DOI: 10.7860/JCDR/2017/23818.9205

Case Report

Dentistry Section

A Rare Case Report of Spindle Cell Ameloblastic Carcinoma Involving the Mandible

MOHAMMAD ASIF KIRESUR[1], ARUNODAYA KUNCHE[2], ANURADHA ANANTHANENI[3], BHAVANA S. BAGALAD[4], PUNEETH HORATTI KUBERAPPA[5]

ABSTRACT

Ameloblastic Carcinoma (AC) is uncommon malignant epithelial odontogenic tumour of jaw, with characteristic histologic features and behavior. Clinically, it has aggressive, infiltrative growth pattern with a distinct predilection for mandible. It exhibits histologic features of ameloblastoma and gets dedifferentiated overtime to culminate in carcinoma. Majority of the cases arise denovo (primary) and only few cases arise from a pre-existing ameloblastoma (secondary). Spindle-cell differentiation in ameloblastic carcinoma is rare; Salter described it as a separate entity "low-grade spindle cell ameloblastic carcinoma. Here we report a case of 32-year-old female patient who presented with a swelling present for past six months. It was diagnosed as Spindle cell Ameloblastic Carcinoma (SpAC), after the hemimandibulectomy the patient was under regular follow up for 14 months, no sign of recurrence was seen.

Keywords: Ameloblastoma, Odontogenic tumour, Spindle cell differentiation

CASE REPORT

A 32-year-old female reported to the outpatient department of St. Joseph Dental College, Eluru, Andhra Pradesh, India, with a chief complaint of swelling on the right side of face present since six months [Table/Fig-1]. The swelling was associated with pain, difficulty in mastication and mouth opening. Patient presented with a history of similar swelling with mobility of teeth in right lower back region two years back for which she was surgically treated. Previous biopsy report confirmed the presence of ameloblastoma.

Clinical examination showed diffuse swelling in the middle third of the face which resulted in gross facial asymmetry. On palpation the swelling was smooth, non tender and firm in consistency. Right submandibular lymph nodes were palpable and non-tender. Intraoral examination showed a diffuse swelling with bicortical expansion extending from the distal side of first molar to the retromolar area obliterating both right buccal and lingual vestibule. The lesion was erythaematous and ulcerated [Table/Fig-2].

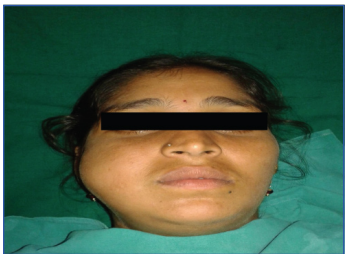
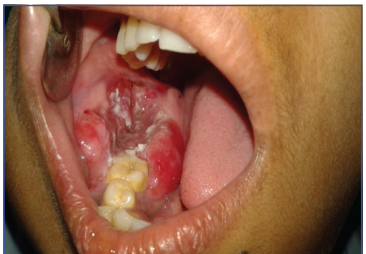
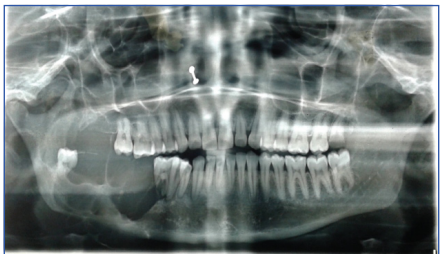
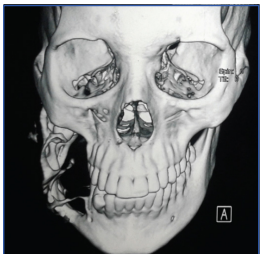
Orthopantomogram (OPG) showed presence of multilocular radiolucency extending from distal aspect of mandibular first molar to ramus of the mandible with displacement of tooth [Table/Fig-3]. The Computed Tomography (CT) scan showed a lobulated, expansile lytic lesion involving body, ramus, and coronoid process of mandible measuring 6x8x7.8 cm in size, with perforation of the cortical plates [Table/Fig-4]. Based on clinical and radiographic features, the lesion was provisionally diagnosed as ameloblastoma (recurrent) incisional biopsy from the lesion was microscopically examined and diagnosed as ameloblastoma. Right hemimandibulectomy was performed.

