An oral presentation of dermatofibromasarcoma protuberans with literature review: A case report

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1. Introduction

Dermatofibromasarcoma protuberans (DFSP) is a rare, low-grade malignancy of the dermis. DFSP accounts for <0.1% of all malignancies and 1–2% of all soft tissue sarcomas [1,2]. The overall annual incidence is 4.2 per million [1]. The average age of presentation ranges from 20 to 50 years but has been known to occur in all ages. There is a higher incidence in women and blacks with poorer survival associated with increased age, male gender, black race and location in the limbs, and head and neck region [3]. It typically presents as a slow-growing, painless, skin-colored cutaneous plaque. Following the early slow growth, a period of rapid growth can occur, resulting in classic protuberant nodules [4]. DFSP most often occurs on the trunk and extremities, with only 10–15% occurring in the head and neck region [5]. The common areas affected in this region are the scalp and neck. Characteristic pathologic features include finger-like projections of irregular fibroblasts interwoven in the subcutaneous or muscular tissues, and entrapment of adnexal structures. An immunohistochemical analysis marker specific to DFSP is CD34 [6].

While metastatic rates of DFSP are low, the tumor is locally invasive with a great potential for destruction of underlying tissue [4]. There is often significant subclinical extension of malignant cells leading to a high rate of local recurrence [2]. The current treatment of choice is wide excision with at least 2–3 cm margins or Mohs micrographic surgery. Wide excision requires the removal of normal tissue 2–3 cm from the gross boundaries of the tumor including skin, subcutaneous tissue, and any underlying fascia, muscle or periosteum, to best ensure achievement of adequate tumor extirpation [4,6]. Wide excision may result in sizeable defects. In the head and neck region this can lead to functionally and aesthetically unacceptable outcomes. We present a rare case of DFSP presenting from within the oral cavity and the surgical challenges that arise with tumor excision and subsequent recon-

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structure. In addition, we present a review of reported cases of DFSP presenting within the oral cavity within the PubMed database. The work has been completed in compliance with the SCARE criteria [7].

2. Methods

A literature review was conducted using the US National Library of Medicine “PubMed” database. We sought to identify case reports that included the following terms “buccal,” “oral cavity,” “lips,” and “dermatofibrosarcoma protubersa.” Of the 40 papers identified 35 were excluded because the reports did not identify intraoral presentations of DFSP.

3. Case report

A 50-year-old Caucasian male presented with the chief complaint of a rapidly enlarging nodule in his mouth for over 5 months. He had a remote history of a tumor on the skin of his left cheek that was excised 16 years prior to this presentation. Pathology analysis from the previous surgery was benign. He has a history of chewing tobacco use and no prior medical issues. As a sequela the patient had a mobile scar on the left cheek. In addition, he reported a history of fullness in his left cheek for years that he attributed to this scar. The new intraoral lesion was biopsied at an outside facility and it was confirmed to be DFSP.

Physical examination revealed no facial deformities or palpable lymphadenopathy. Intraorally, a 3 cm irregularly shaped, bulky submucosal mass was present in the left buccal mucosa extending anteriorly to the vestibule of the mouth (Fig. 1A). The lesion was resected with 2 cm margins in all directions, creating a full thickness defect of the left cheek (Fig. 1B). This defect was reconstructed with a folded radial forearm free flap (Fig. 1C). The final pathology report noted positive margins in both peripheral and central deep subcutaneous tissues (Fig. 2A, B).

Subsequent re-excision was performed with 2 cm additional peripheral and 1.5 cm deep margins (Fig. 1D). The resulting defect was reconstructed with a circumoral rotational flap, to close the left oral commissure defect. Buccal advancement flaps were used for closure of the included intraoral defect. The remaining defect was closed using wide undermining of preplatysmal skin and a rotational skin flap from the left preauricular area. One year postoperatively, there was no evidence of recurrent disease (Fig. 1E). Our patient does not report any oral incompetence or speech issues. He is reasonably satisfied with his aesthetic result aside from wanting hair on his reconstructed flap.

4. Results

We reviewed the pertinent literature covering DFSP presenting in the head and neck region. Only five papers were identified dealing with oral presenting DFSP. The average age of patients from our literature review is 56 years-old (44–72 years-old) with 60% occurring in males. All the articles had a similar presentation consisting of a slowly enlarging oral cavity mass. Treatment consisted of conventional excision. Only 2 papers reported formal surgical margins. 3 papers included reconstructive and closure techniques. Interestingly the margins used in these reports were variable ranging from 1.5 to 2 cm but none reported recurrence. It is unknown how long these patients were seen for follow up [Table 1].
5. Discussion

Dermatofibrosarcoma protubersans (DFSP) is a low-grade mesenchymal cell neoplasm with low metastatic potential but a highly locally invasive nature, leading to frequent recurrence after surgical excision [8]. DFSP classically presents in the third to fifth decades of life as a slow-growing cutaneous plaque. It is most often skin-colored but can be violaceous, erythematous or have a blue-brown discoloration. Following the initial indolent phase of slow growth, there can be a phase of rapid growth resulting in the characteristic protuberant nodules [4]. The diagnosis of DFSP is based on histology. DFSP has a characteristic storiform pattern with spindle-shaped tumor cells arranged in a cartwheeling pattern [13]. Histologic analysis will demonstrate mesenchymal tumor cells arranged around a central area of collagen or vascular space [13]. This classic pattern is diagnostically significant as it has been shown to be unique to DFSP. Despite this well described, invariable histologic presentation, the cell of origin is unknown [5,11,13].

