CASE REPORT

Oral neurovascular hamartoma: an extraordinary verdict in the oral cavity

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SUMMARY

The presence of a neurovascular hamartoma within the oral cavity is truly a rare entity. Scarcely reported in the literature, these hamartomas are smooth, pinkish masses and are painless, and therefore difficult to diagnose. They are benign in nature and apply pressure to their surroundings. The histological diagnosis remains the gold standard as it comprises of neural tissue and vascular components. Treatment is surgical excision with adequate margins. Recurrence is reported in cases of incomplete resection.

BACKGROUND

The term hamartoma is derived from the Greek word 'hamartion' which literally means bodily defect. It was initially used by Albrecht in 1904.¹ It refers to a benign malformation in different areas of the body where growth occurs. The histology consists of an abnormal mixture of cells and tissues localised at the particular site secondary to a developmental error. They do not develop as part of an inflammatory or neoplastic process and have a self limiting proliferation.² On the other hand, they do not have a tendency to regress spontaneously.³

While considering the oral cavity, the hamartoma might result secondary to the proliferation of smooth muscles, skeletal muscles, fat, salivary tissues, vessels and lymphatics.⁴ An oral neurovascular hamartoma is rare because of proliferation of neural tissue solely or in combination with vascular elements.

Described below is an unusual case of an oral hamartoma with neurovascular components.

CASE PRESENTATION

A 14-year-old boy presented to the outpatient department with right cheek swelling for the past 1.5 years. The swelling had been gradually increasing over a period of time and eventually became impossible to ignore. He also reported of decreased mouth opening and a burning sensation within the oral cavity on eating spicy food for 2 years. He did have a significant history of consuming betel nut since the age of 11 years, discontinuing the use once the mouth opening started reducing. His right cheek also had decreased sensation, but there were no symptoms of fever or pain.

The rest of the history was unremarkable.

On examination, the boy appeared to be physically fit with average height and weight. He had a diffuse 2×1.5 cm sized right cheek swelling about two finger breadths below the zygoma. The swelling was soft in consistency, non-tender, non-mobile and non-translucent. The swelling would become

firm on clenching of teeth and mobile on bimanual examination.

Examination of the oral cavity showed significant submucous fibrosis with poor oral hygiene and fullness in the right buccal mucosa. Neck examination was unremarkable as was the systemic examination.

As part of his workup, his complete blood picture was normal. The patient brought with him a CT scan of poor quality and thick cuts. The lesion was not clearly outlined but owing to financial constraint the scan was not repeated (figure 1). No lymphadenopathy was identified on the scan, however.

The family was counselled and an excisional biopsy of the lesion was performed.

INVESTIGATIONS

Histological findings confirmed the diagnosis. The tissue specimen showed haphazard distribution of mature skeletal muscles of varying sizes and nerve fibres closely intertwined with each other on H&E staining (figure 2). The specimen was also evaluated with immunohistochemical stains, which showed positive reactivity with S-100 (figure 3). The lesion was concluded to be neurovascular hamartoma of the oral cavity.

DIFFERENTIAL DIAGNOSIS

The differential diagnosis of the disease included infectious and malignant causes including traumatic hamartoma.

Figure 1 CT image of a lesion in the right masseter muscle.

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Figure 2 High power magnification of the tissue with H&E staining, showing interwoven mature skeletal muscles and nerve fibres.

TREATMENT

Performing a 2 cm incision over the right buccal mucosa, below the Stenson's duct, the dissection was continued using dissection scissors. On reaching the masseter muscle and locating the mass adjacent to the ramus of the mandible, the mass was removed with wide margins (figure 4). The cavity was closed with proper tissue reapproximation using absorbable sutures. Postoperatively, the patient was provided with ice packs to prevent extensive swelling and started on intravenous augmentin 1.2 g/q8h and analgesics for 48 h after which he was switched to oral augmentin in a dose of 625 mg thrice a day and oral analgesics.

