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RESEARCH ARTICLE

INTRAOSSEOUS LOW GRADE MUCOEPIDERMOID CARCINOMA- A RARE CASE REPORT AND BRIEF REVIEW OF LITERATURE

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ABSTRACT

Background: Intraosseous Mucoepidermoid carcinoma(MEC) is a rare tumour in the head and neck region. Its clinical and radiologic features mimic many common lesions in the jaws. The need for an Elective neck dissection (END) along with resection as well as need for adjunctive radiotherapy has been a long standing controversy, thus making treatment modalities a subject of debate. **Objective:** To formulate a correct diagnosis and treatment plan for this lesion in order to prevent recurrence and failure of treatment which may cost the patient dearly. **Case Report:** We present the case of a middle aged man diagnosed with Low Grade Intraosseous Mucoepidermoid Carcinoma in the mandible. Surgical management included resection of the lesion along with an elective neck dissection. Additionally the patient was subjected to radiotherapy. Follow up period of the patient has been 1 year. **Conclusion:** A patient presenting with an osteolytic lesion in the jaw must be thoroughly evaluated. However innocuous the lesion might seem to be, it may turn out to be something far more serious. In low grade intraosseous MEC where surgery is the mainstay of treatment, the tumour size also plays an important role as a prognosticator for the disease. Even if it is a low grade tumour histologically, advanced stages indicated by tumour size or invasion of surrounding structures, change the entire course of the disease. As a result, the management of any such disease should be done judiciously and should include an elective neck dissection and/or radiation.

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INTRODUCTION

Salivary gland cancers account for around 5% of all head and neck cancers. Of these, mucoepidermoid carcinoma (MEC) is the most common. MECs are most commonly seen in the parotid and is the most malignant salivary gland tumour in children. In most cases of MEC, histological grading and clinical characteristic correlate, thus treatment decisions are commonly based on histologic grading. The tumours range from high grade to low grade, with an intermediate grade in between. However determination of the grade is not dependent on histolopathology alone (Marx Robert, 2012).

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The clinical features of each vary and are also important in the determination of the grade (Marx Robert, 2012). Another variant, the intraosseous MEC, first reported in 1939, commonly occurs in the mandible and is almost always low grade in terms of histology and behavior. The decision to proceed with elective neck dissection (END) as well as need for postoperative radiotherapy in these cases is influenced by the probability of finding microscopic disease in a patient without clinical evidence of lymph node metastases. In contrast to squamous cell carcinoma of the head and neck, there is currently no standardized protocol for need of END in salivary gland carcinoma. This is attributed to the relatively low incidence of these tumors as well as their histopathologic diversity. However in recent years great advancements have been achieved with respect to predicting the biology and the behavior of such tumour.

For mucoepidermoid carcinoma (MEC), not only does the grade, but so also the stage influences the treatment outcome.

CASE REPORT:

In January 2019, a 43 years old male patient visited our department with the chief complaint of intermittent pain in his lower left posterior teeth region. A brief history was taken. The patient did not present any comorbidity, was a nonsmoker and a teetotaler. On extraoral physical examination, no signs of swelling or facial asymmetry were seen. On intraoral examination, there was no obvious swelling or buccal and lingual expansion (Fig 1), both on inspection and palpation. There was no dental displacement. An aspiration test was performed and it turned out negative. The patient complained of paresthesia on the left lateral half of chin which was progressing in intensity with time. CBCT of face (Fig 2) was done and it revealed a unilocular hypodense image with a thin sclerotic border in the left ramus of the mandible, commonly seen in odontogenic cysts.

