Original Research Paper

Volume-9 | Issue-1 | January-2019 | PRINT ISSN - 2249-555X

Dental Science

EXTREME AMELOBLASTOMA OF MANDIBLE: THE CHALLENGES AND THE RESOLUTIONS

Dr. Vinay Kumar Sharma Senior Resident, Oral and Maxillofacial Surgery, Deptt of Dentistry, Government Medical College and Hospital, Chandigarh-160030

Dr. Preeti Sharma* Senior Resident, Oral and Maxillofacial Surgery, Deptt of Dentistry, Government Medical College and Hospital, Chandigarh-160030 *Corresponding Author

ABSTRACT Ameloblastomas are locally aggressive tumors of odontogenic origin which if left untreated may acquire enormous sizes. Such lesions pose a treatment challenge because of their size, the sheer amount of bony destruction, the usual poor systemic condition of the affected patient, intraoperative complications and the difficulties associated with reconstruction. Such lesions are called Extreme or Giant Ameloblastomas. We present a rare case of extreme ameloblastoma affecting the entire mandible and the systematic management protocol for the case. We also discuss in detail about challenges associated with such lesions and the step wise management approach.

KEYWORDS : Extreme ameloblastoma, Giant ameloblastoma

INTRODUCTION

Ameloblastoma is a well-recognized prototype of benign odontogenic tumors. Molecular alterations in ameloblastoma are such that it shows relentless growth with infiltration of marrow spaces[1-4]. It can exhibit voluminous size if neglected and left untreated. Complications, like starvation and hypoproteinemia, eventually lead to death in such cases[5]. In today's time, due to easy availability of medical care and increased health awareness, it is uncommon to find such 'extreme' ameloblastoma. Only 10 such cases are reported in literature[6]. We report a rare case of extreme ameloblastoma which was difficult to treat because of its monstruous size and extent.

CASE REPORT

A 50-year-old female reported with complaint of large swelling of left side of the face which had been gradually increasing in size for last 10 years. The patient had not actively sought treatment before. Clinical examination revealed bony hard swelling with indistinct margins involving most of the mandible. Overlying skin was normal and no temperature changes or parasthesia was observed. Intraorally, mandibular arch was edentulous while maxillary arch had multiple root stumps. Mandibular cortex was expanded and perforations were noted throughout.

Orthopantographic evaluation disclosed multilocular radiolucent lesion occupying almost the whole mandible (Fig.1). Cone Beam CT further confirmed the finding showing radiolucent multiple locules destroying the mandibular bony matrix. A provisional diagnosis of ameloblastoma was made. Considering the large size and involvement of whole mandible it was designated to be a Giant or Extreme ameloblastoma.

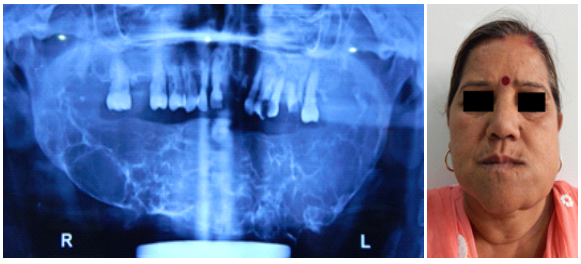

Fig.1 Clinico-radiologic picture of the patient

Surgery was planned under general anesthesia and tracheotomy was done. A submandibular approach combined with intraoral incisions was used to gain access to the tumor. The entire tumor was resected sparing only the proximal condylar stumps on both sides, which were deemed to be disease free and instrumental for reconstruction efforts. A free fibular microvascular graft was then harvested and shaped to create the mandibular outline (Fig 2). It was fixed to the condylar stumps. Layerwise closure was done and meticulous post operative monitoring was executed. The patient recovered well and was discharged after two weeks.

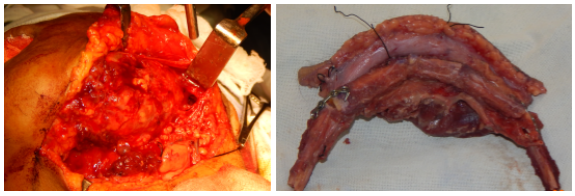

Fig 2. Tumor resected and free fibula harvested

Microscopic examination of hematoxylin and eosin (H&E) stained sections from the specimen showed multiple islands and follicles of odontogenic epithelium in the background of fibrous connective tissue. The epithelial islands were made up of tall columnar cells resembling ameloblasts, arranged in peripheral layers. This was consistent with the diagnosis of follicular ameloblastoma.