The excised lesion was sent for histopathological analysis. On gross examination, the lingual cortical plate was eroded; the tissue within the mandible appeared to be grayish black with irregular surface and firm in consistency [Table/Fig-5].

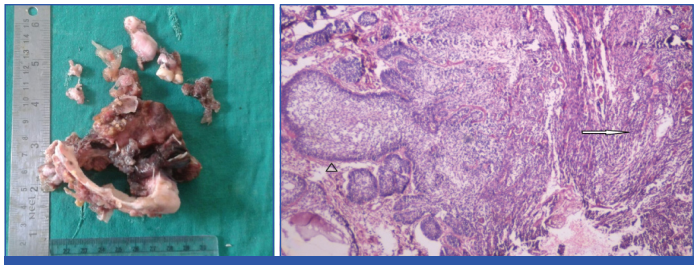
Microscopic examination showed large hypercellular epithelial islands with few cells displaying peripheral tall columnar cells with reverse polarity. Stellate reticulum like cells showed squamous differentiation and keratin pearl formation [Table/Fig-6,7]. Cellular atypia, nuclear hyperchromatism and increased mitosis were observed. Sheets of spindle cells were also observed [Table/Fig-8]. Immunohistochemical (IHC) staining with the cytokeratin-19 showed positivity in the epithelial islands and atypical spindle shaped cells [Table/Fig-9]. Based on the histopathological and IHC findings it was diagnosed as spindle cell variant of AC.

DISCUSSION

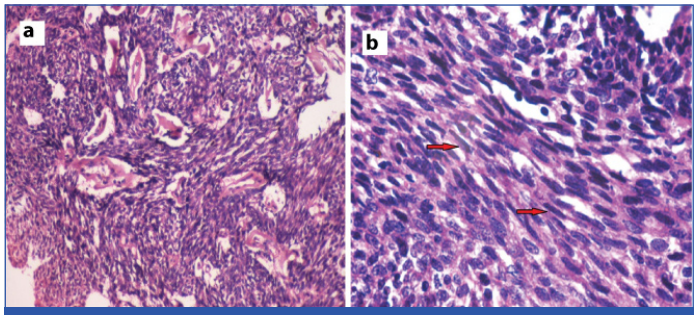
AC is a rare odontogenic tumour that shows the histological features of ameloblastoma with cellular atypia in the absence of metastasis [1]. According to Elzay RP and Slootweg PJ the term AC can be used to label a lesion that shows histologic features of both ameloblastoma and carcinoma [2,3]. AC can arise from odontogenic cysts, ameloblastoma, odontogenic epithelial rests, salivary gland epithelium and epithelium entrapped along embryonic fusion sites [4-6]. AC demonstrate two different variants depending on the differentiation: 1) De-differentiated (secondary) variant characterized by lesions that initially demonstrate the morphology of an ameloblastoma, but which de-differentiate in the course of

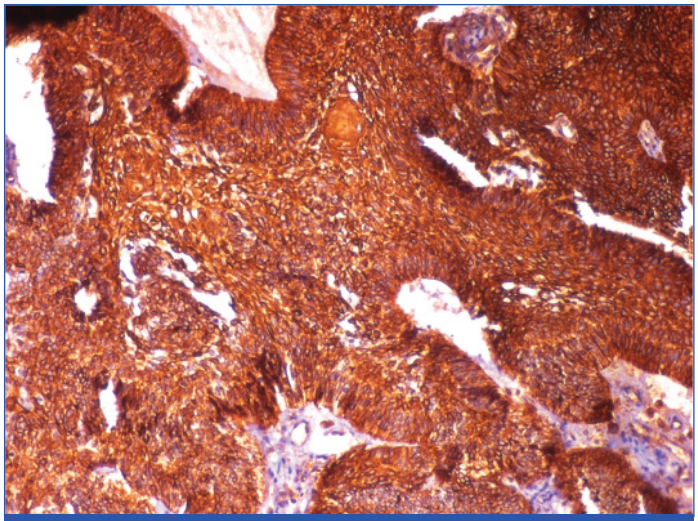
[Table/Fig-1]: Diffuse swelling in the middle third of the face. **[Table/Fig-2]:** Erythaematous and ulcerated intraoral swelling. **[Table/Fig-3]:** OPG with presence of multilocular radiolucency extending from distal aspect of mandibular first molar to ramus of the mandible on the right side. **[Table/Fig-4]:** CT scan showing a lobulated expansile lytic lesion involving body, ramus, and coronoid process of mandible. (Images left to right)



