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Case Report
Clinical Pathology

An ectomesenchymal chondromyxoid tumour on the lateral border of the tongue

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Abstract. Ectomesenchymal chondromyxoid tumour (ECT) is an extremely rare intraoral mesenchymal tumour. Most of these tumours have been identified on the anterior aspect of the dorsal surface of the tongue. ECT is difficult to diagnose because of its rarity. We report a case of ECT arising on the lateral border of the tongue in a 67-year-old woman. The tumour, measuring 20 × 10 mm in size, was surgically removed. Histopathologically, the tumour was composed of small polygonal cells arranged in sheets, with a myxoid or hyalinized stroma. The tumour boundary was clear; however, the tumour showed a multinodular structure expanding along the tongue surface without obvious capsule. Careful examination revealed the tumour nodule to be spreading in a skip lesion-like fashion away from the main part of the tumour in the striated muscle layer. Although there was no evidence of recurrence at 18 months after the surgery, our observations suggest that surgery for ECT resection with a safety margin is more appropriate than enucleation.

Key words: ectomesenchymal chondromyxoid tumour; lateral border of tongue; benign.

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Ectomesenchymal chondromyxoid tumour (ECT) is an extremely rare intraoral mesenchymal tumour and was first described by Smith et al. in 1995[1]. Fewer than 100 cases of this tumour have been reported in the English language literature[2]. Most of these tumours have been identified on the anterior aspect of the dorsal surface of the tongue[1]. In addition, four cases with lesions involving the floor of the mouth[1], two with lesions involving the hard palate, and one with tonsillar bed involvement have been reported[3]. Recurrence has been reported in three cases, with the time to recurrence varying from 3 months to 5 years[1,4]. However, the clear cause of recurrence remains unknown.

Case report

A 67-year-old Japanese woman, who complained of an asymptomatic mass on the tongue that had been present for 2 years, was referred to Naha City Hospital for diagnostic evaluation. On intraoral examination, a semi-hard elastic mass measuring 20 × 10 mm was observed on the left lateral border of the tongue (Fig. 1A). The overlying mucosal surface appeared smooth. Ultrasonography revealed a smooth hypoechoic mass. No lymph node swelling or other tumourous lesions were identified. Magnetic resonance imaging (MRI) was performed to assess the extent of the tumour and determine possible involvement of the regional lymph nodes. The images showed a well-defined, oval lesion with heterogeneous hypointensity on T1-weighted images (data not shown)

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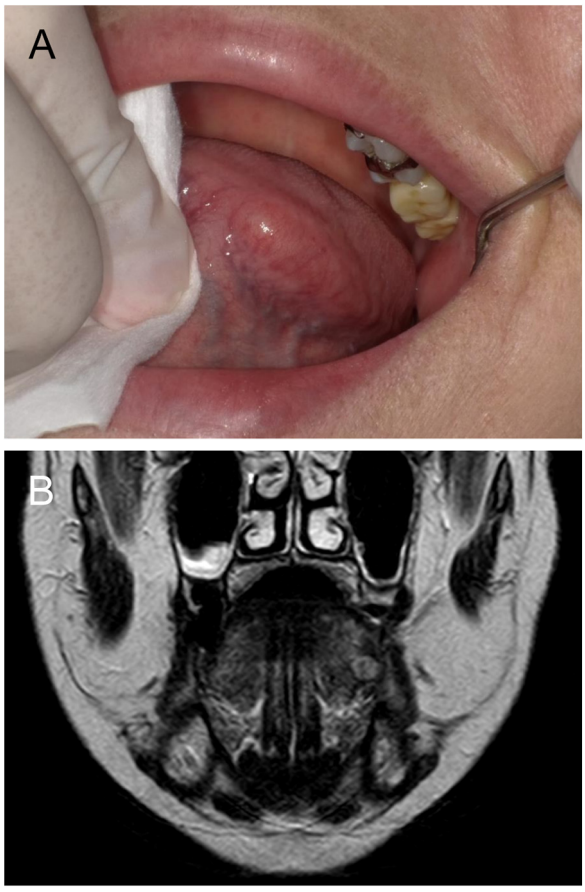

Fig. 1. Clinical and radiological findings of the tumour. (A) Intraoral image of the tumour on the left tongue margin. The submucosal mass measured approximately 20 × 10 mm in size. (B) Coronal T2-weighted magnetic resonance imaging showing a well-defined heterogeneous hyperintensity on the left tongue margin.

and hyperintensity on T2-weighted images (Fig. 1B). On the basis of the clinical features, the lesion was clinically diagnosed as a benign tumour such as schwannoma, and it was excised via an intraoral approach under general anaesthesia. Rapid intraoperative diagnosis was performed and the margin was found to be positive; thus, an additional resection was immediately performed. The margin was negative in the additionally resected specimen.