DISCUSSION

Ameloblastoma is a tumor of the jaw that typically occurs in the third and fourth decades of life and shows no gender predilection. These tumors arise from embryonic remnants of odontogenic cysts, dental lamina, enamel organ, or stratified squamous epithelium of the oral cavity[10]. Although the pathogenesis of ameloblastoma remains unknown, many pathogenic mechanisms have been proposed, and associations with human papillomavirus, chronic trauma and inflammation, malnutrition, and vitamin deficiency have been described[11].

The entity is known for its propensity of growth up to large dimensions and proclivity for recurrence. 'Giant' or 'extreme' ameloblastomas are the lesions which show huge dimensions, causing gross facial disfigurement and functional disturbances. Although benign, such extreme ameloblastomas can become life threatening due to associated problems. Lethal complications related to such voluminous size of tumor are starvation due to restriction from feeding, airway obstruction, and hypoproteinemia due to leakage of proteins into large cystic spaces of tumor.

Studies have shown that ameloblastoma accounts for 40 to 60% of all received specimens of odontogenic tumors in various Institutes in India[7,8,9]. In spite of availability and easy access to the medical care in developing countries, sometimes ameloblastomas of enormous sizes are confronted in practice. At least 10 cases of extreme ameloblastomas are reported till date.

The diagnosis of ameloblastoma is based on histologic and radiologic findings. Unicystic ameloblastomas, especially alveolar tumors, can be confused with cysts, but clinical behavior can be useful in the differential diagnosis. Most multicystic ameloblastomas are readily identified by OPT and CT and typically occur in the mandibular molar region and ascending ramus. Radiologic images often show a “soap-bubble” aspect of large loculations and a “honeycombed” appearance of small loculations[12]. Despite clinical and radiologic suspicion, in our

experience, a biopsy with the patient under local anesthesia is always preferable, especially in case of extensive lesions, to achieve a pathologic diagnosis. Unicystic and peripheral ameloblastomas behave similarly to dentigerous cysts, and the smallest lesions can be treated adequately by curettage (extending to the periosteum for peripheral tumors). However, conventional ameloblastomas tend to infiltrate intact cancellous bone trabeculae at lesion peripheries before bone resorption becomes radiographically evident.

Thus curettage of the tumor often leaves microscopic islands of neoplasm in the bone, which explains the 50% to 90% rate of recurrence after curettage reported in various studies[13]. Most (85%) ameloblastomas are multicystic, showing more aggressive behavior and a greater tendency to recur than unicystic ameloblastoma[14].

The management of very extensive lesions, such as those reported in this study, is difficult. Preservation of thin bone margins increases the risk of fracture and recurrence and prevents resection with safe margins, which are strongly recommended in extensive lesions (the impossibility of intraoperative histologic examination requires resection with a 1-cm margin beyond the macroscopic limits of the tumor). Most affected patients are young, and dental rehabilitation should be considered in the choice of surgical resection and reconstruction approaches. Extensive curettage can compromise tooth stability and necessitate immediate or delayed extractions. In these cases the residual bone is typically inadequate for dental implant placement, and additional surgeries, including bone grafting to restore sufficient bone height, are required[13]; these procedures have unpredictable results and require lengthy oral rehabilitation periods. Thus locally advanced pathology is best treated with radical surgical resection consisting of a partial mandibulectomy and immediate reconstruction of the defect, which ensures complete tumor resection, prevents recurrence, and enables fast and safe dental rehabilitation.

Correct surgical planning is key to successful management. Stereolithographic models enable preoperative planning of the resection and direct modeling of the reconstructive titanium plates, which make flap positioning safer and faster.

Mandibular reconstruction can be performed with bone grafts or bone-containing free flaps. Reconstruction of a segmental mandibulectomy with immediate bone grafting has been widely reported[15,16]. Although satisfactory results can be achieved with this technique for small reconstructions, the use of bone grafting to treat extensive tumors has several disadvantages.

The amount of bone provided limits the ability to reconstruct large resections, and unpredictable bone graft resorption further compromises the final result. Furthermore, bone grafts do not allow the reconstruction of soft tissue defects, which are often created by giant ameloblastomas.

Finally, the impossibility of immediate dental implant placement prolongs rehabilitation time and increases the number of surgical procedures. Thus we reserve the use of bone grafting for the reconstruction of small mandibular defects after subperiosteal resection and prefer to use bone-containing free flaps for extensive reconstructions, such as that reported in this study. Such flaps are our first choice because they provide sufficient bone and allow dental implant placement at the time of reconstruction and because composite flaps can be harvested for soft tissue reconstruction.