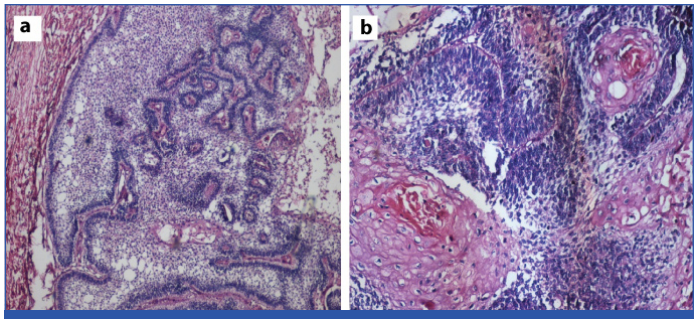
[Table/Fig-5]: Resected part of the mandible showing perforation of cortical bone.
[Table/Fig-6]: Haematoxylin and eosin stained section shows ameloblastic islands (arrow head) and area of spindle cell differentiation (arrow) (4X magnification).



[Table/Fig-8]: a) Sheets of spindle cells with cellular atypia and nuclear hyperchromatism; b) Atypical spindle shaped cells (arrow).



[Table/Fig-9]: Immunohistochemical staining positive with cytokeratin-19 for ameloblastic islands and spindle shaped cells.



[Table/Fig-7]: a) Hypercellular epithelial islands enclosing stellate reticulum (haematoxylin and eosin stain,10X magnification); b) Basal cell hyperplasia wiith keratin pearl formation in stellate reticulum like area (haematoxylin and eosin stain, 40X magnification).

unusual locations reported are maxilla, nasal cavity and anterior skull. The age of occurrence of SpAC is third to seventh decade [Table/Fig-10] [3,7-16]. Cases reported by Lau SK et al., and Ismail SB et al., were seen in 23 and 21-year-old individuals respectively [9,11]. In the present case a 32-years-old female presented with swelling of the right mandible associated with pain, difficulty in mastication and mouth opening, the cervical lymph nodes were palpable and non tender.

AC shows multilocular radiolucency with ill-defined borders and perforation of cortical bone, invasion into the soft tissue, a feature that is not commonly seen in an ameloblastoma. In most cases, radiographs show ill-defined radiolucency, focal areas of radiopacity can be seen in radiolucent area [1]. In the present case OPG showed multilocular radiolucency involving right side of the mandible extending upto the retro molar area. The second molar was missing, possibly it was extracted during the treatment of ameloblastoma.

Histologically, ACs have characters of both a benign ameloblastoma and carcinoma. The growth pattern can be follicular or plexiform, or combination of both. The peripheral tall columnar cells are arranged in a palisaded pattern, reversal of nuclear polarity and hyperchromatism may be evident. Basaloid cells with predominant nuclear hyperchromatism may dominate in the center most areas of the tumour islands, loss of ameloblastic differentiation, spindling, more than two mitotic figures per high-power field can be seen [1]. The cases reported by Jindal C et al., and Kamath VV et al., showed spindle cell associated with cystic epithelium [12,13].

