Neurofibrosarcoma of Facial Nerve

Neurogenic fibrosarcoma of the facial nerve is a very rare tumour. Till date, only three cases have been reported in the literature (Rosenwaesser (1949), Kettel (1950), Guthman and Simon (1951). The main character of this tumour is gradual facial paralysis. Unless the tumour is large enough, hearing is unaffected.

Stout (1948) wrote, that it is unsafe to diagnose a tumour as malignant neurilemmoma, (neuro-fibrosarcoma, Schwannoma or neurogenic sarcoma) by it's histological pattern only, and preferred reasonable gross evidence of origin from a nerve to substantiate such diagnosis. Russel and Rubin-Stein concluded that when a well defined malignant spindle cell tumour arises in the course of a nerve at a site favoured by a neurofibroma there is reasonable argument for accepting it as a malignant tumour of fibrocytes. Strict dependence on the nature and degree of involvement of nerve is indeed necessary, since extraneural soft tissue sarcoma may completely surround a nerve and may infiltrate the epineurium and on rare occasions may actually invade the nerve longitudinally for some distance, simulating a nerve tumour.

Geschickter (1935) held that neurofibrosarcoma represents a malignant change in a solitary benign tumour. But Stout (1948) and Vieta (1951) did not agree with the above conclusion. D'Agostine et al (1963) also denied any relationship between benign encapsulated neurofibroma and neurofibrosarcoma. None of the 24 cases studied histologically by them revealed any trace of cellular pattern reminiscent to benign neurofibroma.

Karagh et al (1960) described 4 cases of neurofibrosarcoma out of 143 benign neurofibromas of head and neck during the period between 1910 to 1957 and none of their cases had involvement of facial nerve either benign or malignant. We are presenting a case of neurofibrosarcoma of intra-temporal part of facial nerve.
Case Report

S.S. 28 years Hindu Male was admitted in E.N.T. wards on 15.5.68 with complaints of gradual facial weakness over left side for the last 2 years. He had otorrhoea and deafness since 9 months. Pain accompanied otorrhoea and was increasing. He developed a firm swelling behind and below the pinna for 6 months prior to consultation. There was no vertigo.

Examination revealed pus and granulation in left external auditory meatus. His posterior meatus wall was bulging throughout its length. His deafness was of mixed type. He had complete infra-nuclear paralysis of facial nerve. Swelling behind the pinna was diffuse measuring 2x2 cms. (Fig. 1) Skin over the swelling was slightly adherent to deeper structure particularly in its lower portion. Biopsy from granulations from E. A. M. was inconclusive. X-ray showed marked bony erosion (Fig. 2) in the region of mastoid cells.

Mastoid was opened through postaural route. Cortex was intact but tip was eroded by firm pinkish growth which was extending below the tip into the soft tissues of the neck. Posterior meatal wall was also found to be eroded and growth was pushing the skin of external canal anteriorly. Cortex was opened and was found full of pinkish mass which was continuous with the growth below. Bony landmark of the middle ear could not be identified. Growth was removed as far as possible. Dura mater in the posterior fossa was seen to be infiltrated with growth. The cavity was closed and dressed. The tumour biopsy material was preserved in 10% formalin-saline and processed as usual. The microsections revealed tumour tissue composed of interwoven fascicles of spindle cells interspersed with wavy collagen fibers. The spindle cells exhibited marked pleomorphic and hyperchromatic nuclei. Their nuclei revealed abnormal mitosis and assumed giant size in some of the cells (Fig. 3). The histological diagnosis was neurogenic fibrosarcoma. Patient refused Radiotherapy and left hospital. He died on 16th Dec. 1968, the cause of death was not known.