The fibula free flap is our first choice for long or lateral defects involving the ramus because it provides an adequate length of bone and can be modeled to simultaneously reproduce the mandibular ramus and body. This flap is very reliable and adaptable, provides a large amount of bone tissue for prosthetic rehabilitation, and has a long vascular pedicle. Its only limitation is that its height (usually 5 mm) may present a prosthetic problem, especially in patients (often young) with ameloblastomas who have undergone partial resection and retain dentition on the healthy side. In these cases, to reach the occlusal plane, the flap can be positioned ideally for prosthetic rehabilitation, with its upper border at the level of the residual alveolar crest, but this position has negative effects on the profile of the lower border. Thus the ideal solution is the use of a double-barrel flap to increase the height of the reconstructed mandible, simultaneously preserving the continuity of the inferior margin and the symmetry of the angles, which is mandatory for a satisfactory cosmetic outcome[15,16,17].

Segmental mandibular resection followed by immediate defect reconstruction with bone-containing free flaps should be considered as the treatment of choice for extreme mandibular ameloblastomas. This approach achieves the best functional and esthetic outcomes and the least recurrence.

REFERENCES

1. Pinheiro JJ, Freitas VM, Moretti AI, Jorge AG, Jaeger RG. Local invasiveness of ameloblastoma. Role played by matrix metalloproteinases and proliferative activity. Histopathol 2004;45(1):65-72.
2. Gortzak RA, Latief BS, Lekkas C, Slootweg PJ. Growth characteristics of large mandibular ameloblastomas: report of 5 cases with implications for the approach to surgery. Int J oral Maxillofac Surg 2006;35(8):691-695.
3. Sathi GS, Nagatsuka H, Tamamura R, Fujii M, Gunduz M, Inoue M, et al. Stromal cells promote bone invasion by suppressing bone formation in ameloblastoma. Histopathol 2008;53(4):458-467.
4. Pinheiro JJ, Nascimento CF, Freitas VM, Siqueira AS, Alves Junior SM, Jaeger RG. Invadopodia proteins, cortactin and membrane type I matrix metalloproteinase are expressed in ameloblastoma. Histopathol 2011;59(6):1266-1269.
5. Marx RE, Sterne D. Oral and maxillofacial pathology. A rationale for diagnosis and treatment. Illinois: Quintessence publishing Co; 2003.
6. Acharya S, Joshi A, Tayaar AS, Gopalkrishnan K. Extreme ameloblastoma of the mandible with hypoproteinemia: a case report and review of clinicopathological features. J Clin Exp Dent 2011;3(4):e343-347.
7. Sriram G, Shetty RP. Odontogenic tumors: a study of 250 cases in an Indian teaching hospital. Oral Surg Oral Med Oral Pathol Oral Radiol 2006;105(6):e14-e21.
8. Krishnapillai R, Angadi PV. A clinical, radiographic, and histologic review of 73 cases of ameloblastoma in an Indian population. Quintessence Int 2010;41(5):e90-e100.
9. Varkhede AS, Tupkari JV, Sardar M. Odontogenic tumors: a study of 120 cases in an Indian teaching hospital. Med Oral Patol Oral Cir Bucal 2011;16(7):e895-e899.
10. Hertog D, Bloemena E, Aartman IH, et al: Histopathology of ameloblastoma of the jaws; some critical observations based on a 40 years single institution experience. Med Oral Patol Oral Cir Bucal 17:e76, 2012
11. Gomes CC, Duarte AP, Diniz MG, et al: Review article: Current concepts of ameloblastoma pathogenesis. J Oral Pathol Med 39:585, 2010
12. Gupta V, Kapoor A, Shah N, et al: Evaluation of panoramic radiography and axial computed tomography in cases of ameloblastoma and central giant cell granuloma of mandible. Indian J Dent Res 12:200, 2001
13. Lau SL, Samman N: Recurrence related to treatment modalities of unicystic ameloblastoma: A systematic review. Int J Oral Maxillofac Surg 35:681, 2006
14. Bachmann AM, Linfesty RL: Ameloblastoma, solid/multicystic type. Head Neck Pathol 3:307, 2009
15. Cuesta-Gil M, Ochandiano Caicoya S, Riba-Garcia F, et al: Oral rehabilitation with osseointegrated implants in oncologic patients. J Oral Maxillofac Surg 67:2485, 2009 Oral Surg Oral Med Oral Pathol Oral Radiol Endod 108:661, 2009
16. Handschel J, Hassanyar H, Depprich RA, et al: Nonvascularized iliac bone grafts for mandibular reconstruction—Requirements and limitations. In Vivo 25:795, 2011
17. Chang YM, Wallace CG, Tsai CY, et al: Dental implant outcome after primary implantation into double-barreled fibula osteoseptocutaneous free flap-reconstructed mandible. Plast Reconstr Surg 128:1220, 2011