Fig 1. Intraoral exam

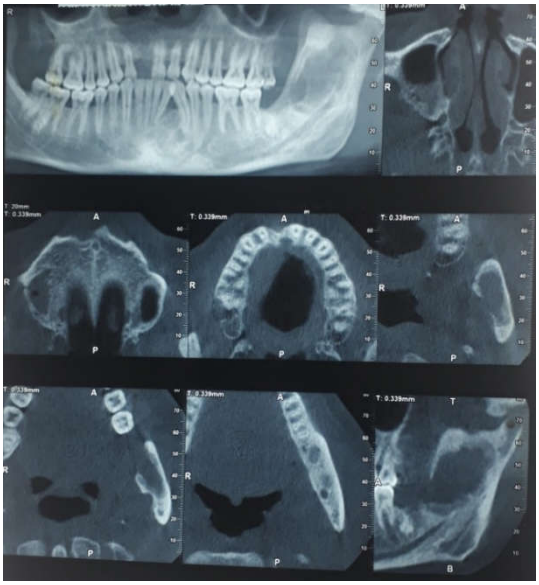

Fig 2. CBCT

However, there was a distinct breach in the lingual cortex, more suggestive of a tumourous process. In addition, paresthesia reported by patient was difficult to ignore as a warning sign in the diagnostic work up as it was suggestive of invasion in the nerve sheath.

After a routine blood investigation, an open biopsy was performed. Histopathological examination was suggestive of low-grade intraosseous mucoepidermoid carcinoma A PET-CT was done to rule out the presence of any cervical and distant metastasis. Neck and distant metastasis was not seen in PET-CT. However, breach of mandibular lingual cortex due to the presenting disease was once again appreciated. The size of the tumour was mentioned as 4.3 cms approximately, in the greatest dimension, thus warranting a neck dissection. A surgical plan of segmental resection of mandible plus disarticulation on the left side along with an elective supra omohyoid neck dissection (SOHND) was planned for the patient considering the fact that the disease in the local area was widespread. Peroperatively a wide margin was ensured on the lingual aspect and the entire inferior alveolar nerve traced and removed along with the surgical specimen and a SOHND was performed on the ipsilateral side (Fig 3,4 and 5). The margins were checked by frozen section and were found to be clear of disease. Histopathological examination of the surgical specimen was confirmatory for low grade MEC and in addition showed perineural invasion too, thus warranting radiotherapy. The immediate and late post operative period showed uneventful healing. The patient has been on periodic follow up (Fig 6) for the last 1 year in our institution and is doing well.

DISCUSSION

Mucoepidermoid carcinoma (MEC) is the most common salivary gland malignancy and accounts for 15% of all salivary gland tumors (Harrison, 2013), whereas the intraosseous variant comprises only 2% to 3% of all MEC (DeFreitas, 2018; Bell, 2016) and occurs more frequently in the posterior region of the mandible especially the third molar and the premolar regions. It was first reported in 1939. Although the exact pathogenesis of Intraosseous MEC is unknown, there are several theories pertaining to its origin. The following may represent origins for these lesions: (a) ectopic salivary gland tissue or remnants of embryonic salivary glands trapped within the bone; (b) transformation of mucous cells found in odontogenic cysts; and (c) maxillary sinuses or submucosal and mucosal glands with intraosseous extension (Johnson, 2008). More recently, intraosseous salivary tissue has been found to be present in 0.3% of the bone specimens of all maxillary bones studied by Bouquot *et al*. (2000) providing new evidence for the origin of intraosseous salivary carcinomas. Although its origin is still a matter of debate, central MEC is now a recognized entity (Pires, 2003). Traditionally, mucoepidermoid carcinomas have been classified into three histopathological grades using the following criteria: quantity of cystic formation, degree of cellular atypia, and relative number of mucous, epidermoid, and intermediate cells. Low-grade tumors exhibit prominent cystic formation, minimal cellular atypia, and a relatively high proportion of mucosal cells. High-grade tumors consist of solid islands of squamous and intermediate cells, which may demonstrate considerable pleomorphism and mitotic activity. Mucus-producing cells may be infrequent, and sometimes, it may be difficult to distinguish the tumor from squamous cell carcinoma. However, those of intermediate grade exhibit characteristics that are located between low and high-grade tumors (Harrison, 2013). Mucoepidermoid carcinomas and the intraosseous variants are more common in middle-aged adults at the age of 40-50 years and have a slight preference for females (DeFreitas, 2018; Pequeno, 2019). They are three times more common in the mandible than in the maxilla (DeFreitas, 2018).

