

Granular cell ameloblastoma – A rare entity with recurrence after 48 years

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Abstract

A 64-year-old female with complaints of swelling right preauricular region was referred to our tertiary cancer center with fine-needle aspiration cytology (FNAC) report of mucoepidermoid carcinoma and radiological differential of malignant salivary gland neoplasm and sarcoma. On examination, there was a mass over her right parotid region. Clinical diagnosis was malignant salivary gland neoplasm. Slide review of FNAC was inconclusive. Biopsy was done. Histopathology showed neoplasm comprising of nests of cells with abundant granular eosinophilic cytoplasm with focal area showing peripherally arranged columnar cells with palisading. On enquiry, the patient gave a history of surgery of right mandible 48 years back. Correlating histopathology and clinical history, a diagnosis of granular cell ameloblastoma was rendered. Radiological evaluation showed a solid-cystic lesion in the right masticator space. Right mandible showed only part of head of mandible consistent with previous surgery. Radical surgery was done. Final report confirmed the biopsy diagnosis.

Keywords: Diagnostic challenge, granular cell ameloblastoma, late recurrence

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INTRODUCTION

Ameloblastoma is a benign progressively growing, locally aggressive intra-osseous epithelial odontogenic neoplasm characterized by expansion and tendency for local recurrence if not removed with an adequate surgical margin, beyond the radiological margin.^[1] Ameloblastoma is the second most common odontogenic tumor after odontoma (11%) and is commonly found in the lower jaw, near the angle of the mandible. It occurs most often between the third and fifth decades of life.^[2] According to clinical and radiographic features, ameloblastomas are classified into conventional solid or multicystic, extraosseous or peripheral, desmoplastic or unicystic

types. Histopathological types include follicular, plexiform, acanthomatous, granular, basaloid and desmoplastic types.

Follicular type is the most common and has a distinctive microscopic appearance characterized by the presence of peripheral columnar cells with hyperchromatic, reversely polarized nuclei, arranged in a palisaded pattern with central loose connective tissue resembling stellate reticulum. Granular cell ameloblastoma (GCA) is a rare histological subtype of ameloblastoma accounting 3%–5% of the total ameloblastomas.^[3] It is characterized by granular changes in stellate-like cells located within the inner portion of the epithelial follicles.

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CASE REPORT

A 64-year-old female was referred to the surgery outpatient wing of our tertiary cancer center with an outside fine-needle aspiration cytology (FNAC) report of mucoepidermoid carcinoma. She complained of swelling and pain on the right side of her face of 6 months duration. On examination, there was a large mass lesion over her right preauricular region measuring 7 cm × 6 cm. There were no enlarged lymph nodes or any other significant clinical findings. A magnetic resonance imaging (MRI) study done from outside center showed a multiloculated solid-cystic mass involving right masticator space and prestyloid and parapharyngeal space. The possibilities considered radiologically were malignant salivary gland neoplasm or malignant peripheral nerve sheath tumor. Slide review of FNAC at our center was not in agreement with outside report of mucoepidermoid carcinoma. We found the smears to be inadequate with few cells with foamy cytoplasm macrophages. With a provisional clinical diagnosis of malignant salivary gland neoplasm, radiological evaluation and biopsy was planned at our center.

Histopathology from biopsy specimen showed a neoplasm comprising of nests of cells with abundant granular eosinophilic cytoplasm with peripherally placed compressed pyknotic nucleus. In focal areas at the periphery of the nests were seen columnar cells with palisading and reverse polarity [Figure 1]. In view of the abundant granular cells, S100 and cytokeratin (CK AE1/AE3) immunostains were done to rule out possibility of granular cell tumor. S100 immunohistochemical stain showed negative staining. CK (AE1/AE3) was diffuse strong positive. On enquiry, the patient gave a history of surgery of her right mandible

