Review

Tumors of Fibrous Tissue Origin

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Abstract

Soft tissue tumors are enormously vast and yet relatively undiscovered. Tumors and tumor like lesions of soft tissue continues to be a challenge to both the surgeons and pathologists due to their diversity in biological behavior and histogenesis. Soft tissue tumors are mesenchymal proliferations that occur in the extraskeletal , non-epithelial tissues of the body ,excluding the viscera, coverings of the brain, and lymphoreticular system. Benign tumors are more common than malignant ones in ratio of 100:1. Hematoxylin & Eosin sections act as a useful diagnostic technique in the initial diagnosis of tumors. The goals of the review is to introduce the most common soft tissue entities of fibrous tissue origin and provide with a basic knowledge regarding the diversity of soft tissue tumors.

Key Words: Soft Tissue Tumors, Collagen, Fibroblasts, Histiocytes, Immunohistochemistry.

Introduction

Soft tissue lesions range from nonneoplastic conditions to benign and malignant tumors. Majority of soft tissue tumors are benign, with a very high cure rate after surgical excision. Malignant mesenchymal neoplasms amount to less than 1% of the overall human burden of malignant tumors but they are life threatening and may pose a significant diagnostic and therapeutic challenge since there are more than 50 histological subtypes of Soft tissue tumors, which are often associated with unique clinical, prognostic and therapeutic features. Fibroblastic /myofibroblastic tumors represent a very large subset of mesenchymal tumors.[1] Many lesions in this category contain cells with both fibroblastic and myofibroblastic features. As a dental practitioner, understanding of soft tissue neoplasms has increased significantly and it is important to gain plenty insight regarding common neoplasms of fibrous tissue origin

Common Tumors in Oral Cavity

FIBROMA (Irritational fibroma, traumatic fibroma, Focal fibrous hyperplasia, Fibrous nodule, fibromatosis fibroma)

Most common reactive hyperplasia of fibrous connective tissue in oral cavity occurs in response to local irritation or trauma.[2] Buccal mucosa is the most common location, along the bite line as a consequence of trauma from biting the cheek. Gingival fibromas represent fibrous maturation of a preexisting pyogenic granuloma. Smooth-surfaced elevated pink nodule that is similar in color to the surrounding mucosa. In rare cases, the surface may appear white due to hyperkeratosis or ulcerated due to continued irritation. Fibromas are sessile or pedunculated. Asymptomatic and common in 4th to 6th decades of life. Common in females. Epithelium is stratified often demonstrates atrophy of the rete ridges because of the underlying fibrous mass with collagenized and fibrous connective tissue in bundles of interlacing collagenous fibers interspersed with varying numbers of fibroblasts or fibrocytes. Collagen bundles may be arranged in a radiating, circular, or haphazard fashion and small blood vessels with chronic inflammation mostly of lymphocytes and plasma cells. CD 34, Vimentin can be used as IHC markers.[3] Conservative surgical excision is the treatment of choice with extremely rare recurrence.

Fibromatosis

Broad group of fibrous proliferations that have a biologic behavior and histopathologic pattern that is intermediate between those of benign fibrous lesions and fibro sarcoma. It is a firm painless mass with no gender predilection. Common oral site is submandibular region. If lesion occurs in children, it's termed as juvenile fibromatosis.[2] If lesion occurs within the bone it's termed as desmoplastic fibroma with mild resorption of underlying bone. Histopathologic Features consists of Characteristic proliferation of spindle-shaped cells fibroblasts that are arranged in streaming fascicles and are associated with variable amount of collagen. Lesion is poorly circumscribed and infiltrates the adjacent tissues. Vimentin, Smooth muscle actin, CD34, Desmin as markers.[4] Wide excision is the treatment. 23% recurrence rate has been reported for oral and para oral fibromatosis, but a higher recurrence rate.[2]

Myofibroma

Rare benign spindle cell neoplasm originates from myofibroblasts. It is a solitary exophytic mass with a diameter of 0.3–5.0 cm which occurs at any age. Most of the lesions are asymptomatic and exhibits rapid enlargement. Common oral location is the mandible, followed by the lips, cheek and tongue. Multicentric myofibromatosis primarily affects neonates and infants who may have tumors of the skin, subcutaneous tissue, muscle, bone and viscera. Intrabony tumors create radiolucent defects that usually tend to be poorly defined. Mandibular lesions are well defined unilocular or multilocular. Histopathology of myofibroma consists of interlacing

bundles of spindle cells with tapered ends and blunted nuclei with eosinophilic cytoplasm that resembled smooth muscle. Scattered mitoses are not uncommon. Centrally, the lesion is often more vascular with a hemangiopericytoma-like appearance. Surgical excision is the treatment of choice with a small percentage of recurrence after treatment. Lesions in Viscera or vital organs in infants can act more aggressively and sometimes proves to be fatal.[2] Tumors cells are positive for smooth muscle actin, muscle- specific actin for immunohistochemistry.

