Round Blue Cell Tumor arising in the Background of Ameloblastic Fibrosarcoma

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ABSTRACT
World Health Organization (WHO) blue book of head and neck tumors reports 64 cases of ameloblastic fibrosarcoma and 14 cases of ameloblastic fibrodentinosarcoma/fibro-odontosarcoma. We present a unique case that does not fit into any of the WHO classified odontogenic sarcomas. The patient presented in 2006 with a densely sclerotic lesion arising from the left sphenoid bone, displacing the orbital contents and extending up to the inferior margin of the maxillary sinus. He was treated with left maxillectomy and orbital exenteration. He relapsed 6 years later, salvage surgery was unsuccessful and subsequent care was supportive. Histologically the malignant neoplasm showed follicles comprising of peripheral columnar cells lined up in palisaded fashion with hyperchromatic nuclei displaced away from basement membrane. Center of the follicles comprised of spindle-shaped cells resembling stellate reticulum. Intervening areas showed fibrosarcomatous areas. Other areas revealed predominant sheets of round cells having hyperchromatic nuclei, inconspicuous nucleoli and scant cytoplasm. These round cells revealed strong membranous positivity for CD99.

Keywords: Ameloblastic carcinoma, Round blue cell tumor, Head and neck.


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INTRODUCTION
Malignant odontogenic tumors are generally rare. Odontogenic sarcomas comprise of ameloblastic fibrosarcoma, ameloblastic fibrodentinosarcoma and ameloblastic fibro-odontosarcoma.1 WHO blue book of head and neck tumors reports 64 cases of ameloblastic fibrosarcoma and 14 cases of ameloblastic fibrodentinosarcoma/fibro-odontosarcoma.2 Ameloblastic fibrosarcoma has locally aggressive behavior with extremely low potential for distant metastasis. Ameloblastic fibrodentinosarcoma/fibro-odontosarcoma has better prognosis as compared to other jaw sarcomas.3 We present a unique case that does not fit into any of the WHO classified odontogenic sarcomas and label it as round blue cell tumor arising in the background of ameloblastic fibrosarcoma.

CASE REPORT
A 30 years old male presented with a 1 year history of left-sided facial swelling in 2006. Computed tomographic (CT) scan showed a densely sclerotic lesion arising from the left sphenoid bone, displacing the orbital contents and extending up to the inferior margin of the maxillary sinus (Fig. 1). Patient underwent left maxillectomy and orbital exenteration. The pathology specimen was oriented with single stitch on superior margin, double stitches on medial margin, triple stitches on optic nerve resection margin, four stitches on temporalis margin and five stitches on zygomatic arch margin. The whole of the specimen measured 8 × 5 × 3 cm. Serial sectioning revealed a well- circumscribed tumor measuring 6 × 4 × 3 cm located at a distance of 4 cm from superior margin, 0.2 cm from inferior margin, 1.5 cm from medial margin, 0.5 cm from lateral margin, 0.3 cm from posterior margin and 2 cm each from temporalis and zygomatic resection margins. Eyeball was not involved by the tumor. Cut surface of the tumor was gray white, firm and multilobulated. Histological sections revealed a malignant neoplasm showing follicles comprising peripheral columnar cells lined up in palisaded fashion. Nuclei were hyperchromatic and displaced away from basement membrane. Center of the follicles comprised of spindle-shaped cells resembling stellate reticulum (Fig. 2). Intervening areas showed fibrosarcomatous areas comprising proliferated spindle cells showing moderate atypia and atypical mitoses (Fig. 3). Other areas revealed predominant sheets of round cells having hyperchromatic nuclei, inconspicuous nucleoli and scant cytoplasm (Fig. 4). These round cells revealed strong membranous positivity for CD 99 (Fig. 5). We labeled this case as round blue cell tumor arising in the background of ameloblastic fibrosarcoma.
arising in the background of ameloblastic fibrosarcoma. Postoperative CT scan did not show any residual tumor and radiation therapy 60 Gy in 30 fractions were delivered to the operative bed. He remained symptom free for 6 years and re-presented with a large recurrent mass involving ethmoid sinuses. Biopsy of the recurrent lesion again revealed round blue cell tumor arising in the background of ameloblastic fibrosarcoma.

**DISCUSSION**

Odontogenic sarcomas are classified as ameloblastic fibrosarcomas and ameloblastic fibrodentinosarcoma and fibro-odontosarcomas. They usually occur in a wide age range (3-89 years) with median age in the third decade. Most of ameloblastic fibrosarcomas occur in mandible. Usually these sarcomas are locally aggressive. These have better prognosis than other jaw sarcomas and are generally regarded as low grade sarcomas. Ameloblastic sarcoma was first reported by Heath in 1887. It has then been reported by many authors over a period of many years from all around the world. Previously ameloblastic sarcoma had been used as a general term to include all types of odontogenic sarcomas. Later on it was further classified as ameloblastic fibrosarcomas and ameloblastic fibrodentino- and fibro-odontosarcomas. This segregation was based on the presence or absence of dentin or dentin or enamel in the lesion. Ameloblastic fibrosarcoma has been reported by Chromette in 1982, Goldstein G in 1976, Pinborg in 1960, Kousar in 2009, Demoor-Goldschmidt in 2012 and many other authors. About 14 cases of fibrodentino- and odontosarcomas have been reported. In majority of the cases reported patients were middle aged and most common location was mandible followed by maxilla. Most of the cases showed locally aggressive behavior. Surgical treatment was curative in majority of the cases. Only one case showed metastasis to mediastinal nodes.

**Fig. 2:** Follicles comprising peripheral columnar cells lined up in palisaded fashion. Center of the follicles comprised of spindle-shaped cells resembling stellate reticulum

**Fig. 3:** Fibrosarcomatous areas comprising proliferated spindle cells showing moderate atypia and atypical mitoses

**Fig. 4:** Predominant sheets of round cells having hyperchromatic nuclei, inconspicuous nucleoli and scant cytoplasm

**Fig. 5:** Round cells revealed strong membranous positivity for CD99
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Our case is unique due to the fact that it involved maxilla and it was different from ameloblastic fibrosarcomas and ameloblastic fibrodentinosarcoma fibro-odonto- sarcomas. This tumor had a component of ameloblastoma and the other components comprising fibrosarcoma and round blue cell tumor component. The latter component had cells with increased nuclear to cytoplasmic ratio, hyperchromatic nuclei and frequent atypical mitoses. These cells were positive for CD99. This tumor behaved aggressively. It recurred after a period of 6 years despite complete surgical resection and radiotherapy. We labeled our case as round blue cell tumor arising in a background of ameloblastic fibrosarcoma. We have not been able to find such a case in the literature.

Usually, odontogenic tumors are low grade sarcomas but if round blue cell component occurs in background of ameloblastoma it can behave in an aggressive fashion.

REFERENCES


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