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Answer of the Last Issue's Case Question

Geçen Sayının Bilmece Olgusunun Yanıtı

An Unusual Mass of Oral Cavity: Parachordoma

Sıradışı Bir Oral Kavite Tümörü: Parakordoma

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Anahtar Sözcükler
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Parachordama represents a soft tissue tumor consisting of cells with histological and ultrastructural characteristics similar to those of chordoma cells. On the other hand, with immunohistochemistry, they have features similar to that of chondroid tumor cells.[1,2] This tumor most often arises from upper or lower limbs. Parachordomas are treated by complete removal of lesion.[3] Sometimes, these tumors are locally destructive and tend to recur if incompletely excised.[4-6] In this study, we described a parachordoma seen on the tongue. Parachordoma on the tongue or in oral cavity has not been previously reported in the medical literature.

Parachordoma was first reported by Dabska in 1977.[1] Parachordomas typically develop adjacent to tendon, synovium, or osseous structures within extremities and present as slowly growing locally destructive, lobulated neoplasms that are prone to recurrence if incompletely excised.[3] These tumors are commonly localized in soft tissue, particularly at upper or lower limbs. It might derive from an ectopic notochord, like chordomas or other neuron-related cells.[7] A role for trauma is implied in the etiopathogenesis of parachordomas.[3] The parachordoma case in the present study also reinforces the role of trauma. Generally, these tumors are considered to be a benign mass; however, they may also be locally aggressive or lead to distant metastasis.[1,4]

On macroscopic evaluation, parachordomas are frequently seen as a well-defined nodular mass, ranging from 3 to 7 cm in diameter (Figures 1,2). They have a yellow-white cross-sectional surface with myxoid or gelatinous condition.[4]

Histopathologically, parachordomas were characterized by well circumscribed lobules composed of small cellular aggregates, and single, large, vacuolated cells embedded within a hyalin and chondroid matrix divided by fibrous trabeculae. The tumor was composed of cells with clear eosinophilic cytoplasm and an epithelioid appearance in a myxoid stroma separated by fibrous tissue with mild pleomorphism and mitotic activity.[3]

Parachordomas show two distinct patterns: cohesive nests or ribbons of cells with abundant, clear cytoplasm and a high nuclear cytoplasmic ratio set in a large myxoid background (Figure 3); and chondroid areas, comprising 15-20% of the mass area, composed of clear cells set in a cartilagenous matrix that stained prominently with Alcian blue. Positive immunohistochemical staining is observed among 25% to 50% of the tumor cells.[7]

In the immunohistochemistry, parachordoma cells express cytokeratin 8/18, focal chromogranin A, epithelial membrane antigen (EMA) (Figures 4,5), S-100 protein and vimentin. However, CD 34, actin, glial fibrillary acidic protein (GFAP) and calponin are typically negative.[3,5,6,8]

Electron microscopic evaluation indicates that the cells display features of incomplete epithelial differentiation such as primitive cell-ligaments, fragmental basal lamina and microvillus apophysis.

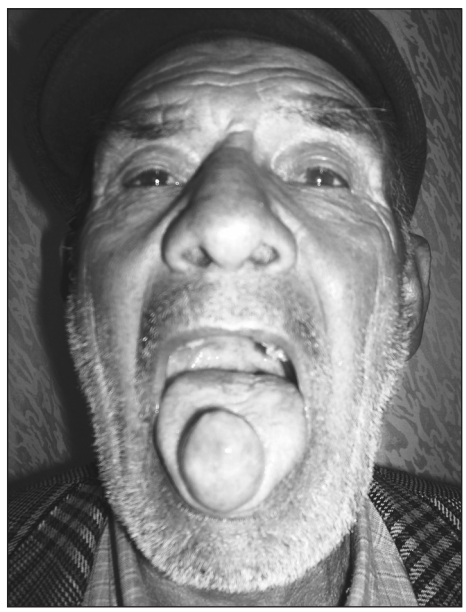

Figure 1a. Anterior view of parachordoma on the tongue.

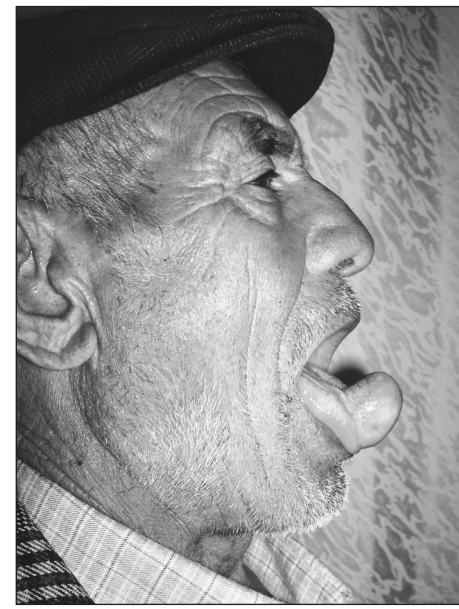

Figure 1b. Lateral view of parachordoma on the tongue.