On the cut surface of the surgical specimen, a solid and lobulated tumour was observed under the mucosa (Fig. 2A). The tumour was whitish, multinodular, and myxoid. Histopathologically, it measured 18 × 8 mm and extended from the submucosal layer to the striated muscle layer (Fig. 2B). Although the tumour boundary was clear, there was no obvious capsule (Fig. 2C). The tumour was composed of small polygonal cells arranged in sheets, with a myxoid stroma. Bundles of striated muscles and peripheral nerve fibres were present within the tumour (Fig. 2C, arrows). In the central area, eosinophilic hyalinized matrices were abundantly deposited among the tumour cells, some of which appeared to be surrounded by chondroid matrices (Fig. 2D). The tumour nodule spread in a skip lesion-like fashion within the muscle layer, and was exposed at the resection margin (Fig. 2B, arrowhead). The tumour cells showed eosinophilic cytoplasm and blunt, rounded nuclei. There were no mitotic figures, nuclear pleomorphism, or vascular invasion.

Immunohistochemistry revealed that the tumour cells were diffusely positive for S-100 protein (Fig. 2E) and weakly positive for glial fibrillary acidic protein (data not shown). There were no positive signals for pan-cytokeratin, epithelial membrane antigen, α-smooth muscle actin, CD34, HMB45, Melan-A, or SOX10 (data not shown). The Ki-67-positive cell index was less than 1%. On the basis of these pathological findings, the tumour was diagnosed as ECT. The patient showed no functional disturbance or evidence of recurrence at 18 months after the surgery.

Discussion

ECT is an extremely rare intraoral mesenchymal tumour, and most of these lesions have been identified on the anterior aspect of the dorsal surface of the tongue[1]. In this report, we described a case of ECT arising on the lateral border of the tongue, which was difficult to diagnose from the clinical and pathological aspects. In addition, rapid intraoperative diagnosis was useful to assess the margin status, because the tumour spread in a skip lesion-like pattern.

Despite showing specific histopathological findings, ECT is difficult to diagnose both clinically and pathologically because it is an extremely rare lesion, with fewer than 100 cases reported in the literature[2]. Clinically, the differential diagnoses include benign mesenchymal tumours such as schwannoma and tumours originating from the minor salivary glands. Pathologically, ECT exhibits a distinct morphology and immunophenotype and appears as a well-demarcated, non-encapsulated, lobular lesion with a myxoid or chondromyxoid background[1]; these features were also observed in the present case.

In this case, differentiation of ECT from schwannoma, glomus tumour, solitary fibrous tumour, and ossifying fibromyxoid tumour was facilitated by immunohistochemistry. Schwannoma, a common mesenchymal tumour originating in peripheral nerve tissue, was ruled out because SOX10 positivity is observed in almost all cases of schwannoma[5]; the tumour cells in the present case were not positive for SOX10. Glomus tumour, an uncommon mesenchymal neoplasm, was ruled out because these tumours are usually negative for S-100[6]. Solitary fibrous tumour is an unusual spindle cell neoplasm that shows CD34 positivity[7]; this was ruled out because of the CD34 negativity in the present case. Ossifying fibromyxoid tumour is a rare, morphologically distinct neoplasm composed of small polygonal cells with bone trabeculae. This tumour also shows immunopositivity for S-100[8]. However, it was ruled out because of the absence of bone formation in our case.

The multinodular growth pattern of ECT may make complete surgical resection difficult. It is considered that the histopathogenesis of ECT begins with the migration of ectomesenchymal cells from the neural crest of the first branchial arch to the tongue[9]. The mechanism by which skip lesion-like multinodular

