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Extraskeletal myxoid chondrosarcoma of maxilla: A rare entity

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CASE REPORT

A 45-year-old male patient reported with a slow growing swelling on the left side of the palate for 3 months. On examination, proliferative growth was seen on the left posterior palatal gingival region extending from the left second premolar to second molar. The growth was 2 cm × 1.5 cm × 0.5 cm in the greatest dimension, soft in consistency with irregular borders and was pinkish in color. The growth was painless and nontender. Orthopantomograph showed unilocular radiolucency with diffused borders in the same region. Lymph nodes were tender. The patient had the habit of tobacco chewing since last 6 years. Medical history was not contributory. Clinically, it was suspected to be a benign lesion. Incisional biopsy was done and the lesion was reported as pyogenic granuloma. On excisional biopsy, the lesion was diagnosed as extraskeletal myxoid chondrosarcoma (ESMC).

Histopathology

Under scanner view, the tumor showed multiple lobular patterns separated by dense fibrocellular septae [Figure 1]. On higher magnification, the tumor cells were seen arranged at the periphery of the lobules assuming a radial pattern of columns, cords or strands [Figure 2]. The tumor cells were polygonal, spindle- or stellate-shaped and had round to oval, bland nuclei and scant to moderate amount of deeply eosinophilic cytoplasm [Figure 2]. In the center of the lobules, abundant myxoid stroma was seen [Figure 2]. In addition, the cells in the center were loosely arranged with some forming small nests or clusters resembling chondroblasts [Figure 3]. These cells were periodic acid–Schiff [Figure 4] and vimentin [Figure 5a] positive and negative for pan-cytokeratin [Figure 5b], S-100 [Figure 5c] and epithelial membrane antigen (EMA) [Figure 5d]. The myxoid stroma was strongly positive for alcian blue [Figure 6] and mucicarmine stains [Figure 7].

DISCUSSION

ESMC is chondroblastic in origin and relatively a rare tumor. ESMC accounts for 6–12% of all chondrosarcomas.[1] It is predominantly a tumor of the extremities, with the head and neck region involved in only approximately 5% of the cases.[2] Age of occurrence ranges from 1 to 92 years with the mean age of 45 years. Men are more commonly affected than women.[3] Clinical signs and symptoms are mostly nonspecific, as reported in our case which was painless and nontender. However, some cases have been reported with painful slow growing mass. Radiography, computed tomography scans and magnetic resonance imaging show a soft tissue mass with no distinctive radiologic feature that can differentiate it from other soft tissue sarcomas.[2,3] Histopathological findings and staining characteristics makes this tumor distinct from other sarcomas. Microscopically, a characteristic multilobular pattern is clearly evident. The individual lobules are separated by fibrovascular septae and consist of round and slightly elongated cells of uniform shape and size separated by variable amount of myxoid material. Individual cells have small darkly-stained nuclei and a rim of deeply eosinophilic cytoplasm, features characteristic of chondroblasts. Occasionally, cells are vacuolated. Typical chondrocytes within lacunae are rare, as seen more commonly in skeletal chondrosarcomas. Mitotic figures are rare in ESMC, but commonly observed in skeletal chondrosarcomas.[4] The individual cells are arranged in cords, strands or pseudoacini, creating a lace-like appearance. In ESMC, extracellular myxomatous material stains deeply

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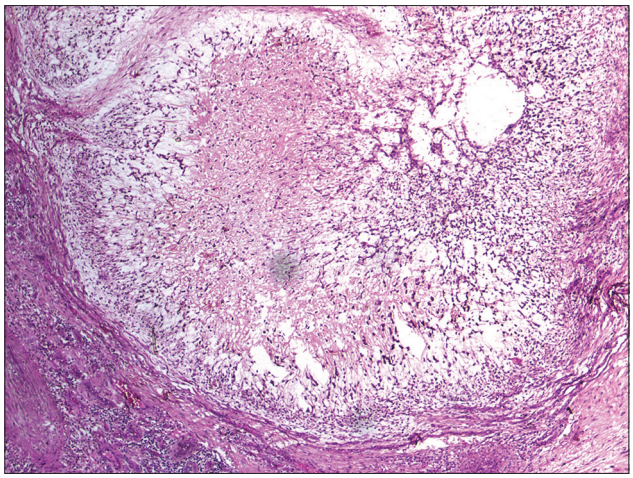

Figure 1: Lobular structure of the tumor cells. (H&E stain, x40)