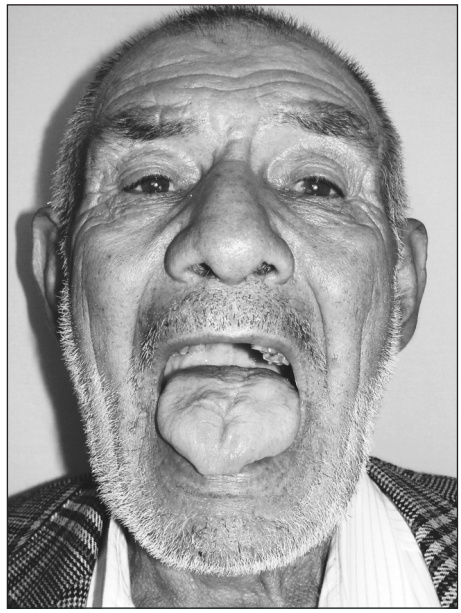

Figure 2. Postoperatif view of the tongue after six months.

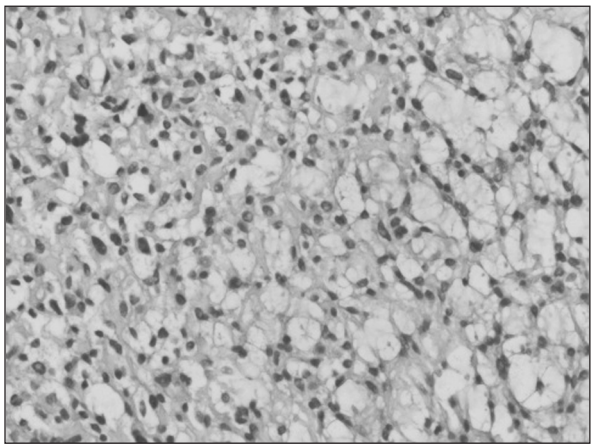

Figure 3. Parachordoma: Epithelioid tumor cells in myxoid matrix (HE, x100).

This is the first paper that presents a parachordoma arising from a tongue dorsum. Clinical differential diagnosis has to be made with chordoma, giant contact granuloma, mucus retention cysts and the other lingual lesions. The other neoplasms often considered in the diferantial diagnosis of parachordomas are extraskeletal myxoid chondrosarcomas, cartilaginous tumors in soft tissue, and myxoid liposarcomas.[3] Furthermore, pathological differential diagnosis of parachordoma has to be made with chondrosarcoma.[1] Parachordomas are less aggressive compared with chordomas. They can be totally removed in most cases, and generally do not show metastasis.[7] Therefore, the prognosis of parachordoma is better than chordoma. Conversely, chordoma's location often makes it difficult to remove totally, so its recurrence rate is high.[6] However, sometimes parachordomas are locally destructive and tend to recur if incompletely excised.[4] Karakaya et al. reported a recurrent parachordoma on shoulder.[6] There are reports of a chest wall parachordoma with lymph node metastasis and a forearm parachordoma with lung metastasis in the literature.[1,5,6]

Non-malignant parachordomas can be treated by complete removal of the lesion. Complete resection of the tumour with a clear free surgical margin can be considered as a curative treatment.[1-3,6] In the present case, no relapse was observed within 6 months follow-up after local surgical excision of the parachordoma on the tongue.

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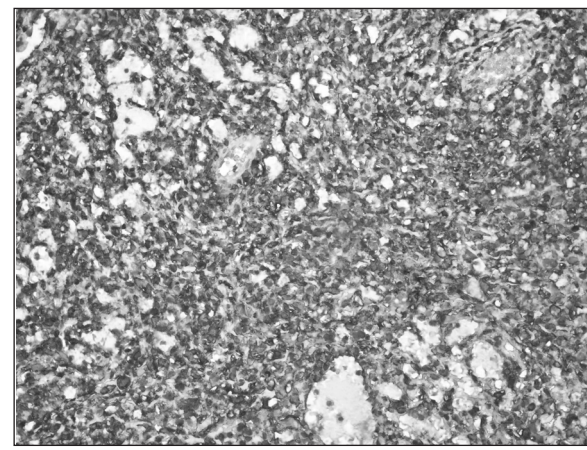

Figure 4. S100 immunohistochemical stain; diffuse and strong positive (HE, x200).

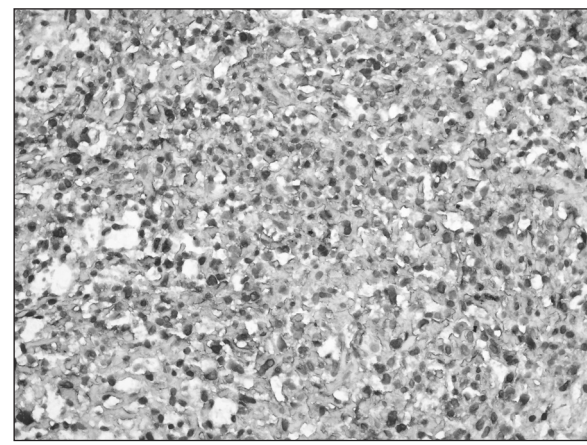

Figure 5. EMA immunohistochemical stain; membranous and moderate patern (HE, x200).

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