Fig 3: Peroperative photographs

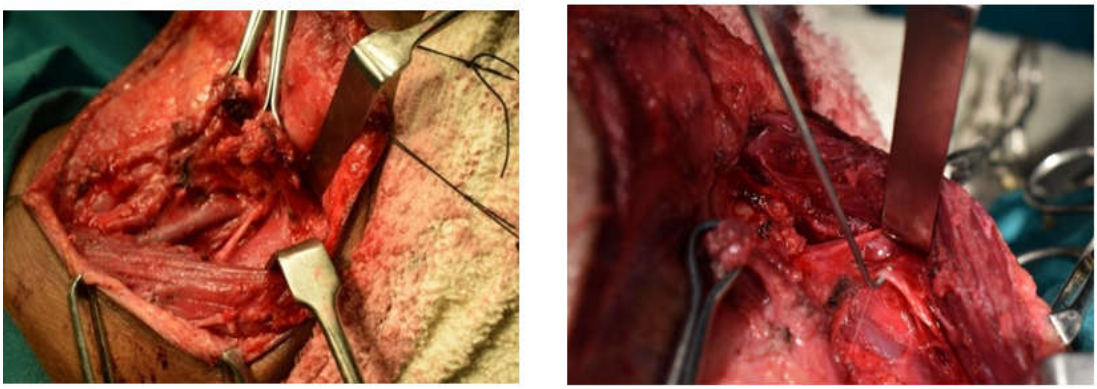

Fig 4: Peroperative photographs showing neck dissection being performed and the Internal Jugular vein and Spinal Accessory Nerve in the surgical bed

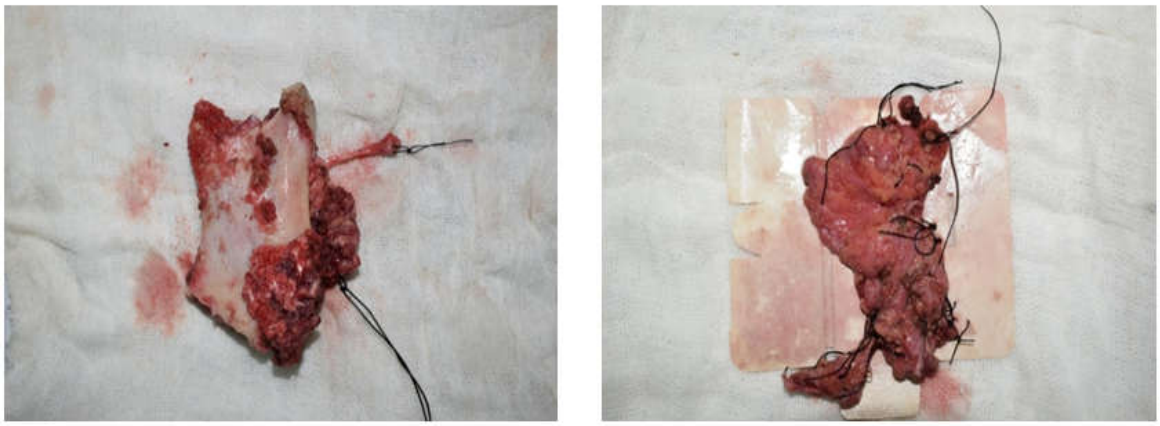

Fig 5. Surgical specimen showing the resected portion of the mandible along with the inferior alveolar nerve and the excised ipsilateral neck nodes (Level I to Level III)

Fig 6. Follow up

They present frequently with cortical bulging, although some lesions may be discovered as an accidental finding on radiographs. Pain, trismus, and paresthesia although seen, are rare in these lesions. Metastases have been reported in 12% of the cases (DeFreitas, 2018).They occur primarily to regional lymph nodes and rarely to the ipsilateral clavicle, lung, and brain especially in long standing lesions. Imaging is important in the detection and differentiation of MEC. Radiographically, one half will present as a well circumscribed unilocular radiolucency (Marx Robert, 2012) with a sclerotic periphery, the other half will present as a multilocular radiolucency with imaging characteristics similar to those of other lesions. The diagnostic list often includes ameloblastoma, glandular odontogenic cyst, keratocystic odontogenic tumour, dentigerous cyst, central giant cell tumour (Marx Robert, 2012).