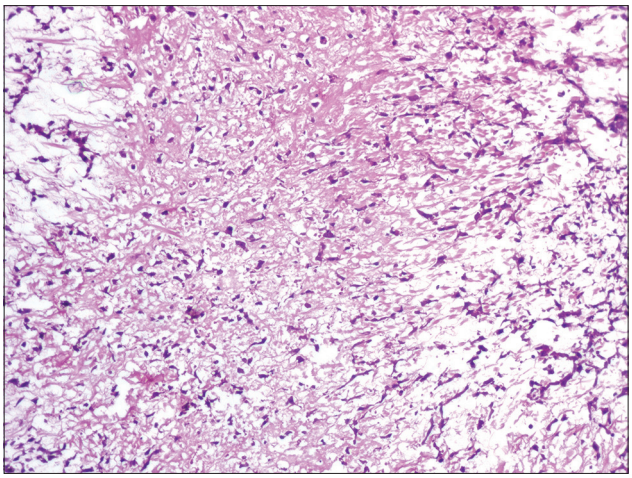

Figure 2: Myxoid areas and tumor cells resembling chondroblasts (H&E stain, ×100)

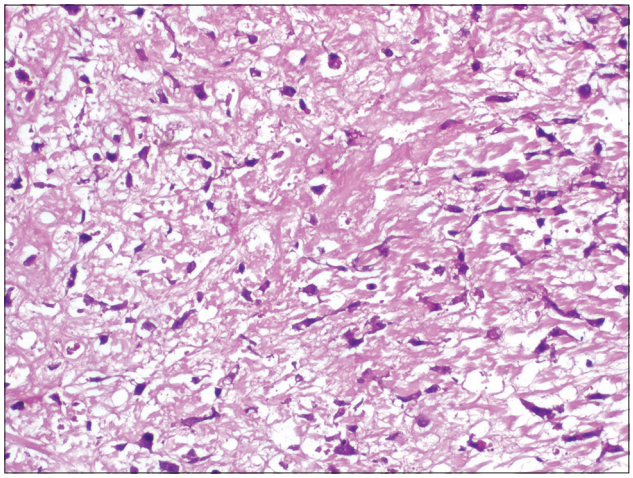

Figure 3: High power view showing loosely arranged tumor cells resembling chondroblasts (H&E stain, ×400)

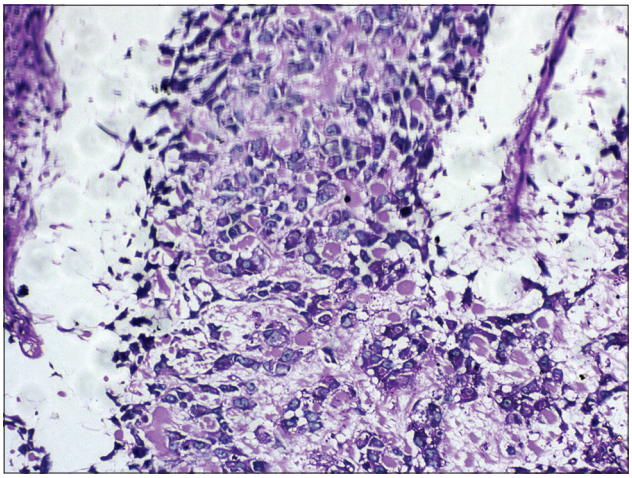

Figure 4: Tumor cells positive for intracytoplasmic glycogen (PAS stain, ×200)

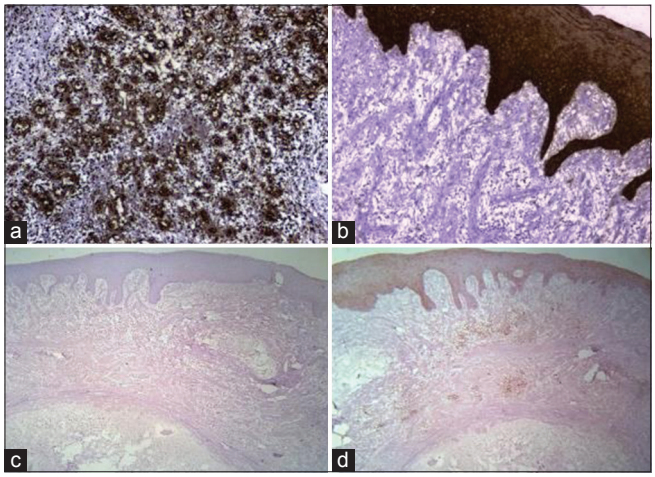

Figure 5: Immunohistochemical analysis of extraskeletal myxoid chondrosarcoma (a) tumor cells strongly reactive to vimentin (IHC stain, ×200) (b) tumor cells nonreactive to pan-cytokeratin (IHC stain, ×200) (c) tumor cells nonreactive to S-100 (IHC stain, ×100) (d) tumor cells nonreactive to epithelial membrane antigen (IHC stain, ×100)

