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A Rare Case Of Proliferative Fasciitis Of Head And Neck As A Complication Of Chronic Otitis Media- A Case Report

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ABSTRACT
Proliferative fasciitis is a beningn pseudosarcomatous myoproliferative lesion with ganglion cells in subcutaneous region. The lesion is more common in upper limb, particularly forearm, followed by lower limb. Very rare presentations are seen in head and neck. Though records of other myofibroblastic tumours are seen in literature, extensive proliferative fasciitis of temporal and occipital regions is not reported in literature. We present a case of proliferative fasciitis as a complication of chronic otitis media in a 52 years old male who presented with recurrent ear discharge and mastoid abscess on right side.

Keywords: *Proliferative fasciitis, myofibroblasts, chronic otitis media, temporal bone*

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I. INTRODUCTION

Proliferative fasciitis is an inflammatory myofibroblastic tumour. Chung and Enzinger have proposed that proliferative fasciitis are peculiar subcutaneous and fascial lesion of bizarre histology, which are cutaneous counterpart of proliferative myositis. It manifests as rapidly growing, tender masses that usually arise in extremity, especially forearm and thigh. Majority of head and neck myofibroblastic tumours were seen in oral cavity, zygoma, cheek, orbit, and neck(1). Subclinical trauma could be an etiologic factor. Histopathology and immunohistochemistry aids in diagnosis(2). Despite its rapid growth and bizarre microscopic appearance, the tumour can be adequately treated by local excision.

II. CASE REPORT

A 45 year old male patient presented with a rapidly progressive swelling behind right ear in a span of 2 months duration with hard of hearing and ear discharge for 2 years

On examination, a mass was seen in right external auditory canal arising from middle ear. Tympanic membrane was not visualized. A firm to soft ill defined swelling was seen in temporal and occipital regions. Facial nerve examination was normal. Pure Tone Audiometry revealed 105 db mixed hearing loss in right ear.

Imaging revealed expansile soft tissue density lesion with mixed osteolysis and sclerosis and ground glass appearance noted in right atticoantral cavity, squamous and mastoid portions of temporal bone with erosion of outer and inner tables of squamous temporal bone with mild epidural component, also involving body, clivus, greater and lesser wings of right sphenoid bone and occipital bone. Lesion was crossing and extending into left cranial bones.

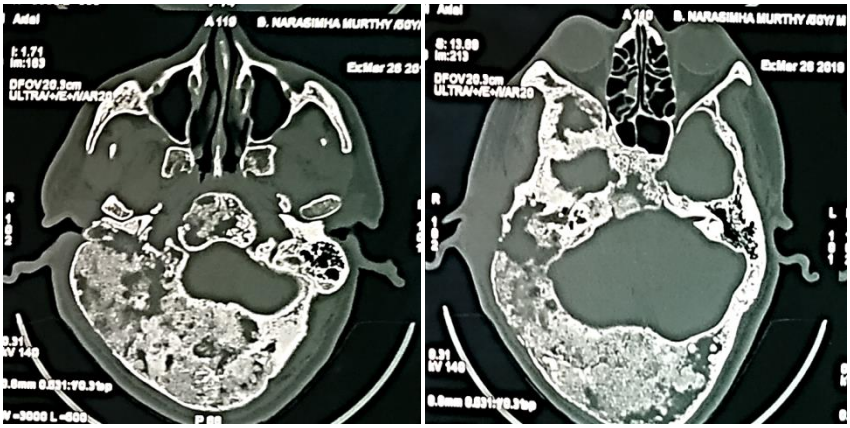

FIG 1. High resolution bone algorithm CT scans. Expansile soft tissue density with mixed osteolysis & sclerosis with ground glass appearance involving right sphenoid, temporal and occipital bone.

Based on clinical and radiological findings, patient was diagnosed as Fibrous dysplasia with chronic otitis media and proceeded for surgery.

The patient subsequently underwent surgery through a postaural incision with extension over occipital region. Modified radical mastoidectomy was done and secondary acquired cholesteatoma in middle ear and mastoid cleared and wide meatoplasty done. Fibrous tissue over root of zygoma, temporal and occipital regions excised and specimen sent for histopathological evaluation.

