Squamous Cell Carcinoma of Mandible in a 10-year-old girl

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ABSTRACT

Aim: To study the importance of early recognition of oral cancer in children and its most suitable management.

Background: Head and neck cancers are the sixth most common cancer worldwide with rare incidence in children. It is associated with a poor prognosis.

Case description: A 10-year-old healthy girl with good oral hygiene presented with complaints of lesion in right mandible for 3 months duration. Incision biopsy of lesion showed squamous cell carcinoma. Computerized tomography showed cortical erosion of mandible, with no significant neck nodes. Case was discussed in multidisciplinary team and options were weighed. After discussing with patients' relatives, it was decided to proceed for surgery.

Conclusion: Surgery was undertaken, patient underwent wide excision of lesion with segmental mandibulectomy, neck dissection and free fibular reconstruction of the defect. She is on regular follow-up for the past one year, with no evidence of lesion.

Clinical Significance: Squamous cell carcinoma of mandible in children is very rare. Treating physician should be aware of such unusual presentation of highly mortal disease. Literature review on treatment being sparse and surgery followed optimal reconstruction is a major challenge.

Keywords: Free fibular flap, Malignancy in children, Oral cancer, Pediatrics, Squammous cell carcinoma.

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BACKGROUND

Head and neck cancers are the sixth most common cancer worldwide, with annual reported cases being 630,000

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and it amounts to 350,0000 death every year. Among these malignancies, more than 90% are squamous cell carcinoma (SCC), with such a huge figures being depicted there were only 217 cases paediatric head and neck SCC reported, till June 2016, of which only 5% have involved the mandible. Only 20.6% patients with head and neck SCC belonged to age group less than 10 years.^{1,2}

Generally SCC in children is thought to be aggressive and has a poor prognosis. Here we report a case of SCC involving mandible in a 10-year-old girl and how the case was managed.

CASE DESCRIPTION

A 10-year-old healthy girl presented to our institution with complaints of lesion in right lower alveolus for 3 months (Fig. 1), which was associated with loosening of tooth. There was no history of similar illness in the family. Clinical examination showed 3 x 2 cm proliferative growth in right lower alveolus extending from right premolar region involving adjacent lower gingivobuccal sulcus, floor of mouth was free. No clinically significant neck node was palpable. Systemic examination was within normal limits. Clinically it was a T4aN0M0 disease of right lower alveolus.

Biopsy from the lesion was reported as moderately differentiated squamous cell carcinoma. Computerized tomography (Fig. 2) showed a 3x2 cm lesion in mandible with cortical breach, there was no clinically significant neck node. She was taken for proposed surgery with informed and written consent.

Wide excision of lesion (Fig. 3) with adequate soft tissue and bony margin amounting to segmental mandibulectomy (part of arch of mandible was also resected), right extended supraomohyoid neck dissection and surgical defect reconstruction with free fibular flap harvested from left lower limb was done on April 2017 (Fig. 4).

Histopathological examination was reported as 2x2.5x0.1 cm well differentiated squamous cell carcinoma of right lower alveolus with noncohesive tumour front. Perineural invasion present no lymphovascular emboli seen, and all margins of excision are adequate. Decalcified section of bone was free of tumor. Lymphnode 37/37 showed only reactive change.

Postoperative period was uneventful. She was kept of close follow-up. She is on regular follow-up with no evidence of disease (Fig. 5) or any problem related to disease or cosmesis till date.





