Case Report

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SINONASAL SQUAMOUS CELL CARCINOMA IN A PEDIATRIC PATIENT WITH INTELLECTUAL DISABILITY: A RARE ENTITY

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ABSTRACT:

Sinonasal squamous cell carcinoma is one of the rarest epithelial neoplasms mainly affecting older individuals. Squamous cell carcinoma of head and neck is rare in pediatric patients i.e. fewer than 2% of all pediatric head and neck malignancies. Among these, tongue and lower lip are the most frequently reported sites and sinonasal malignancies are exceedingly rare. In pediatric patient with intellectual disability poor communication, feeding habits and poor oral hygiene are thought to be considered as risk factors. Inflammatory-like, rapid growing lesions of the oral cavity can be considered to be squamous cell carcinoma as one of the differential diagnosis although it's occurrence in pediatric patients is rare. In this article we have reported a case of sinonasal well differentiated squamous cell carcinoma in 14 years old child with Intellectual Disability.

Key Words: Sinonasal squamous cell carcinoma, Intellectual Disability

INTRODUCTION:

Incidence of nasal cavity and paranasal sinus carcinomas account for 0.2-0.8% of all human malignant neoplasms. Among them, sinonasal squamous cell carcinoma (SNSCC) is one of the rarest epithelial neoplasms and represents about 3% of all malignancies of the head and neck region. In respect to SCC from maxillary sinus it mainly affects mid-aged men (55-65 years old) from Eastern countries.[1]

Although chronic upper respiratory tract infections, nasal congestive symptoms, epistaxis, and rhinosinusitis are much more prevalent in the pediatric population (defined by American Academy of Pediatrics as patients under age 21) and they manifest symptoms that overlap with those of sinus malignancy.[2] Here we are presenting an extremely rare case of SNSCC in 14 years old orphan male with an intellectual disability (ID).[3]

CASE DETAIL:

A 14 years old male patient with an ID from an orphanage of mentally challenged children run by Delhi Government reported to Department of Oral Medicine & Radiology, Maulana Azad Institute of Dental Sciences, New Delhi due to pain and swelling in right cheek region since one month. Patient was apparently well 45 days back to start with pain and

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swelling with right maxillary first, second premolars and first molar. Associated teeth were mobile and painful, extracted 30 days prior to reporting us considering periapical periodontitis as diagnosis. Even after extraction, patient did not get relief, rather swelling & discomfort increased.

Clinical examination revealed a single, well defined (approximate size of 5 x 5cm), shiny, soft to firm, febrile, tender extraoral swelling in middle-third of right maxillary region, extending from right zygomatic arch to right angle of mouth (Figure 1). Patient didn't have any special risk factor or habit. The right submandibular lymph node was palpable, mobile and tender, no other cervical lymphadenopathy was evident.

Intraoral examination showed a proliferative growth with approximate size of 3cm x 3cm encompassing the area of right maxillary premolars and first molar alveolar ridge region with grade 2 mobile canine and second molar. Growth was fixed to underlying tissue, tender with indurated margins and obliterating the buccal vestibule (Figure 2).

Panoramic radiograph showed extensive osteolytic lesion involving complete right maxillary sinus & alveolus with destruction of floor and other walls of maxillary sinus (Figure 3). Contrast Enhanced Computed Tomography of head and neck showed heterogeneously enhancing soft tissue mass lesion in right maxillary sinus with osseous erosions of hard palate, medial and lateral pterygoid plates, inferior orbital wall with extension of soft tissue into sphenoid sinus, right side of nasal cavity (Figure 4, 5).

The blood investigation reports of the patient were within normal physiological range except highly increased Erythrocyte Sedimentation Rate i.e. 80 mm. Hence tuberculosis was ruled out.

Fine Needle Aspiration Cytology from right cheek swelling was suggestive of possibility of SCC. After an incisional biopsy from intraoral growth; a diagnosis of well-differentiated SCC was made (Figure 6, 7). Biopsied tissue was assessed for Human Papilloma Virus (HPV) and Epstein Barr Virus (EBV) detection, but both found negative. Considering histopathology report and computed tomography findings final diagnosis of sinonasal well differentiated SCC was made.

DISCUSSION:

SNSCC is one of the rarest epithelial neoplasms of the head and neck region. It predominantly occurs within the maxillary sinus (60-70%)[1] which was consistent with our case. Very few cases of SCC of sinonasal region in pediatric patients have been reported in the medical literature, but most common reported sites are tongue and lip.[4–8] The most common pediatric head and neck malignancies include non-Hodgkin lymphomas, Hodgkin lymphomas, rhabdomyosarcomas, thyroid malignancies, nasopharyngeal carcinomas, salivary gland malignancies, and neuroblastomas.[2,9,10] A relatively rare location for SCC is sinonasal region. Santos et al (2014)[1] assessed all maxillary sinus SCC patients in year 1997 to 2006 in

single Brazilian institution, concluded mean age of patient 58.7 years. In our case SCC was present in sinonasal region in 14 years old orphan patient with ID which is an unusual finding.