Fibrosarcoma

Most common soft tissue sarcoma. Tumor of mesenchymal cell origin that is composed of malignant fibroblasts in a collagenous background. Common in the extremities, only 10% occur in the head and neck region. Primary fibrosarcoma is central, arising within the medullary canal, or peripheral, arising from the periosteum that produces variable amounts of collagen. Secondary fibrosarcoma aggressive tumor with poorer prognosis, arises from a preexisting lesion or after radiotherapy to an area of bone or soft tissue. Tumor associated with genetic mutation from the translocation t (12;15)(p13;q25) giving rise to ETV6-NTRK3 (ETS variant gene 6; neurotrophic tyrosine kinase receptor type 3) gene fusion.[5]

Slow-growing masses that reach considerable size before they produce pain. Commonly in patients of fourth decade of life. Histopath features consists of fascicles of spindle-shaped fibroblasts that classically form a "herringbone" pattern. Fibroblasts are plump with pale eosinophilic cytoplasm and deeply staining spindled nuclei with tapered ends. Cellular and nuclear pleomorphism along with more frequent mitotic activity is noted. Moderate amount of mature collagen may be produced, with areas of hyalinization surgical excision including a wide margin of adjacent normal tissue is the treatment.

Recurrence is noted in about half of cases, and survival rates range from 40% to 70%. IHC Positivity for smooth muscle actin, desmin, S100 protein, and CD34, vimentin. vimentin and negativity for S-100 protein, CD 68, cytokeratin cocktail, HMB-45, CD34, pan actin HHF 35, desmin, smooth muscle actin and epithelial membrane antigen (EMA).[6]

Fibrous Histiocytoma

(Dermatofibroma, sclerosing hemangioma, fibroxanthoma & nodular subepidermal fibrosis)

A benign but diverse group of neoplasms which exhibit both fibroblastic and histiocytic differentiation. Small, firm nodule occur in middle-aged and older adults Characterized by a submucosal, Cellular aggregation of spindle-shaped, fibroblast like cells with relatively pale, oval nuclei. spindled cells may be arranged randomly but usually large areas with tumor cells streaming in interlacing fascicles from a central nidus and intersecting with cells from adjacent aggregates, imparting a storiform or crisscross pattern. lesional stroma is occasionally very densely fibrotic or hyalinized. Scattered rounded foamy histiocytic cells and touton-type multinucleated giant cells are also present. Chronic inflammatory cells, especially lymphocytes, are usually scattered throughout the tumor in small numbers. Few lesions occasionally show areas of calcification or bone metaplasia or present clear granular, balloon, or signet-ring cells.[7] Local surgical excision is the treatment. 5–10% of cases recurring locally. CD34 and Factor xiiia positivity in IHC.[8]

Malignant Fibrous Histiocytoma

(Pleomorphic sarcoma)

Sarcoma with both fibroblastic and histiocytic features. A tumor of older age group. The most common complaint is an expanding mass that may or may not be painful or ulcerated. Histopathalogical features consist of Spindled fibroblast like cells which tend to be arranged in short woven fascicles or bundles. Spindle cells may be long and thin with minimal atypia, but there are usually areas with plump cells containing enlarged, hyperchromatic and irregular nuclei. Rounded, polygonal and irregularly shaped histiocyte like cells may dominate some areas of the lesion. Chronic inflammatory cells often scattered sparsely throughout the tumor including lymphocytes and plasma cells. An aggressive tumor that is usually treated by radical surgical resection. 40% of patients have local recurrences approximately. Vimentin, CD68, CD38, CD34, SMA is positivity for IHC.[7]

Conclusion

Soft-tissue tumors are tumor like lesions which are encountered often in dental practice. These tumors can occur anywhere in the body but few are locally aggressive and tend to recur in head and neck region due to the proximity of vital structures. Hence, it is important to diagnose soft tissue neoplasms by evaluating clinically, histologically or by using IHC markers which help in establishing the cell origin because treatment is challenging in few lesions like fibrosarcoma, malignant fibrous histocytoma due to its aggressive clinical behavior and propensity for local infiltration with a tendency for recurrence. So, it is important to be aware of few soft tissue tumors of fibrous origin with the ultimate goal of improving patient management and outcome with close follow-up.

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