, and followed up and the patient did not report any signs of reoccurrence in the past 1 year. During follow-up, AFP should be monitored. In our case, all the AFP levels were found to be within the normal range.

The overall prognosis for oral teratomas is good. Though benign most of the time, these may result in a high degree of mortality and morbidity especially in new-borns due to variations in their size and location. Similarly in new-borns, if the lesion is large enough, it may be the cause of airway obstruction, respiratory distress, dysphagia, difficulty in eating, and pain due mostly to infection in the lesion. In malignant teratoma, radio-chemotherapy is used after surgical removal of the tumour [12] They are very less likely to produce any problems antenatally. Postnatally, early surgical intervention is necessary. Early correct histopathological diagnosis, the establishment of a good airway, complete excision of the tumour, and timely follow-up should be the code to deal with any oral teratoma be it in new-borns or adults.

**Conclusion**

Some lesions that occur in the maxillofacial region have a more common cystic radiographic appearance. It is difficult to diagnose these lesions just based on just clinical and radiographic features. To formulate a precise final diagnosis a detailed case history, evaluation of associated signs and symptoms, radiographic imaging and early histopathological examination are vital.

As seen in this case, a simple case of suspected dentigerous cyst turned out to be a monstrous lesion of immature teratoma with cystic malformation arising from the maxillary sinus which was successfully excised and reported no malignant transformation or recurrence till 1 year of follow-up.

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**Legend Figures**

Figure 1

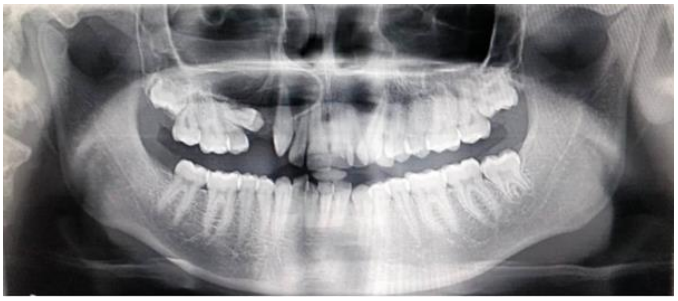

Figure 2

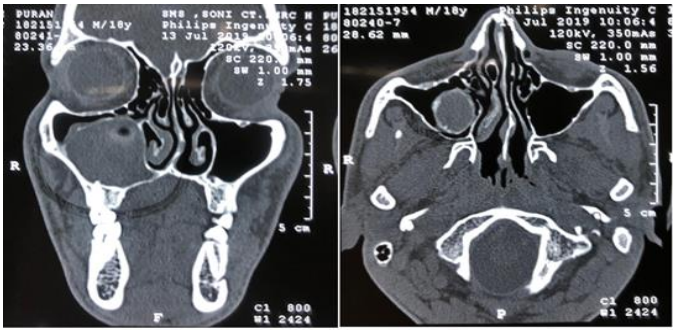

Figure 3

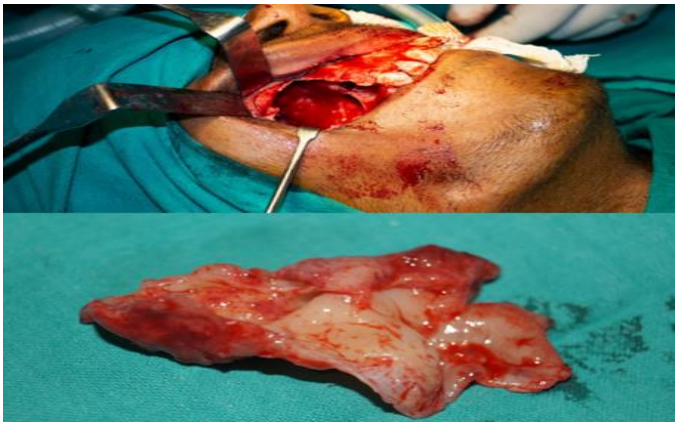

Figure 4

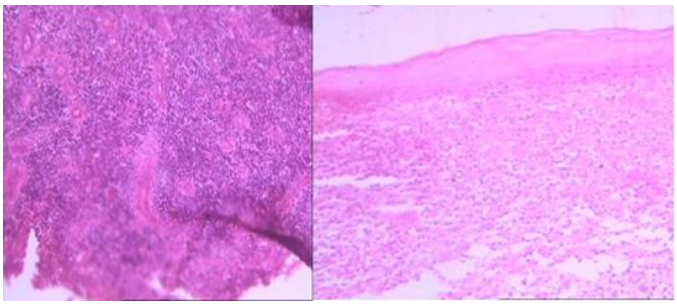

Figure 5