Author	Age/Gender	Location	Treatment	Recurrence/Follow up
Slootweg PJ et al., [3]	75/Male	Mandible	Chemotherapy and radiation	Dead, one year after treatment
Nagai N et al., [7]	50/Male	Mandible	Surgery	NA
Infante-Cossio P et al., [8]	69/Female	Maxilla	Surgery and radiation	Alive, five years after surgery
Lau SK et al., [9]	23/Male	Mandible	Surgery	Alive, five years after surgery
Kawauchi S et al., [10]	67/Male	Mandible	Surgery	Dead, one year and 10 months after surgery
Ismail SB et al., [11]	21/Female	Mandible	Surgery with resection of lymph node	Absent
Jindal C et al., [12]	60/Female	Mandible	Surgery	Absent
Kamath VV et al., [13]	75/Male	Mandible	Surgery	Patient died two weeks after surgery
Ansari HA et al., [14]	60/Male	Maxilla	Surgery followed by radiotherapy	NA
Mc Naught MJ et al., [15]	53/Male	Mandible	Surgery	Absent/Alive
Matsushita Y et al., [16]	69/Male	Mandible	Surgery followed by radiotherapy	Absent/Alive 23 months of follow up
Present report	35/Female	Mandible	Surgery	Absent/Alive 14 months follow up

[Table/Fig-10]: Cases reported of SpAC.

time. De-differentiation can be spontaneous or related with surgical procedures or radiaotherapy: 2) Less-differentiated (primary) variant is comprised of histologic features of an ameloblastoma associated with less differentiated areas. As the patient was surgically treated for ameloblastoma two years back this case can be considered as de-differentiated or secondary in origin [7].

AC is commonly seen in the posterior part of the mandible, the

Ansari HA et al., [14] reported a case of SpAC with presence of nests of tumour cells arranged in cribriform pattern with luminal eosinophilic hyaline-like material, which resembled adenoid cystic carcinoma.

In the present case, histopathologic examination showed few characteristic ameloblastic follicles. Stellate reticulum like area showed squamous differentiation and keratin pearl formation.

Numerous areas of necrosis were present, pseudosarcomatous areas were predominated by sheets of spindle cells. The spindle cells showed nuclear hyperchromatism and pleomorphism, scattered mitotic figures, two to five mitotic figures were present in the high power field. IHC stain showed positivity for cytokeratin 19 in the epithelial islands and in the spindle shaped cell area; positive reaction with cytokeratin 19 confirmed the epithelial origin of the spindle cells. Similar IHC findings were seen in cases reported by Kawauchi S et al., [10] and Ismail SB et al., [11]. Thus, it was diagnosed as SpAC.

Resection with complete removal of the tumour is the treatment of choice. Radiotherapy with regular follow up is advised when the resection is impossible. Prognosis is very poor and metastasis can occur to lungs, few years after the treatment [1,3]. It should be treated aggressively as early as possible with regular follow up. In the present case right hemi-mandibulectomy was done with complete excision of lesion and the patient was followed for 14 months and periodic follow up was advised.

CONCLUSION

SpAC is a rare odontogenic epithelial neoplasm of the jaw that shows combination of histological features of an ameloblastoma with features of cytological atypia, with a poor prognosis. Due to lack of literature, the biological behavior, recurrence and metastatatic nature of this lesion is not known. This case report described a SpAC of mandible that originated from a pre-existing ameloblastoma with prompt treatment and regular follow up.

REFERENCES

[1] Barnes L, Eveson JW, Reichart P, Sidransky D, editors. World Health Organization Classification of Tumours. Pathology and Genetics of Head and Neck Tumours. IARC Press; Lyon: 2005. Odontogenic carcinomas; pp. 287–93.

[2] Elzay RP. Primary intraosseous carcinoma of the jaws. Review and update of odontogenic carcinomas. Oral Surg Oral Med Oral Pathol. 1982;54:299–303.

[3] Slootweg PJ, Muller H. Malignant ameloblastoma or ameloblastic carcinoma. Oral Surg Oral Med Oral Pathol. 1984;57:168–76.

[4] Corio RL, Goldblatt LI, Edwards PA, Hartman KS. Ameloblastic carcinoma: A clinicopathologic study and assessment of eight cases. Oral Surg Oral Med Oral Pathol. 1987;64:570–76.

[5] Yoon HJ, Hong SP, Lee JI, Lee SS, Hong SD. Ameloblastic Carcinoma: An analysis of 6 cases with review of the literature. Oral Surg Oral Med Oral Pathol Oral Radiol Endod. 2009;108:904–13.