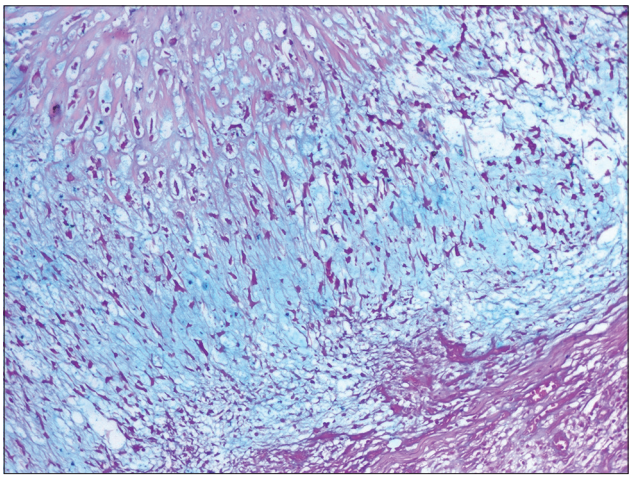

Figure 6: Myxoid matrix positive for mucin (alcian blue stain, ×100)

with colloidal iron, mucicarmine and alcian blue stains.[5] Immunohistochemical (IHC) analysis shows that this tumor

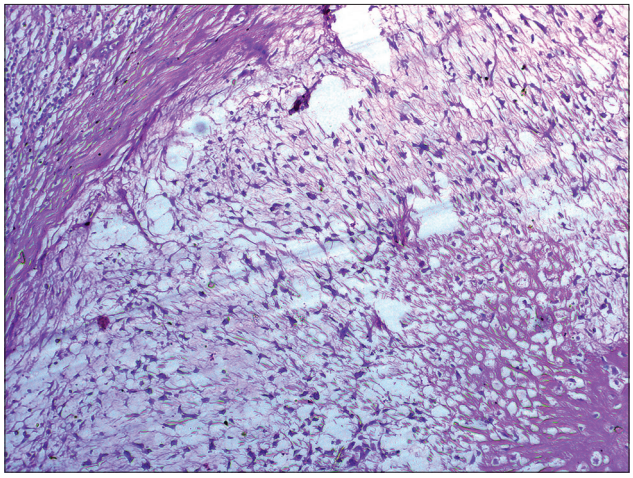

Figure 7: Myxoid matrix positive for mucin (mucicarmine stain, ×100)

is strongly reactive to vimentin.[6] As the lesion shows focal or very less amount of cartilage formation, the S-100 reactivity is focal or negative.[4] Some rare lesions can show focal reactivity toward cytokeratins, otherwise many are negative to it.[6,7] Other IHC panel of markers studied with this lesion includes SOX-9, neuron-specific enolase, chromogranin or synaptophysin.[7]

Differential diagnosis includes osteosarcoma. Diagnosis of ESMC should be made only after ruling out the presence of malignant osteoid. In the head and neck region, the most important histological differential diagnosis includes chordoma. Chordomas were initially wrongly reported as ESMCs. The cells of chordoma, in contrast to those of ESMC, are reactive for cytokeratin and EMA.

ESMC is differentiated from the classic skeletal chondrosarcomas by the following features (1) Clinically, ESMC presents with painless swelling located in soft tissue. (2) Microscopically, typical chondrocytes within the lacunae are seen in skeletal chondrosarcoma. (3) Mitotic figures are commonly seen in skeletal chondrosarcomas. (4) Cartilage formation is commonly seen in skeletal chondrosarcomas. (5) Myxoid differentiation is abundantly seen in ESMC. (6) Microscopically, multilobular pattern is characteristic for ESMC. (7) ESMC has far better prognosis than its skeletal counterpart.

In our case, the swelling was located in the palatal gingiva (soft tissue). Microscopically, the tumor showed characteristic multilobular pattern with abundant myxoid tissue, which was positive for alcian blue and mucicarmine stains. The typical chondrocytes within the lacunae were rare. Mitotic figures were also rarely seen within the tumor. The tumor was devoid of cartilage formation, as supported by its negativity toward S-100 protein. Therefore, taking into consideration, clinical features, histopathological findings, special staining and IHC analysis, the lesion was diagnosed as ESMC.

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Conflicts of interest

There are no conflicts of interest.

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