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Desmoid tumor of the tongue

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Abstract

Desmoid tumors are rare neoplasms accounting for 0.03% of all neoplasms and have an estimated incidence of 2 to 4 per million per year. World Health Organization currently refers to all of the deep types of fibromatosis as desmoid-type of fibromatoses. The term "desmoid" refers to the hard, tendon-like appearance of the tumor. The etiological factors suggested include genetic, endocrine and physical factors. About fifty percent of desmoid tumors arise in the abdominal region. The extra-abdominal desmoid tumors present a difficult problem in recognition and management especially because of the striking discrepancy between its deceptively harmless microscopic appearance and its potential to attain a large size, to recur, and to infiltrate neighboring tissues in the manner of a fibrosarcoma. Desmoid tumors are very rare in the oral cavity with less than 5% of cases constituting oral soft tissue fibromatosis. A 22-year old mentally retarded female patient with desmoid tumor occurring in the tongue is presented here.

Key words: *Desmoid tumors, fibromatosis, desmoid-type of fibromatoses.*

Introduction

Desmoid tumors originating from musculoaponeurotic system is referred to as deep type fibromatosis, which are group of non-metastasizing benign fibrous proliferation (1), however, the World Health Organization currently refers to all of the deep types of fibromatosis as desmoid-type of fibromatoses (2).

Desmoid tumors are rare accounting for 0.03% of all neoplasms and have an estimated incidence of 2-4 per million per year (3). Many studies have shown that about 37% to 50% of desmoids arise in the abdominal region (4-7) the most common extra-abdominal sites include the shoulder girdle, chest wall and inguinal regions (8). Desmoid tumors are very rare in the oral cavity. In a series of 24 cases with desmoid tumors reported in the head and neck, only one tumor affected the tongue (9). An extensive search of the English literature revealed that less than 5% of cases constituted oral soft tissue fibromatosis. The radiographic impression of some desmoid tumors often is over-called as suspicious for malignancy. The cytomorphology is non-specific often resulting in descriptive diagnosis (10). A case of desmoid tumor occurring in the tongue is presented.

Case Report

A 22-year old mentally retarded female patient presented to the outpatient clinic with a complaint of swelling on the tongue of 3-4 months duration. The patient's mother first noticed the swelling and recorded its gradual increase in size. The patient herself had no complaints as regards its presence. No other relevant history was noted.

A general survey and examination revealed no relevant finding. On intra-oral examination except for the tongue swelling, no other abnormalities of the hard and soft tissues were noted. Examination of the tongue revealed a single, fairly well-circumscribed spherical swelling on both the dorsal and ventral aspects of the anterior part of the tongue (Fig. 1). The swelling appeared reddish in color. Papillae were present on the tongue dorsum while

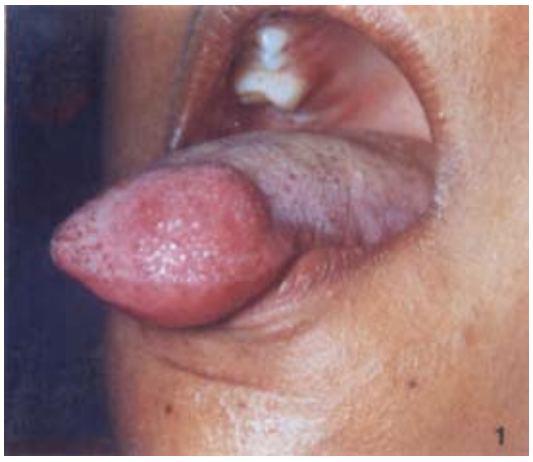

Fig. 1. Clinical photograph showing a spherical swelling in the anterior part of the tongue.

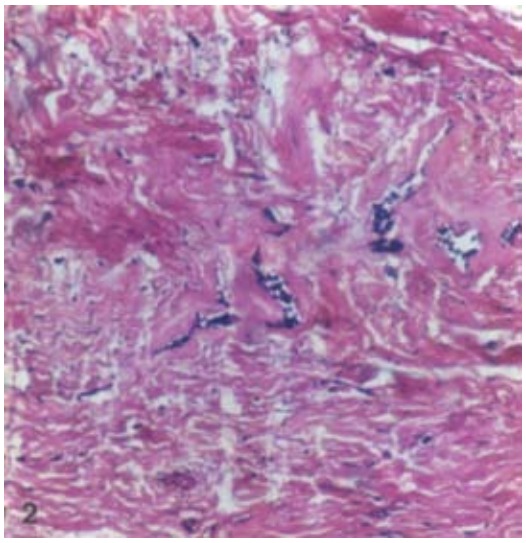

Fig. 2. Photomicrograph showing desmoplasia of fibrous connective tissue (H & E Stain. Original magnification x 10).

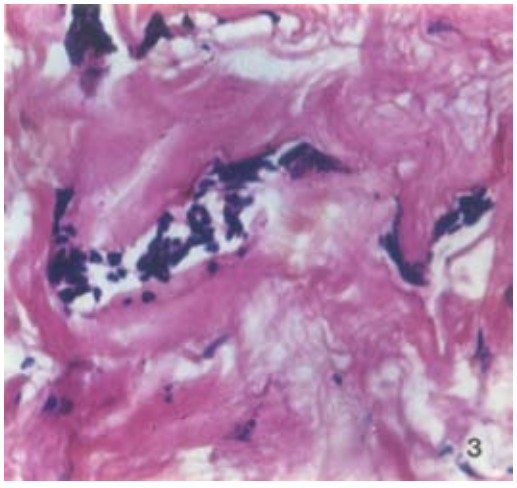

Fig. 3. Photomicrograph showing hyalinization of fibrous tissue around vascular spaces (H & E Stain. Original magnification x 25).

the ventral surface appeared smooth. Palpation revealed a non-tender firm mass approximately 3 x 3 cm in diameter on the dorsal surface and about 3.5 cm x 3.5 cm in diameter on the ventral surface. The consistency was uniform throughout the swelling and was non pulsatile.