Histopathology showed dense proliferation of fibrocollaginous stroma with spindle cells and myofibroblasts, arranged in short fascicles. Foci of foreign body giant cell, large ganglion like cells and thick congested blood vessels with perivascular collection of lymphocytes and plasma cells were seen. Lesion was infiltrating skeletal muscle and cartilaginous area. Immunohistochemistry revealed stain positive for SMA specifying proliferative fasciitis. Patient was followed up and no recurrence was found.

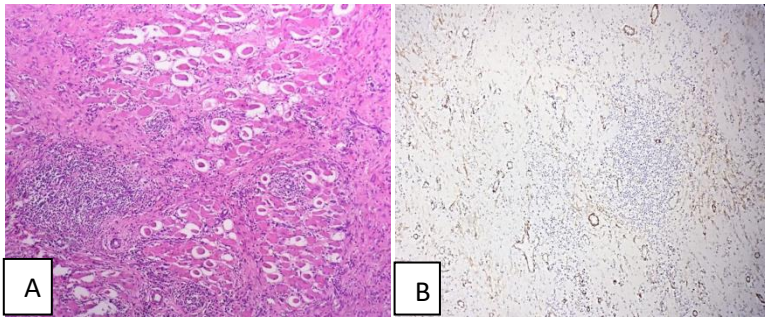

FIG 2

A- Proliferation of fibrocollaginous tissue with spindle cells and myofibroblast. Foci of foreign body giant cells & thick congested blood vessels seen.
B- Immunohistochemistry- SMA POSITIVE

III. DISCUSSION

Proliferative fasciitis is a beningn, non neoplastic well encapsulated lesion, arising from the deep fascia, the dense fibrous connective tissue that interpenetrates and surrounds the muscles, bones, nerves and blood vessels of the body. The 1994 World Health Organization classification of soft tissue tumours propose proper definition for this inflammatory myofibroblastic tumour, which refers to “a tumour composed of differentiated myofibroblastic spindle cells usually accompanied by numerous plasma cells and / or lymphocytes”(3).

Proliferative fasciitis was initially thought to be a variant of nodular fasciitis. It was formally recognized as a distinct entity from nodular fasciitis in 1975. Nodular fasciitis presents as small and well circumscribed lesion while proliferative fasciitis being relatively large and not well circumscribed. So it can be mistaken for a malignant condition considering the rapid growth.They are beningn reparative lesions. The average age of presentation is 54 years and the lesion is uncommon in children. The sex distribution is equal in males and females. Studies show subclinical trauma to be an inciting factor for the lesion.

Proliferative fasciitis develop more frequently in volar aspect of forearm and thigh. In head and neck, inflammatory myofibroblastic tumours often encounter as chronic inflammatory lesion of orbit without invasion

of bony walls(4), and also have been reported in extraorbital locations like maxillary sinus, infratemporal fossa , nasopharynx, pterygopalatine fossa, and skull base(5-8). Mulder et al described three cases of fibroinflammatory pseudotumours of ear, the symptoms being otalgia, otorrhoea, hearing loss and vertigo.

Microscopy reveals poorly circumscribed mass in subcutaneous region which may extend horizontally along fascia. The rare childhood variant is better circumscribed. Histopathology shows fibroblastic/ myofibroblastic spindle cells and ganglion cells which are large cells with rounded nuclei, prominent nucleoli, and abundant amphophilic to basophilic cytoplasm. Mitotic figures are found. The stroma varies from myxoid to collaginous. Immunohistology is positive for SMA and negative for desmin.

CT is insufficient to describe its inherent structure of lytic lesion. MR imaging may show homogenous enhancement with contrast- enhanced T1 weighted images and low signal intensity on T2 weighted images (7). Acute lesions respond to high dose of steroids while chronic lesions, which are more fibrosed don't respond to medical theraphy. Marginal excision is generally curative. Local recurrences are rare, even in lesions which are incompletely excised and they do not metastasize.

IV. CONCLUSION

Proliferative fasciitis is a rare beningn condition found in extremities of the body and rarely presents in head and neck. It can mimic a malignancy given the nature of its rapid growth. Although uncommon diagnosis, otolarngologists should be aware of this beningn condition which is malignant mimicking and tend to radical surgery. Preoperative imaging especially MRI is essential for the diagnosis. Surgical excision is curative and recurrence is uncommon. Proliferative fasciitis as a complication of otitis media is very rare and its presentation in temporal and occipital regions is unusual and has not been reported in literature so far.

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