Fig. 1: Preoperative intaoral photograph of lesion

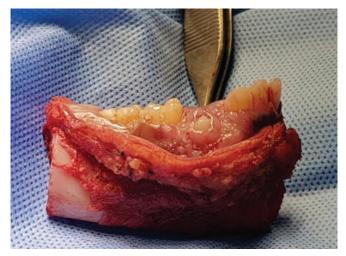


Fig. 3: Surgical specimen



Fig. 5: Six months post surgery

DISCUSSION

SCC is the most common neoplasm affecting head and neck region which commonly affects person between fifth and seventh decade of life. Lesion affecting adolescence is uncommon, but those involving first decade of life is a rare entity. In most of the studies, there is

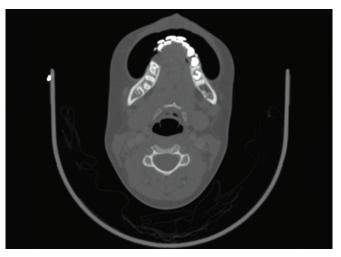


Fig. 2: Computerized tomography showing erosion of mandible



Fig. 4: Post surgery, day 2, photograph

a neglectable sex predilection towards male side. The most common site of SCC of oral cavity in children is the tongue followed by gingiva.²⁻⁴

The most common part in oral cavity involved by squamous cell carcinoma in children is tongue followed by gingiva, because it has alcove of stem cells with a rapid turnover and hence chance of evolution into disease. The other cause of children developing malignancy is mainly because of genetics, due to flaws in DNA repair mechanism. One of the main genetic cause is fanconi's anemia, it can also be due to xeroderma pigmentosum or keratitis, icthyosis, deafness (KID syndrome) which will predispose to SCC. Other possible reasons being early consumption of tobacco in childhood, passive smoking, viruses like human papilloma virus and herpes virus.^{5,6}

SCC in children has generally a poor prognosis compared to adults; this might be due late presentation of disease by the child or might be related to poor tumor behavior, ultimately leading to reduced survival.⁷

The earliest age where a case of SCC oral cavity got diagnosed, was on the 12th day of life, in a neonate, way

back in 1932, who underwent surgery on day 14 which was followed by radiotherapy which was started on 22nd day of birth.⁸

Various modalities of treatment are available, in a curative intent, which includes su rgery alone, radiotherapy alone or surgery followed by adjuvant radiotherapy or concurrent chemoradiotherapy. In our case, surgery alone was adequate with a close follow-up.

Reconstruction of mandible, when needed, following mandibulectomy is very important aspect in this surgery, because of esthetic and psychosocial reasons. Mandible help to assist in mastication, deglutition and speech. Continuity of this bony skeleton is the essence of lower framework of face, thus playing an important role in facial appearance and also facial expression.

Mandibulectomy can be a marginal mandibulectomy or segmental mandibulectomy. Segmental manbidulectomy will disrupt the continuity of bone mandating reconstruction. Several options are available to establish bony continuity following segmental mandibulectomy which includes regional pedicled flaps, skin graft, reconstruction plates and free flaps. Free flaps are the preferred option for reconstruction of mandible, with donor site being fibula, radius, iliac bone, scapula etc. In our case, segmental mandibulectomy was done because of the bone expansile type of lesion, and CT scan showed cortical bone erosion and we used fibula for reconstruction, which was a workhorse of mandible reconstruction.

Free flap in children will pose challenges especially due to small caliber of vessel, airway issues and anesthesia related problems, which all warrant a careful proceeding of surgery. Fibular flap has the several advantages main being simultaneous surgery by two team, plastic surgeon and oncosurgeon, it can be used to reconstruct entire mandible, long vascular pedicle, osseointegrated dental implants can be placed. The major disadvantage being: donor site morbidity, fibula does not grow concomitantly with the facial skeleton, thus might necessitate revision surgery, atrophy of the graft over time, but it did not proceed to an extent were osseointegrated was not possible. The optimal duration for osseointegration is not explained.

CONCLUSION

SCC of mandible in children is rare. Diagnosis of which should be promptly made. Undue prolongation of diagnosis by physician will lead to upstaging of disease and a bad prognosis. Treatment of SCC mandible is difficult with most difficult part being the reconstruction. Despite many advancement in management, these tumors tend to be aggressive, and overall survival still remains poor.

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