Panoramic radiography and conventional computed tomography (CT) are commonly used as imaging procedures. The presence of tooth dislocation and root resorption are common findings. Cortical bone perforation and extension to surrounding soft tissues is indicative of aggressive behavior (Chan, 2013). Brookstone and Huvos have prepared a classification system based on the radiographic properties of the tumor, and this classification system is now used for determining the prognosis. According to this classification, the lesions with an intact cortex layer and without any bony expansions are categorized as stage 1 and have the best prognosis (Brookstone, 1992; Başaran Bora, 2018). Lesions that expand the bone but do not disturb the integrity of the cortex are stage 2 (Brookstone, 1992; Başaran Bora, 2018). Lesions that disrupt the integrity of the periosteum or cause cortical perforation with or without nodal involvement and masses with nodal involvement are accepted as stage 3, with the poorest prognosis (Brookstone, 1992; Başaran Bora, 2018). Accordingly, our case was a Stage 3. Although confinement to bone is a diagnostic criterion for intraosseous MEC, absence of prominent soft tissue invasion in presence of cortical perforation is sufficient to prove its central origins. The final diagnosis along with the treatment plan should be based on all of clinical, radiographic, and histopathological characteristics and not on clinical and radiological character alone. Central MECs do not differ much from their soft tissue counterparts and most of them are low grade (Marx Robert, 2012). Although FNAC is difficult to perform in such cases (Başaran Bora, 2018), when done it is better as a diagnostic aid for high and intermediate grade MECs compared to low grade MECs. Surgery is the mainstay of treatment. Central MECs usually have a 90% cure rate with prompt and definitive surgery. These lesions are best treated with a resection of bone with 1 to1.5 cms margins. Unless the nerve is at a distance of more than 1 cms from the tumour, the nerve should be included in the resection. Post operative radiotherapy and/or neck dissection is not required unless the clinical behavior or histologic grading is more advanced than low grade[1]. Multiple authors have reported that for salivary gland cancer, postoperative radiation improves prognosis for patients with tumors over 4 cm, but is of minimum benefit for patients with smaller tumors. Tumors over 4 cm and those with a positive margin post surgery are suggested to be an absolute indication for adjuvant treatment in salivary gland cancer (Kokemueller, 2005). Thus in our case, a tumour size of approximately 4.3 cms in the greatest dimension, along with perineural invasion, together contributed to our decision making regarding post operative radiotherapy.

In 1992, Armstrong *et al*. (1992) published a landmark study in which the rates of lymph node metastases were evaluated retrospectively in 474 salivary gland cancer patients. Clinically positive nodes were present in 14% of the patients and clinically occult, pathologically positive lymphadenopathy was reported in 12%. They inferred that only size and grade were significant risk factors. Tumors greater than 4 cms in size were at a greater risk for occult metastasis and so also high grade tumours, as a result of which END should be done in such cases. Thus, in our case, the tumour size being greater than 4 cms in the greatest dimension,a decision of performing END was made. In 2001, Santos *et al*. (2001) further investigated the indications for END in clinically N_0 patients with a retrospective analysis of 145 parotid cancer patients, 29 of whom had MEC (20%). Multivariate analysis, just as in Armstrong's study confirmed a statistically significant greater risk of cervical metastases for high-grade tumors. Advanced tumor (T) stage was identified as another independent risk factor. Thus in agreement with Armstrong's study, the authors suggested performing an END only in the presence of these prognostic indicators. Stennert *et al*. (2003) in their study state that incidence of occult metastasis in low risk salivary gland cancers might be greater than originally thought. Spiro (178) in a clinicopathologic study of 367 cases states that whenever there is a discrepancy between the grade and the stage of the tumour (that is high grade, Stage1 or low grade, Stage 3), the outcome was most influenced by the clinical stage rather than by the histological appearance. Herein, Spiro emphatically

states that RND should be considered in any patient with a T3 tumour, without evidence of node metastasis, regardless of the histologic appearance (Spiro, 1978).

CONCLUSION

The histologic grade of MEC is an effective indicator of the risk of lymph node metastases across multiple standardized grading systems. Along with the histological grade, the size of the tumour also seems to be an effective tool in decision making as to whether END is indicated or not. Advanced T stage has emerged as an important risk factor for occult disease. Elective neck dissection may additionally spare radiation dose to the neck in patients and thus keep them free from painful side effects of radiation. Low and intermediate grade MEC in the absence of other high-risk factors, have a lower propensity of nodal metastases, and under these circumstances do not require END. Elective radiation may be an effective alternative to END in the clinically N_0 MEC patient and should be considered when patient comorbidities or other factors dictate a minimalistic surgical approach. Therapeutic decisions cannot and should not be made on the basis of histological appearance alone.