The presentation of DFSP in the head and neck region is a relatively uncommon occurrence as it only accounts for 7% of all head and neck sarcomas [1,13]. DFSP of the head and neck most commonly presents as a single nodular area of painless, firm cutaneous swelling and is rarely multinodular [4,13]. In the head and neck the tumor presents in third and fourth decades of life, arising as a firm skin nodule on the scalp or neck over a period of months to years. Of note, as in our case 10%-20% of patients have a history of prior trauma, surgical or burn scars and vaccination at the site of the tumor, although a causal relationship has not been determined [8,9]. The average size of DFSP tumors in the head and neck range from 2 to 5 cm at time of presentation [2,13]. Stojudinovic et al. found the median tumor size to be 2 cm, with the four largest tumors presenting as multinodular plaques.

Classical DFSP is diagnosed when the histological examination showed tumor cells arising in the dermis with the characteristic storiform pattern [13]. DFSP predominantly grows horizontally, typically only involving the dermis and subcutaneous tissue. However, long standing untreated tumors can invade into the deep fascia, muscle and bone. DFSP presenting primarily as an intraoral mass with no involvement of the overlying dermis is extremely rare. Meehan et al. and Nemenjani et al. describe cases of intraoral DFSP. In both cases, the patient presented with a solid, solitary intraoral nodule with no involvement of the dermal tissue of the overlying cheek. In both instances, the histologic analysis demonstrated spindle-shaped tumor cells arising from the submucosa, without any involvement of the adjacent buccal mucosa [7,10]. Similarly, Gonzaga et al. reported a case of buccal mucosa DFSP that presented as a solid, yellow, solitary intraoral mass that was found to be located entirely within the submucosal plane [5]. All
of these cases were classified as intraoral or buccal mucosa DFSP based on the lack of involvement of the overlying dermis and the primary presentation as an intraoral mass [5,7,10]. Although the tumor cells originated in the subcutaneous tissue in this case, it similarly had no involvement of the dermis both grossly and on microscopic analysis and it presented as a solitary intraoral mass, which led us to classify it as an intraoral presenting DFSP.

DFSP presenting as an intraoral mass is extremely rare. This fact in addition to the nonspecific clinical presentation of DFSP frequently leads to inadequate biopsy and delays in diagnosis [5,7,10]. The differential diagnosis of intraoral spindle cell tumors include benign and malignant fibrous histiocytoma, desmoplastic melanoma, neurofibroma, Kaposi sarcoma, fibrosarcoma, and leiomyosarcoma [5,7,10]. DFSP can be differentiated from these tumors based on its characteristic histologic appearance, CD34 positivity which is seen in 85–100% of DFSP cases [2,4,5,8–13].

The surgical management of DFSP has evolved into two routes, conventional excision and Moh’s micrographic surgery. Surgical excision with margins of 2 cm or larger has been shown to be appropriate with a low risk of recurrence [4]. Recently, Moh’s micrographic surgery has been shown to be an effective modality with fewer recurrences rates than conventional excision with smaller margins in cosmetically sensitive areas [9,10]. Imatinib has emerged as an alternative in cases where DFSP cannot be controlled locally [11]. A systematic review found that when used as neoadjuvant therapy, imatinib in conjunction with surgery has potential for tumor removal with negative margins [12].

While DFSP has a propensity for regional invasion, it rarely metastasizes. Both local metastasis to lymph nodes and distant hematogenous metastasis are rare, occurring in less than 5% of DFSP cases [2,5,13]. Factors associated with increased risk of metastatic disease include age greater than 50, increased cellularity, high mitotic index, multiple recurrences, positive microscopic margins, location in the head and neck region and large size 14,111,213. Survival rates are reported over 95% over the course of 5–15 years [9]. The nonspecific appearance, and often asymptomatic presentation often results in initial misdiagnosis and incomplete excision [8]. These tumors can be mistakenly diagnosed as keloids, hypertrophic scars or benign soft tissue tumors such a lipomas [4,13].

6. Conclusion

DFSP is a tumor that rarely can present from within the oral cavity. If a patient presents with a slow growing mass, biopsy and appropriate staining is crucial in determining the correct diagnosis. The literature review is consistent in that excision requires wide margins or Moh’s surgery to reduce recurrence. Furthermore, close follow up is always indicated as there is a high rate of recurrence. Using a multi-disciplinary team approach is essential to diagnose, treat and follow up patients. In the head and neck region, total excision can pose difficult reconstruction needs due to the sensitive and aesthetic nature of facial reconstruction.

Declaration of Competing Interest

There are no conflicts of interest to disclose

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Ethical approval

This study is exempt from ethical approval in our institution

Consent

Written informed consent was obtained from the patient for publication of this case report and accompanying images. A copy of the written consent is available for review by the Editor-in-Chief of this journal on request.

Author contribution

Carlos Martinez: Writing – Original draft preparation, Writing – Reviewing and Editing, Investigation.

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Joram Gilstrap: Writing – Reviewing and Editing, Supervision, Performed Surgery/Intervention.

Harold I. Friedman: Writing – Reviewing and Editing, Supervision.

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Registration of Research Studies

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Guarantor

All mentioned authors were involved in the preparation of this case report and accept full responsibility for the work, had access to the data and controlled the decision to publish.

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