PACINIAN NEUROFIBROMA OF SCALP (A Case Report)

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Introduction

Although conventional neurofibromas may contain occasional structures that resemble specialized tactile receptors (Wegner-Meissner bodies) or pressure receptors (Pacinian corpuscles), it is unusual to find a neurofibroma in which these elements predominate [1]. Pacinian neurofibroma consists of predominant proliferation of Pacinian corpuscles and we came across such a neurofibroma involving the scalp region.

Case Report

A 20-year-old male sustained cricket ball injury to the left temporal region 2 years ago. The injury was followed by slight swelling over the region. The swelling persisted and was growing slowly. There was no pain or tenderness over the region. There was no history of aural discharge. Examination showed an averagely built and nourished individual. Significant 'café-au-lait' spots were present over the trunk. Local examination of left temporal region showed a diffuse, spongy, nontender, non-pulsatile, and nonreducible swelling over the temporal region extending over the mastoid antrum and adjoining area and pushing the ear lobe forward. No bruit was detected. Neurological examination did not reveal any abnormality. Skull radiography suggested a possibility of dysplasia of medial sphenoid wing on left side. Cranial CT showed a diffuse lesion superficial to left temporalis muscle over the temporal, parietal and occipital regions. The lesion enhanced uniformly with intravenous contrast. Doppler studies of cervical carotids and temporal vessels did not reveal any abnormality.

The patient underwent left temporoparietal exploration and excision of scalp tumour. The lesion was found to be diffuse, soft, relatively avascular and could be separated from underlying muscle. The tumour was removed *in toto* and sent for histopathological examination. The tumour was firm in consistency and measured 12x3x4 cm. The external surface was smooth and grayish brown in colour. No areas of necrosis or haemorrhage could be seen. Microscopic examination (Figs 1 and 2) showed well-defined spherical or oval bodies (Pacinian corpuscles) predominantly, with a background of interlacing

bundles of elongated cells with wavy nuclei (Schwann cells). These interlacing bundles were separated by collagenous stroma. The Pacinian corpuscles comprised of a central zone of polygonal cells flanked by a peripheral zone of concentrically flattened cells. There was no evidence of cellular pleomor-

Fig. 1 : Multiple, spherical to oval Pacinian corpuscles on background of elongated Schwann cells (100x).

Fig. 2 : Pacinian corpuscles at higher magnification showing elongated cells arranged in concentric manner.

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phism, anaplasia or atypical mitotic figures. Based on these histological findings a diagnosis of Pacinian neurofibroma was made.

Discussion

Pacinian neurofibromas were first reported by Prichard and Custer in 1952 followed by Prose et al in 1957 [3]. These tumours are benign tumours and conventionally have been considered to be neurofibromas by Enzinger and Weiss [1]. However, the same authors state that paucity of cases prevents arriving at any meaningful conclusion about its histogenesis. Pure Paciniomas bearing no resemblance to neurofibromas have also been reported in literature [4]. Weiser carried out electron microscopic study of Pacinian neurofibromas and demonstrated that these tumours are characterized by proliferation of the perineural cells and these cells differ from Schwann cells as there is absence of mesoaxons and fragmented basal lamina [5]. Immunohistochemistry studies carried out by Iwanga et al [6] and Blumber et al [7] however, demonstrated a S-100 protein, neuron specific enolase and neurofilament protein favouring an origin from Schwann cells.

Pacinian neurofibromas are usually seen in solitary forms but McCormack et al came across multiple tumours of buttocks [8] and Levi and Curi have also reported multiple neurofibromas of fingers in form of pacinian neurofibromatosis [9]. These benign tumours usually lack cellular pleomorphism and mitotic figures but King and Barr reported a case of pacinian neurofibroma with cellular pleomorphism and mitotic figures and proposed a new term for such forms as 'bizarre cutaneous neurofibromas' [10]. These tumours are usually seen in adolescents and young adults [1] and the commonest site is finger tips [11-13]. Rare sites such as mandible [14], maxilla [15], sacrococcygeal region [4], buttocks [6] and middle ear [16] have also been reported. A Medlar search for

the year 1969 to 1995 was carried out but we did not come across a single report of Pacinian neurofibroma arising in the scalp.

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