Glossary of Abbreviations

END: Elective Neck Dissection
CBCT: Cone Beam Computed Tomography
MEC: Mucoepidermoid Carcinoma
PET-CT: Positron Emission Tomography-Computed Tomography
SOHND: Supra Omohyoid Neck Dissection

REFERENCES

Marx Robert E, 2012. Stern Diane, Oral and maxillofacial pathology: A Rationale for diagnosis and treatment, 2nd Edition

Harrison BL, Sessions RB, Kies MS, 2013. Head and Neck Cancer: A Multidisciplinary Approach,4th Edition

DeFreitas GB *et al*. 2018. Intraosseous Mucoepidermoid carcinoma in the mandible. Case Reports in Dentistry, Vol, ID 9348540.

Bell D, Lewis C, El Naggar AK, Weber RS. 2016. Primary intraosseous mucoepidermoid carcinoma of the jaw: reappraisal of the MD Anderson Cancer Centre Experience, Head and Neck, Vol 38, no S1,ppE1312-E1317

Johnson B, Velez I. 2008. Central mucoepidermoid carcinoma with an atypical radiographic appearance. *Oral Surg Oral Med Oral Pathol Oral Radiol Endod.,* 106:e51-3.

Bouquot JE, Gnepp DR, Dardick I, Hietanen JH. 2000. Intraosseous salivary tissue: jaw bone examples of choristomas, hamartomas, embryonic rests, and inflammatory entrapment: another histogenetic source for intraosseous adenocarcinoma. *Oral Surg Oral Med Oral Pathol Oral Radiol Endod.,* 90:205-17.

Pires FR, Paes de Almeida O, Lopes MA, Elias da Cruz Perez D, Kowalski LP. 2003. Central mucoepidermoid carcinoma of the mandible: report of four cases with long-term follow-up. *Int J Oral Maxillofac Surg.,* 32:378-82.

Pequeno Ana Luiza Viana *et al*. 2019. Intraosseous mucoepidermoid carcinoma: *Arch Head Neck Surg.,* 48(2):e00122019.

Chan KC, Pharoah M , Lee L , Weinreb I , Perez-Ordonez B. 2013. Intraosseous mucoepidermoid carcinoma: a review of the diagnostic imaging features of four jaw cases, *Dentomaxillofacial Radiology*., 42, 20110162

Brookstone MS, Huvos AG. 1992. Central salivary tumors of the maxilla and mandible: A clinicopathologic study of 11 cases with an analysis of the literature. *J Oral Maxillofac Surg.,* 50: 229-36.

Başaran Bora, Doruk Can, Yılmaz Eren, Sünnetçioğlu Ecem, Bilgiç Bilge, 2018. Intraosseous Mucoepidermoid Carcinoma of the Jaw: Report of Three Cases. *Turk Arch Otorhinolaryngol.,* 56: 42-6

Kokemueller, H., Brueggemann, N., Swennen, G., & Eckardt, A. 2005. Mucoepidermoid carcinoma of the salivary glands—clinical review of 42 cases. Oral Oncology, 41(1).

Armstrong JG, Harrison LB, Thaler HT, *et al*., 1992. The indications for elective treatment of the neck in cancer of the major salivary glands. *Cancer.,* 69:615–619.

Regis De Brito Santos I, Kowalski LP, Cavalcante De Araujo V, Flavia Logullo A, Magrin J. 2001. Multivariate analysis of risk factors for neck metastases in surgically treated parotid carcinomas. *Arch Otolaryngol Head Neck Surg.,* 127:56–60.

Stennert E, Kisner D, Jungehuelsing M, *et al*. 2003. High incidence of lymph node metastasis in major salivary gland cancer. *Arch Otolaryngol Head Neck Surg*.,129:720

Spiro RH, Huvos AG, Berk R, *et al*. 1978. Mucoepidermoid carcinoma of salivary gland origin. A clinicopathologic study of 367 cases. Am J Surg 136:461.

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