He was discharged on the second postoperative day and advised a soft diet for the next 3 days.

OUTCOME AND FOLLOW-UP

On follow-up a week later he was pain-free with the wound healing well.

He was counselled regarding the disease and recommended a six to eight monthly follow-up.

Reviewed in the outpatient department a year later, the boy was symptom and disease free on clinical examination.

Figure 3 Immunohistochemical stain of the same, showing positive staining for S-100.

Figure 4 Excision of the mass using an intraoral incision.

DISCUSSION

Hamartomas arising within the oral cavity are unusual and may show a variety of clinical presentations and histological and growth patterns. The biological behaviour is benign with reasons for development vague. While epithelial and mesenchymal hamartomas involving the oral cavity are rare, one cannot deny that the presence of a neural or a neurovascular component in a hamartoma is even more infrequent.^{4 5} The literature has only reported a handful of these cases.

In 2007, Takeyama *et al*⁵ reported of a hamartoma on the hard palate associated with corpus callosum agenesis, microphthalmia and skin malformation. Prior to that, Goldsmith *et al*⁶ described a case of leiomyomatous hamartoma of the posterior tongue leading to dysphagia in a 16-year-old, whereas Miyamoto *et al*⁷ documented a lingual hamartoma in a child with a cleft palate. Al Habeeb *et al*⁸ reported a solitary but cutaneous neural hamartoma in a middle-aged individual. The lesion had histological characteristics comprising of numerous mature nerve bundles within the papillary dermis. Prior to that, a case of congenital neural hamartoma on the leg was reported by Argenyi *et al*.⁹ Vascular hamartomas are most common of all in the head and neck.¹⁰

As well as being sporadic, hamartomas can present as a part of a syndrome. Epithelial hamartomas can be a part of Cronkhite-Canada syndrome or orofacial digital syndrome,¹¹ whereas neuromuscular and vascular hamartomas may become apparent in Crohn's disease.¹²

Oral neurovascular hamartomas present as smooth surfaced exophytic lesions usually less than 1 cm in diameter. Being pedunculated or wide based, their colour may range from pink to yellow. They are painless lesions leading to general discomfort in the patient.

Histologically, there is no clear boundary between the lesions and the surrounding normal structures as they are non-encapsulated and poorly circumscribed. Composed of hypocellular connective tissue containing aggregates of loose vessels closely packed with small to medium sized nerve bundles, they are not easily distinguishable from the surrounding non-hamartomatous tissues. These nerve bundles are reactive with S-100 protein. This feature can be easily used as a criterion for diagnosis while examining.¹³ This reactivity was also pivotal in our case to reach the final diagnosis. Distinction from quite a few oral benign lesions is still hard. Oral neurovascular hamartomas resemble traumatic hamartomas and can be distinguished

based on the fact that the blood vessels in the former are very closely entwined with the neural component. In traumatic neuroomas the two components are separate.⁴

Once the diagnosis is confirmed the clinical management comprises of surgical excision. Local excision is curative; a management plan similar to most benign oral lesions. A recurrence only follows an incomplete excision and one should not mistake it with malignant transformation.¹⁴ Close clinical follow-up is absolutely essential. The literature does not say much regarding local recurrence of oral neurovascular hamartomas but oral mesenchymal hamartomas arising from the cheek have a reported local recurrence rate of 20%.¹⁵

CONCLUSION

Although oral neurovascular hamartomas are rare entities, they should be considered in the differential diagnosis of oral lesions. It is essential, therefore, to know their clinical presentation and treatment plan.

Learning points

- Oral neurovascular hamartoma is a well-recognised entity.
- Knowledge regarding its diagnosis, histological findings and treatment should be at hand despite it being a rare condition.
- Surgical excision cures the disease and recurrence is only in cases of incomplete resection.

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Competing interests None.

Patient consent Obtained.

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