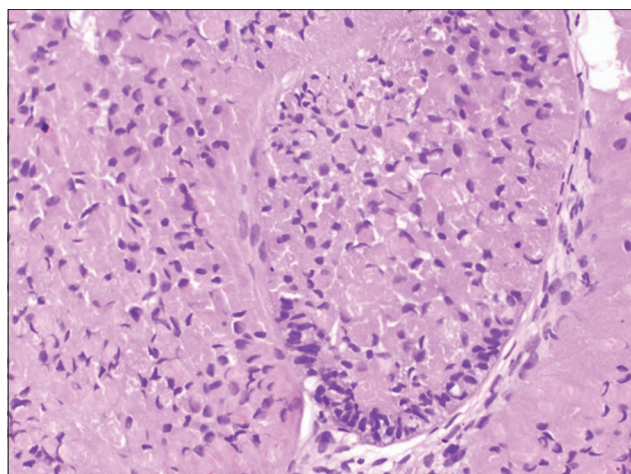


Figure 1: Neoplasm comprising of nests of cells with abundant granular eosinophilic cytoplasm and peripherally placed nucleus. In focal areas at the periphery of the nests were seen columnar cells with palisading and reverse polarity (H&E, ×200)

48 years back following a tooth extraction. Details were not available. Correlating histopathology and clinical history a diagnosis of GCA was offered. Computed tomography and MRI study at our center showed a solid-cystic lesion measuring 6.2 cm × 6 cm × 4.9 cm centered in the right masticator space with compression and displacement of parapharyngeal space and doubtful infiltration of parotid. Right mandible showed only part of head of mandible, rest of coronoid and conoid process was not seen which was consistent with previous surgery [Figure 2]. The patient was taken up for radical surgery.

Peroperatively, there was a hard encapsulated mass measuring 6 cm × 7 cm involving the right infra temporal fossa, right parapharyngeal space and right temporal fossa. Mass was located medial to the parotid. Mass was excised in toto and parotid preserved. The gross specimen was a solid cystic mass measuring 10 cm × 6 cm × 5 cm. Cut section gray-white, solid-cystic with cystic area showing internal septations [Figure 3]. Histopathology showed a neoplasm composed predominantly of nests of large granular cells with abundant eosinophilic granular cytoplasm and peripherally compressed nucleus. The periphery of these nests showed columnar cells with hyper chromatic, reversely polarized nuclei typical of odontogenic epithelium, arranged in a palisaded pattern [Figure 4a and b]. Also were seen focal areas showing more common pattern of ameloblastoma: The plexiform type of pattern composed of anastomosing strands of ameloblastomatous epithelium with an inconspicuous central stellate reticulum and cyst like stromal degeneration. Correlating clinical history, radiological details and histopathological findings, a diagnosis of GCA which was in concordance with the biopsy report was reached.



Figure 2: Computed tomography showing solid-cystic lesion (asterisk) measuring 6.2 cm × 6 cm × 4.9 cm centered in the right masticator space. Right mandible ramus not visualized

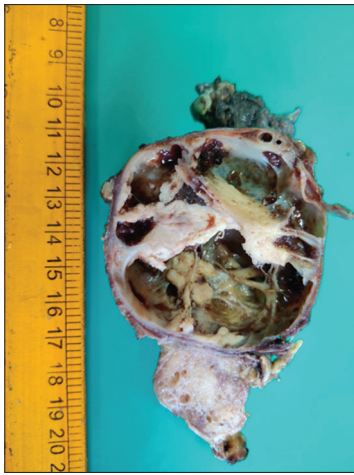


Figure 3: Solid cystic mass measuring 10 cm × 6 cm × 5 cm. Cut section gray-white with cystic area showing internal septations