Etiologic factors associated with oral SCC in adults includes tobacco, alcohol, viruses and nutritional deficiencies, that do not necessarily apply to pediatric cases as exposure time would be comparatively short for the establishment of a cause-effect relation.[4,11,12] Thus, factors should be investigated such as: genetic predisposition, previous viral infection, feeding habits, and immunodeficiency states, occupational exposure to the carcinogenic factor (wood, leather dust), socioeconomic condition, oral hygiene and trauma.[13–16] As our patient was from an orphanage and with ID his oral hygiene, feeding habits were compromised, can be considered as possible etiological factors.

Lack of communication is a common factor in the overall poor quality of health care for people with ID and cancer.[17] It has a detrimental effect in relation to late diagnosis of cancer and subsequent misdiagnosis of associated symptoms, which was seen in our case.

Toner and O'Regan (2009)[18] concluded that non-smokers males with oropharyngeal cancer, related to high risk HPV infection. Yukashi Yamashita et al (2015)[19] said that HPV infection has important role in tumorigenesis , malignant transformation. Doescher J et al (2015)[20] stated that EBV infection is strictly associated with the metastasis of SNSCC. Hence in our case, biopsied sample was studied for HPV and EBV but both were negative.

S.A.H. Stolk-Liefferink et al (2008)[21] reported a case of 11 year old boy with histopathological diagnosis of OSCC with maxillary gingiva and summarized case reports of OSCC in pediatric patients from 1970—2005 , with most common sites were tongue, gingiva. He concluded although rare, but SCC does occur in pediatric patients hence it should be included in the differential diagnosis of inflammatory-like, rapid growing lesions of the oral cavity. Similarly in our case, habit history was negative and patient was misdiagnosed as periapical pathology with associated teeth lead to delayed diagnosis.

Nina L. Shapiro (2009)[2] extracted cases of pediatric sinus cancer from the Surveillance, Epidemiology, and End Results database (1988–2005). 63 pediatric sinus cancers were identified with a mean age at diagnosis of 10.5 years and a 1:1 male to female ratio. Rhabdomyosarcoma was the most common followed by sarcoma and olfactory neuroblastoma and concluded maxillary sinus the most common site for sinonasal cancers. So SNSCC is an unusual finding.

Aziz Binahmed (2007)[4] stated a case of SCC of the maxillary gingiva and alveolus in a 10-year-old girl and suggested biopsies of nonhealing lesions in the oral cavity are essential. The prognosis of the disease can be altered by early diagnosis and surgical treatment. In our case both

factors age and ID resulted in delayed presentation of SNSCC.

As very few case reports are presented in literature regarding SCC in pediatric age group and specially SNSCC in pediatric patient with ID is not reported so far, as found in our case.

This case report indicates that SNSCC can occur in pediatric patients. As this is the rarest entity misdiagnosis leads to delay in diagnosis and further management for same. After confirmation of diagnosis patient was undertaken for chemotherapy followed by radiotherapy. A strategic, coordinated management of services including the development of specific policies and protocols which is essential in delivering effective care for people with both cancer and ID.

CONCLUSION

A poorer prognosis in pediatric patients with ID could be due to the presentational and diagnostic delays because of the reduced expectation of cancer in this age group and poor communication with such patients. To manage the patient with ID effective communication skill is mandatory. Although rare, SCC does occur in pediatric patients and should therefore be included in the differential diagnosis of inflammatory-like, rapid growing lesions of the oral cavity.