[6] Yoshioka Y, Toratani S, Ogawa I, Okamoto T. Ameloblastic carcinoma, secondary type, of the mandible: A case report. J Oral Maxillofac Surg. 2013;71:58–62.

[7] Nagai N, Takeshita N, Nagatsuka H, Inoue M, Nishijima K, Nojima T, et al. Ameloblastic carcinoma: Case report and review. J Oral Pathol Med. 1991;20:460–63.

[8] Infante-Cossio P, Hernandez-Guisado JM, Fernandez-Machin P, Garcia-Perla A, Rollon- Mayordomo A, Gutierrez-Perez JL. Ameloblastic carcinoma of the maxilla: A report of 3 cases. J Craniomaxillofac Surg. 1998;26:159-62.

[9] Lau SK, Tideman H, Wu PC. Ameloblastic carcinoma of the jaws. A report of 2 cases. Oral Surg Oral Med Oral Pathol Oral Radiol Endod. 2004;85:78–81.

[10] Kawauchi S, Hayatsu Y, Takahashi M, Furuya T, Oga A, Niwa S, et al. Spindle-cell ameloblastic carcinoma: A case report with immunohistochemical, ultrastructural, and comparative genomic hybridization analyses. Oncol Rep. 2003;10:31–34.

[11] Ismail SB, Zain RB, Yaacob HB, Abraham MT. Ameloblastic carcinoma (spindle cell variant). Pathology. 2009;41:292–95.

[12] Jindal C, Palaskar S, Kaur H, Shankari M. Low-grade spindle-cell ameloblastic carcinoma: Report of an unusual case with immunohistochemical findings and review of the literature. Curr Oncol. 2010;17:52–57.

[13] Kamath VV, Satelur K, Yerlagudda K. Spindle cell variant of ameloblastic carcinoma arising from an unicystic amelobastoma: Report of a rare case. Dent Res J. 2012;9:328-33.

[14] Ansari HA, Ray PS, Khan N, Khan AH. Spindle-cell ameloblastic carcinoma of the maxilla with adenoid cystic carcinoma - like areas: A new variant? Indian J Pathol Microbiol. 2015;58:513-15.

[15] McNaught MJ, Turella SJ, Fallah DM, Demsar WJ. Spindle cell variant of ameloblastic carcinoma: A case report and review of literature. Mil Med. 2015;180(5):614-17.

[16] Matsushita Y, Fujita S, Yanamoto S, Yamada S, Rokutanda S, Yamashita K, et al. Spindle cell variant of ameloblastic carcinoma: A case report and literature review. Oral Surg Oral Med Oral Pathol Oral Radiol. 2016;121(3):54-61.

PARTICULARS OF CONTRIBUTORS:

1. Senior Lecturer, Department of Oral and Maxillofacial Pathology, St. Joseph Dental College, Eluru, Andhra Pradesh, India.
2. Postgraduate Student, Department of Oral and Maxillofacial Pathology, St. Joseph Dental College, Eluru, Andhra Pradesh, India.
3. Professor, Department of Oral and Maxillofacial Pathology, St. Joseph Dental College, Eluru, Andhra Pradesh, India.
4. Senior Lecturer, Department of Oral and Maxillofacial Pathology, St. Joseph Dental College, Eluru, Andhra Pradesh, India.
5. Senior Lecturer, Department of Oral and Maxillofacial Pathology, St. Joseph Dental College, Eluru, Andhra Pradesh, India.

NAME, ADDRESS, E-MAIL ID OF THE CORRESPONDING AUTHOR:

Dr. Mohammad Asif Kiresur,
Senior Lecturer, Department of Oral and Maxillofacial Pathology, St. Joseph Dental College, Eluru-534004, Andhra Pradesh, India.
E-mail: mdasifkiresur7@gmail.com

FINANCIAL OR OTHER COMPETING INTERESTS: None.

Date of Submission: **Aug 30, 2016**
Date of Peer Review: **Oct 04, 2016**
Date of Acceptance: **Nov 03, 2016**
Date of Publishing: **Jan 01, 2017**