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CASE REPORT

Ectomesenchymal chondromyxoid tumor of the hard palate – a case report

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Ectomesenchymal chondromyxoid tumor (ECMT) is a rare tumor. Only 26 cases of ECMT have been reported, all occurred in the anterior tongue. We present a case of a 30-year-old male with a nodule in the hard palate, which was reported as ECMT on histopathology. The differential diagnosis considered included are extraskeletal myxoid chondroma, ECMT, pleomorphic adenoma, oral focal mucinosis, chondroid choristoma, and ossifying fibromyxoid tumor. After serial sections no other component was observed and a diagnosis of ECMT was made by exclusion. The patient is asymptomatic on follow up. A review of existing literature is also presented here.
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A 30-year-old male patient presented with history of a small, painless nodule in the hard palate for 4 years. There was no significant increase in size or any other oral complaints. There was no family or past history of a similar lesion. On examination, a 0.5 cm diameter non-tender nodule was seen located on posterior end of the hard palate in line with the retromolar trigone, away from the upper incisors. The overlying mucosa was unremarkable. The nodule was excised in toto under local anesthesia.

Gross-examination revealed a single mucosa covered soft tissue measuring $0.5 \times 0.3 \times 0.3$ cm. The hematoxylin and eosin-stained sections showed a polypoid, well-circumscribed but unencapsulated lesion (Fig. 1). The margins were not sharply demarcated from the surrounding tissue. The tumor had low cellularity and was arranged in lobules of chondromyxoid tissue (Fig. 2). The tumor was composed of two types of cells – round to polygonal cells in a chondroid background and stellate cells in a myxoid background (Fig. 2a,b). No nuclear atypia, necrosis, hemorrhage calcification or osteoid was seen. Occasional binucleate cell was seen. A linear proliferation of small-sized vessels was seen at the periphery, which was also traversing to the center of one of the lobules (Fig. 1). The overlying mucosa was unremarkable. The resected margins were free. Histological differential diagnosis (D/d) considered included are extraskeletal myxoid chondroma (ESMC), ectomesenchymal chondromyxoid tumor (ECMT),

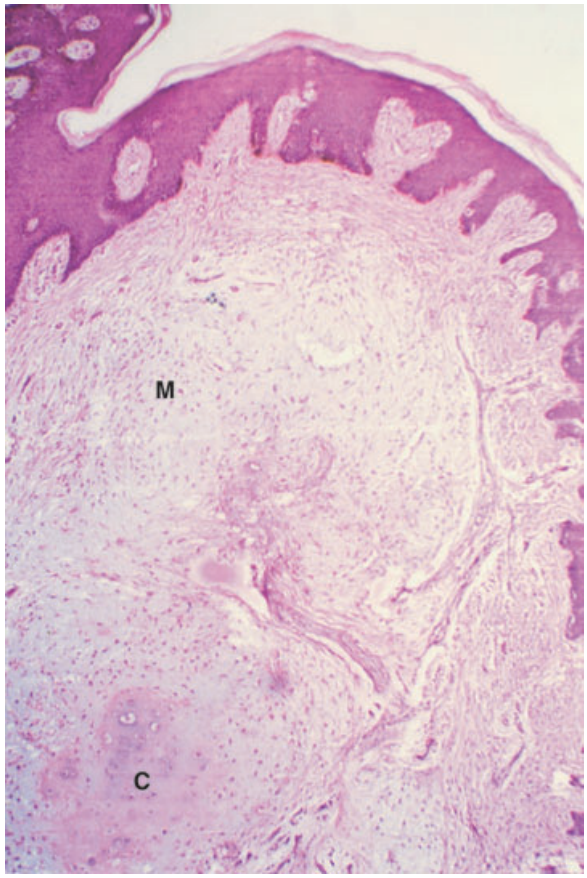

Figure 1 Tumor with lobules of chondromyxoid tissue. The cellularity is low. The chondroid tissue (C) is seen in the center, whereas myxoid (M) tissue predominates at the periphery [hematoxylin and eosin (H&E), ×100].

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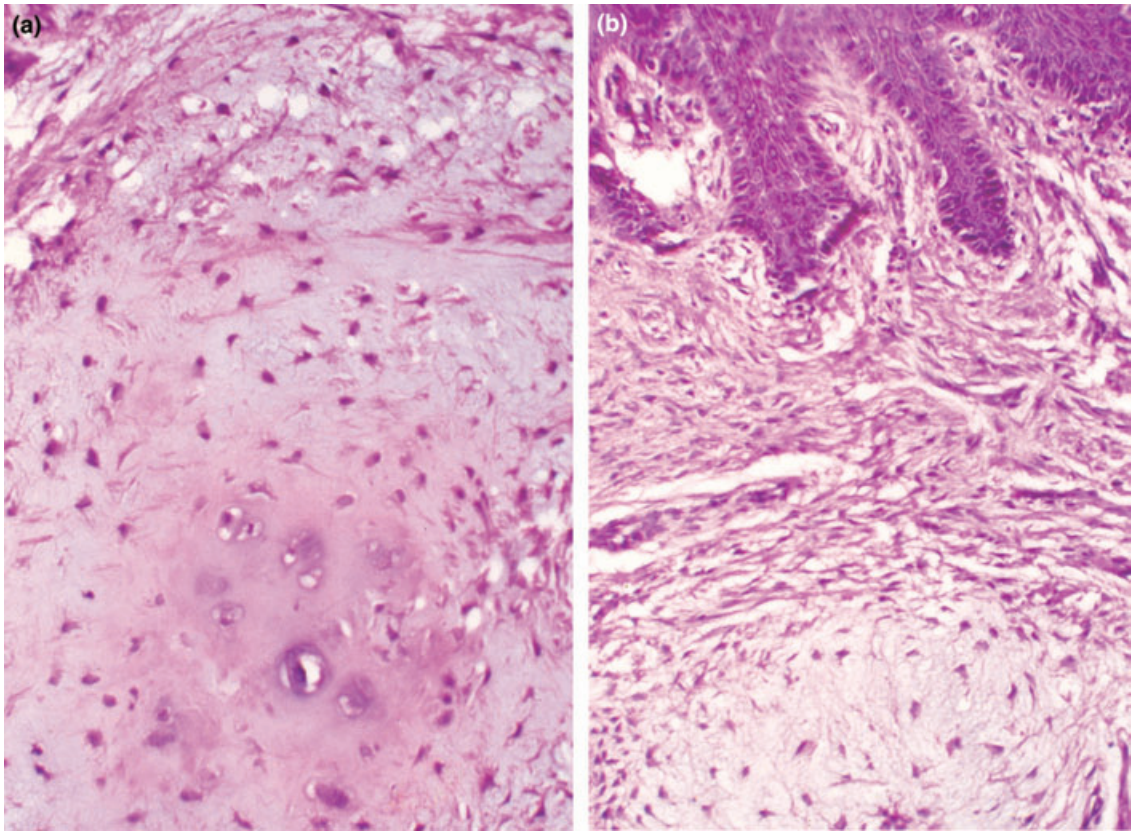