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FIGURES

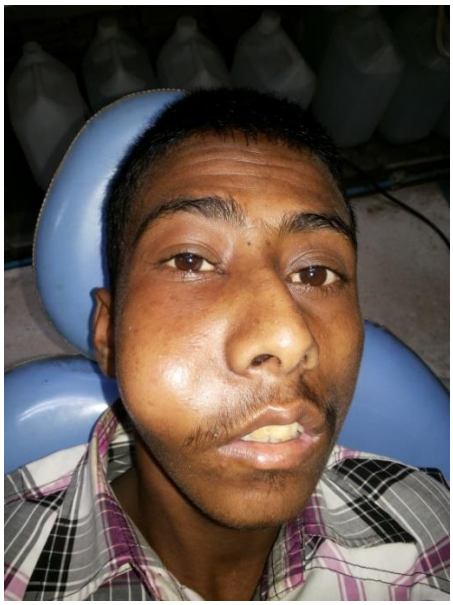


Figure 1: Profile photograph

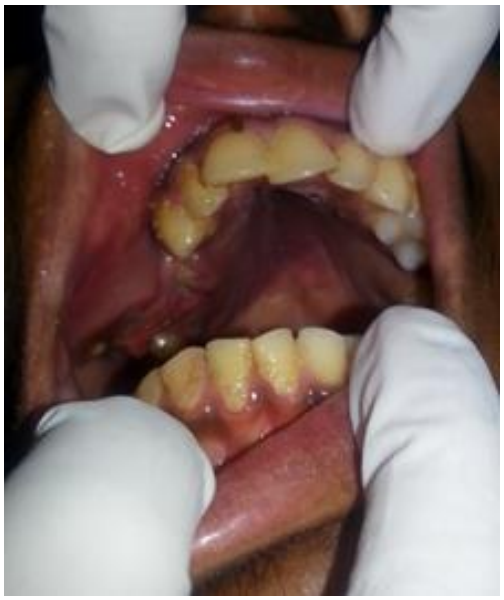


Figure 2: Intraoral photograph

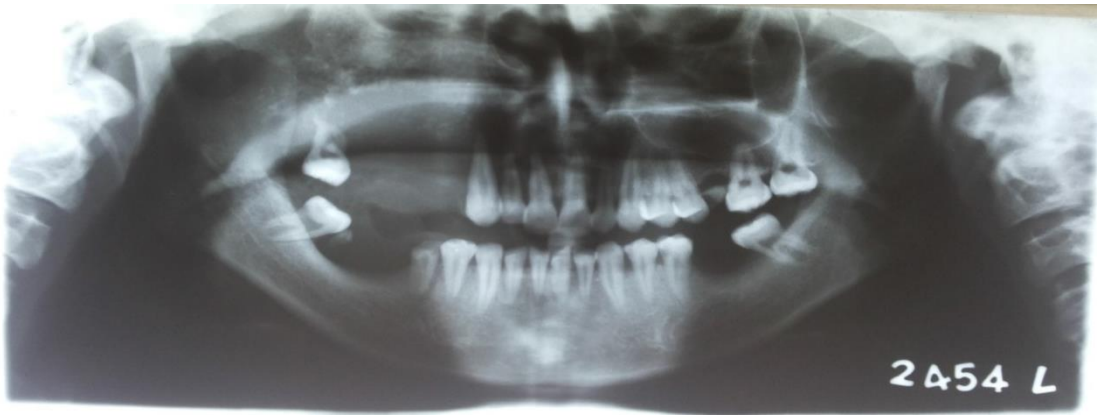


Figure 3 : Panoramic radiograph showed extensive osteolytic lesion involving complete right maxillary sinus & alveolus

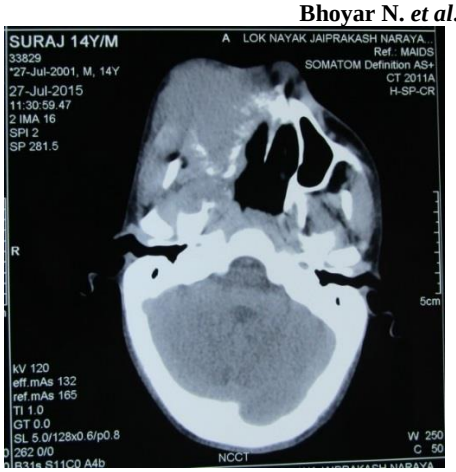


Figure 4: Axial section

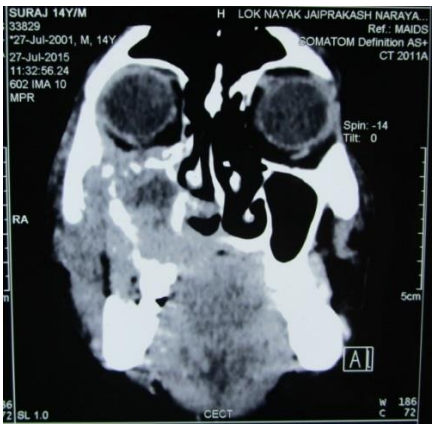
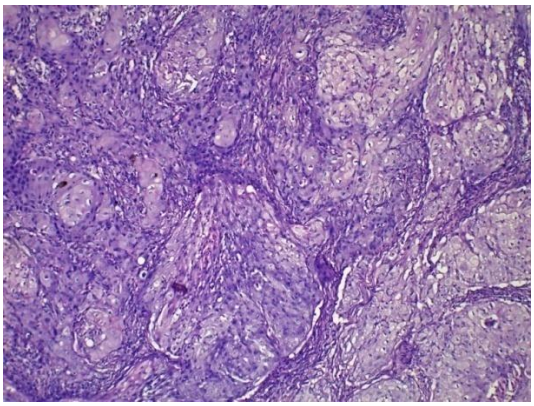


Figure 5: Coronal section

Figure 4 & 5: CECT showed heterogeneously enhancing soft tissue mass lesion in right maxillary sinus with osseous erosions and extending into right nasal cavity



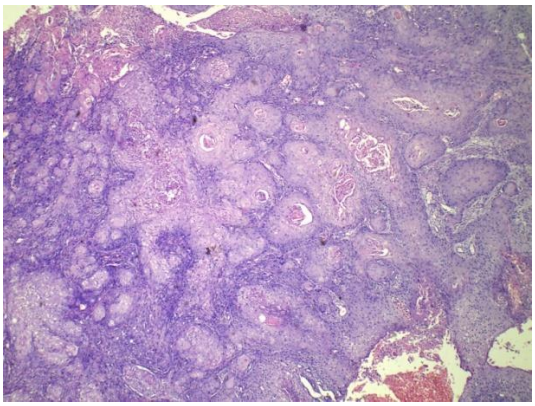


Figure 6,7 : Photomicrograph of H & E stained tissue section showing dysplastic cells, mitotic figures and keratin pearls