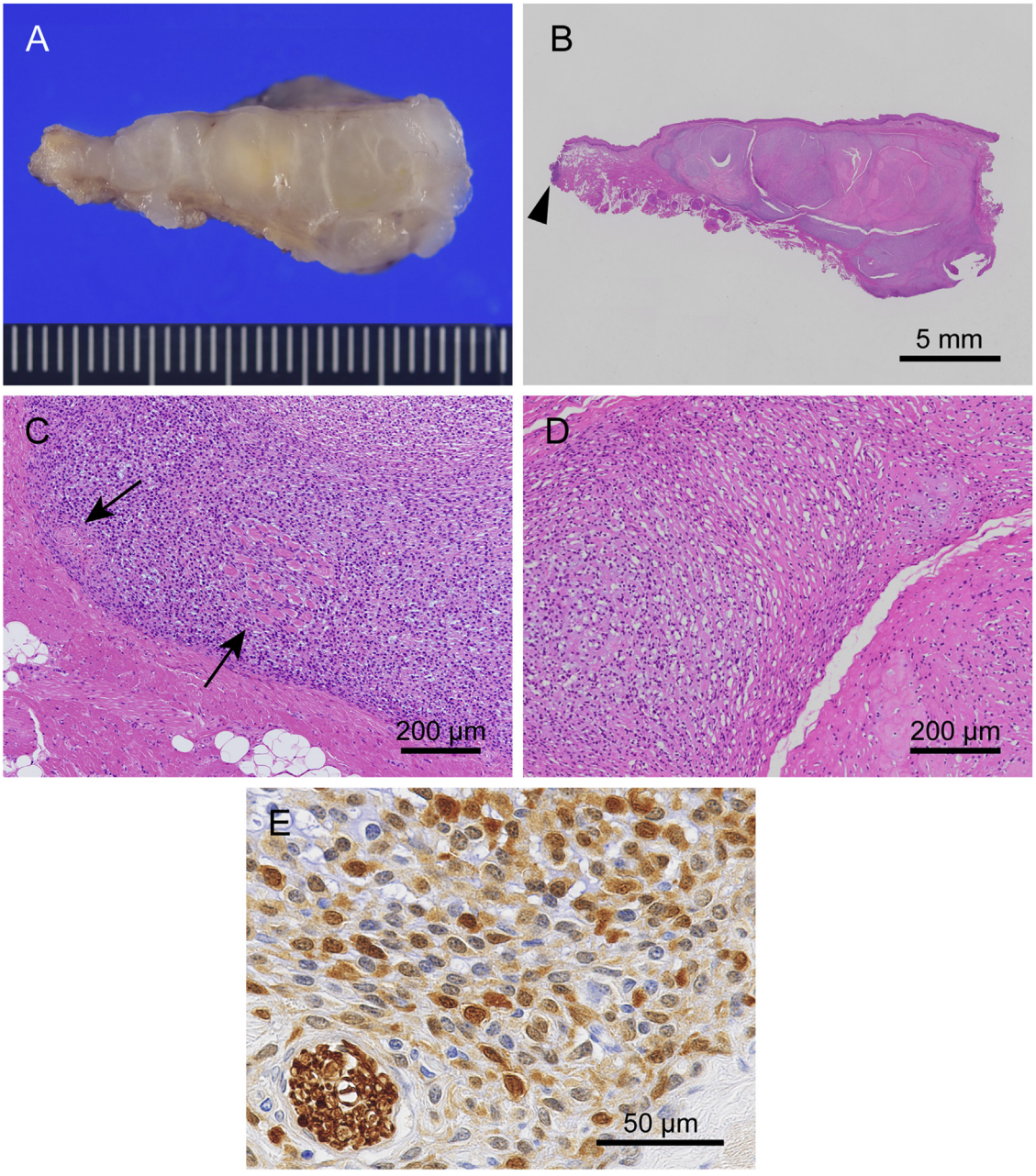

Fig. 2. Gross and histopathological findings of the surgical specimen. (A) Gross findings of the surgical specimen. The cut surface of the surgical specimen. The tumour was whitish, multinodular, and myxoid. (B) Loupe view of the cut surface. The solid and well-demarcated tumour extended from the submucosal layer to the striated muscle layer. A skip lesion-like nodule was observed apart from the main part of the tumour in the muscle layer (arrowhead). Scale bar, 5 mm. (C) There was no obvious capsule. The tumour was composed of small polygonal cells arranged in sheets with a myxoid stroma, and bundles of striated muscle and peripheral nerve fibres were involved within the tumour (arrows). Scale bar, 200 μm. (D) In the central area of the tumour, eosinophilic hyalinized matrices and chondroid matrices were observed among the tumour cells. Scale bar, 200 μm. (E) The tumour cells were diffusely positive for S-100 protein. Scale bar, 50 μm.

masses occur is presumed to be related to the migration of neural crest cells. If such lesions develop and are left behind after surgical removal, recurrence may occur. Recurrence has been reported in three cases[1], although we could not find detailed descriptions of the surgical procedures or margin status. Therefore, the cause of recurrence remains unknown. For instance, Portnof et al. reported recurrence after 5 years in a case of ECT with no capsular structure at the primary excisional biopsy[4]. In contrast, despite the fact that postoperative pathological findings showed a residual tumour, Closmann et al. found no recurrence at 6 months after surgery[10]. However, we believe that long-term follow-up is necessary, because the time to recurrence varies from 3 months to 5 years[1,4].

With regard to ECT removal, surgical resection with a safety margin may be more appropriate than enucleation, and evaluation of the surgical margin by rapid intraoperative diagnosis may be important. For the elucidation of ECT, it is necessary to accumulate knowledge regarding the relationship between pathological findings and recurrence. Moreover, co-operation with specialized institutions

is necessary for rare diseases that are difficult to diagnose.

Ethical approval

XXX.

Patient consent

Written patient consent was obtained.

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Competing interests

We have no competing interests.

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