Figure 2 (a) Chondroid tissue showing benign cells [hematoxylin and eosin (H&E), ×400]. (b) Circumscribed myxoid tissue with bland stellate cells (H&E, ×400).

pleomorphic adenoma, oral focal mucinosis, chondroid choristoma, and ossifying fibromyxoid tumor. Serial sections did not reveal any other component and the diagnosis of ECMT were made by exclusion. Immunohistochemistry could not be carried out as the tumor got exhausted during the first serial sectioning.

The patient is asymptomatic and free of recurrence 2 years after surgery.

Comments

Ectomesenchymal chondromyxoid tumor is a rare tumor of the oral cavity, described recently by Smith et al. (1). Only 26 cases of ECMT have been reported so far, all of which occurred the anterior tongue (1–4). Histologically, the lesion is characterized by a well-circumscribed lobular proliferation of cells within a chondromyxoid background. The cells of ECMT have round to ovoid or fusiform nuclei, with occasional multilobated or atypical nuclei and rare nuclear inclusions, arranged in cords or sheets (1–4). We report a case of ECMT seen for the first time in hard palate.

The ECMT appears clinically as slow-growing, painless, firm, well-circumscribed small lesions, of 0.5–2 cm size (1) as also seen in our case. It has been reported over a wide age (range: 9–78 years), with no sex predilection (1–4). The duration of symptoms at the initial diagnosis is also variable, ranging from 1 month to 20 years (1). Our patient had noticed the nodule for 4 years.

The ECMT is a diagnosis of exclusion, it is very important to rule out other D/d-like ESMC, oral focal mucinosis, pleomorphic adenoma, myoepithelioma, mucocele, glial/chondroid choristoma, and ossifying fibromyxoid tumor (3). Extraskeletal myxoid chondroma is a very close D/d of ECMT. However, the ESMC usually occurs in hands or feet, and is often seen in relation to tendon, its sheath or the joint capsule (5). Histologically ESMC also has a lobular pattern but its borders are sharp. ESMC is often associated with calcification which is more prominent in the center. A granuloma-like proliferation of epitheloid cells and multinucleate giant cells may be seen in ESMC (5). These features were not seen in the present case. Further, the periphery of ESMC has more mature and less cellular cartilage as opposed to the central chondroid tissue seen in our case (5). Mucocele shows granulation tissue and extravasated mucin pools. Oral focal mucinosis is a circumscribed lesion with stellate cells in myxoid background; however, chondroid areas are absent. Myoepithelioma of minor salivary gland can show both chondroid and myxoid stroma, the lesional cells are, however spindled or plasmacytoid (3). The absence of astrocytes, ependymal and ganglion cell excludes the diagnosis of glial choristoma. Presence of myxoid areas merging with mature chondroid tissue ruled out cartilaginous

choristoma (3). Ossifying fibromyxoid tumor of soft parts would show a similar background, but presence of bone in the periphery and cells, which may be in lacunae, clinches the diagnosis (3).

The ECMT is a lesion of uncertain histogenesis. ECMT probably arises from a proliferation of uncommitted ectomesencymal cells from which the lesion derives a part of its name (1, 3). The multipotentiality of these cells is explained by the presence of both myxoid and chondroid areas. This is corroborated further by immunohistochemistry. The tumor cells express desmin and vimentin suggesting a mesenchymal origin (4). The expression of S100 and Glial fibrillary acidic protein supports a neurogenic origin (4). The tumor is negative for epithelial markers like keratins and carcinoembryonic antigen; however, Smith et al. (1) have reported variable positivity for cytokeratin (1, 4).

The small-size, long duration and well-circumscribed nature suggests a benign behavior. The reported literature has also proved it to be a benign lesion so far (1, 2, 4). However, recurrence has been observed in two patients, occurring after 2 and 19 months (1). No malignant change is reported (1). No recurrence was observed in our case on 24 months follow up. The treatment of choice is complete excision with tumor-free margins. This is usually possible easily, as these lesions are small and relatively well circumscribed.

We present this case because of its rarity. This is the first case of ECMT arising in hard palate. We advocate that a D/d of ECMT may be considered in small, painless, slow-growing lesions of hard palate. However histologically, the diagnosis should be made after ruling out other chondromyxoid lesions of oral cavity, especially of ESMC.

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