DISCUSSION

Ameloblastoma with granular cell pattern is an unusual histological variant of the odontogenic tumor ameloblastoma. Focal areas of granular cell change can occur in other common histological types of ameloblastoma such as follicular or plexiform types. However, to categorize as granular cell variant, there should be extensive granular cell change which is a very rare finding. The central stellate reticulum like cells show extensive granular cell change with peripherally located pyknotic nucleus. The “granular change” is reported to happen due to a dysfunctional status of neoplastic cells, and the pathogenesis of this tumor seems to be age-related.^[4,5] However, whether the granular cell change in ameloblastoma is a degenerative process or a harbinger of a more aggressive course remains unclear.^[6]

Granular cell change in classic ameloblastoma is a well-recognized phenomenon. Ever since granular cell change has been noted in ameloblastoma, there has been curiosity to establish the nature of these granular cells. The granular cells acquire peripherally pushed nuclei and abundant cytoplasm filled with coarse eosinophilic granules indicating that there is an apoptotic process taking place. Several immunohistochemical studies in the literature showed increased apoptotic cells and decreased expression of antiapoptotic factors such as Bcl-2 and p53 proteins in GCA.^[7] The granular appearance has been ascribed to numerous lysosomes based on histochemical and electron microscopic findings. It is speculated that unnecessary or aged components in the cytoplasm of some tumor cells accumulate with age; however, the ability of lysosomes to digest these materials decreases with age, and as a result, the cytoplasm of some tumor cells become packed with lysosomal granules.^[8]

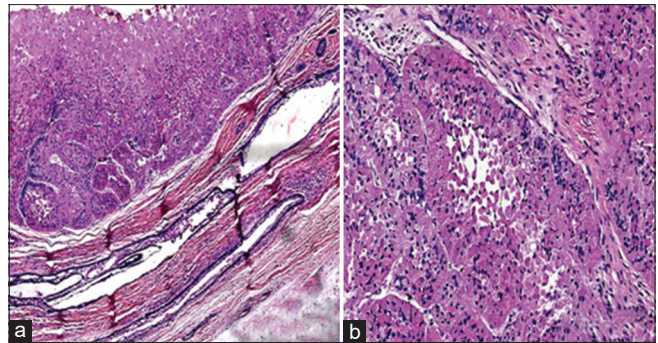


Figure 4: (a) Neoplasm composed of nests of granular cells with abundant eosinophilic granular cytoplasm and peripherally compressed nucleus. Periphery showing the plexiform pattern of ameloblastoma (H&E, ×100). (b) The periphery of these nests of granular cells showed columnar cells with hyperchromatic, reversely polarized nuclei, arranged in a palisaded pattern (H&E, ×200)

The biological behavior of GCA does not seem to differ from the other histologic subtypes of ameloblastoma if diagnosed early. However, there are reports in the literature of GCA being locally aggressive and recurrent. Reichart *et al.* reported a 33.3% recurrence rate for this lesion, which was higher than the more common follicular, plexiform and acanthomatous subtypes.^[9] To recognize this, rare histological type especially in biopsies is a challenge. This is because unlike other common types the ameloblastomatous epithelium is barely identifiable amidst the sheets of granular cells.

GCA should also be differentiated from other oral lesions with granular cells namely granular cell odontogenic tumor, granular cell tumor and congenital epulis.^[10] Although the morphology of all granular cells is similar, the tissue of origin is particular to each condition. Clinical radiological and histopathological details are essential to reach a correct diagnosis. Furthermore, importantly, these lesions present different biological pathways and should be discriminated from GCA.

CONCLUSION

GCA is a rare histological type of ameloblastoma as compared to more common types such as follicular, plexiform and acanthomatous types. This case is being reported to highlight the diagnostic difficulty, especially in biopsies due to the minimal ameloblastomatous epithelium seen. Furthermore, although ameloblastomas are known to recur, especially after incomplete excisions our case has reported 48 years after her initial surgery. To our knowledge, recurrence after this long a period has not been reported in the literature. Regular follow-ups are required to pick up recurrences as early as possible.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient (s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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Nil.

Conflicts of interest

There are no conflicts of interest.

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