An incisional biopsy was done of the tongue swelling. The section consisted of condensed fibrous tissue showing glossy, eosinophilic, hyalinised areas with a perivascular distribution at places. In some areas the condensed fibrous tissue showed moderate cellularity with focal areas of mild inflammatory reaction. Slit-like vascular spaces were seen scattered throughout the connective tissue (Fig. 2 and 3) (Special stains were used to rule out amyloidosis). The histopathological findings confirmed the lesion as desmoid tumor. The treatment employed was complete surgical excision of the lesion. During this period of follow-up (10 years) of the patient, no recurrence was noted.

Discussion

Historically, the term "desmoid" refers to the hard, tendon-like appearance of the tumor. Paget in 1856 recognised a very near affinity between the recurring fibroid tumors of the abdominal wall and similar tumors in an extra-abdominal location. However Nichols in 1923 was the first to recognize and define extra-abdominal desmoid tumors (11).

Masson and Soule (12) believed the term "desmoid tumor" to be acceptable when referring to a specific form of an infiltrative fibrous growth that may appear in various locations, often recurs locally, and does not metastasize, but under certain extenuating circumstances may cause loss of a limb or more rarely death.

The etiological factors suggested include genetic, endocrine and physical factors. Enzinger and Weiss (13) have reviewed cases that have occurred in a surgical scar (cicatricial fibromatosis) and in a previously irradiated area (post-irradiation fibromatosis).

Clinically, it usually presents as a deep-seated, firm, poorly circumscribed mass that grows insidiously over several weeks and causes little or no pain. Tenderness or pain may occur at a later stage of the disease and is usually associated with motion of the involved muscle or muscle group. On reviewing 45 cases affecting the head and neck area (including our case) the mean age was found to be 34.6 years. The lesion shows a female predominance (male:female ratio is 1:2). Table 1 shows the clinical features of 7 patients with desmoid tumors in the oral and peri-oral region (including our case) (9, 14).

A provisional diagnosis of granular cell tumor was made on the basis of the clinical findings. This is the most common lesion on the tongue and the clinical appearance of the lesion was also favorable. Various lesions can be considered in the differential diagnosis. The site is favorable for a lymphangioma and the closely related haemangioma. Although lymphangioma usually presents as papillary le-

Table 1. Clinical characteristics of 7 patients with desmoid tumors of the oral and peri-oral region.

Case no.	Age	Sex	Location	Size	Treatment	Recurrence	Follow up	Ref.
1	50	F	Buccal mucosa	4cm.	Excision with Rχ*	No	10.4 Years	9
2	35	M	Buccal mucosa	3cm.	Excision	Yes in 1st year	9.3 Years	9
3	40	F	Submandibular	2cm.	Excision	No	9.2 Years	9
4	6	F	Submandibular	5cm.	Excision	No	2 Years	9
5	40	F	Retromandibular	5-10cm.	Excision with Rχ**	No	2 years	14
6	36	M	Tongue	1cm.	Excision	No	5.7 Years	9
Present case	22	F	Tongue	3cm.	Excision	No	10 Years	

Rχ*: Radiotherapy for other malignancy (lymphoma) associated with the desmoid tumor
Rχ**: External Beam Radiation for the desmoid tumor

sions and the haemangioma as a superficial, bluish colored pulsatile lesion, deeper lesions of both entities may appear as nodules or masses without any changes in surface texture and color. The mucous retention cyst can occur on the tongue, manifest as a swelling and be deep seated and can thus be included in the differential diagnosis.

The tongue is a common site for the deep granulomatous infections (tuberculosis, leprosy, botryomycosis and histoplasmosis) which can present as nodular lesions. Another granulomatous lesion, sarcoidosis, can present a similar picture.

Amyloidosis and lipoid proteinosis can involve the tongue. However, lipoid proteinosis more commonly produces a diffuse involvement of the oral cavity.

The tongue may be the preferred site for a neurilemoma, neurofibroma, fibroma or myxoma which can present as a single, painless, circumscribed nodule of varying size.

Localization and encystment of parasites, particularly the larval stages of the pork tapeworm, Taenia solium (cysticercosis), and round worm, Trichinella spiralis (trichinosis) have been reported in the tongue musculature. These entities are therefore also considered in the differential diagnosis.

After taking the biopsy and making the histopathological diagnosis, all lesions included in the differential diagnosis were ruled out and the diagnosis of desmoid tumor was confirmed.

The different treatment modalities suggested include:

(i) Prompt radical excision, including excision of a wide margin of uninvolved structures due to the high recurrence rate with local excision (1, 15).

(ii) Postoperative radiotherapy in incompletely excised primary tumors, in recurrent tumors and in tumors in which radical excision is impossible without major loss of function or significant morbidity (16, 17).

(iii) Mega voltage radiation therapy with a median dose of 50 Gy has achieved local control of the lesion (18, 19).

(iv) Zelefsky et al. (20) have shown excellent long-term functional results with Iridium-192 implantations.

(v) The therapeutic usefulness of non-invasive antiestrogen therapy especially tamoxifen (21), progesterone (22) and non-steroidal anti-inflammatory prostaglandin–inhibiting drugs (indomethacin, sulindac) (23), is not fully established, but reports indicate that they may be an effective treatment modality.

Conclusion

A case of desmoid tumor affecting the tongue musculature is presented in this report. The importance of considering this rare entity in the differential diagnosis of tongue swellings is emphasized.

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