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PLEXIFORM NEUROFIBROMA OF FOREARM: REPORT OF AN UNUSUAL CASE.

Neurology

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ABSTRACT

Plexiform neurofibromas represent an uncommon variant of neurofibromatosis type 1 (NF-1) in which neurofibromas arise from multiple nerves as bulging and deforming masses involving also connective tissue and skin folds. The treatment is usually surgical if it is interfering with daily work. A case of solitary swelling over right forearm on anterio-lateral aspect extending to wrist in skin & subcutaneous plane clinically diagnosed as lipoma turned out to be plexiform neurofibroma. Plexiform neurifibromas are type of Neurofibromatosis Type1. Here we present a similar case of forearm Neurofibroma associated with restricted daily activity which needed surgical intervention.

KEYWORDS

Plexiform, Neurofibroma; Neurofibromatosis Type 1 (NF-1).

Introduction -

Plexiform neurofibromas are debilitating complication of neurofibromatosis type 1 (NF1) [1].It is most prevalent dominantly inherited genetic diseases of the nervous system. Common sites being craniofacial involving trigeminal nerve other sites are glutial region & thighs. They can be substantially morbid, cause disfigurement, functional impairment. May even be life threatening, since piexiform neurofibromas are likely to transform in to malignant peripheral nerve sheath tumour (MPNST).This particular complication is difficult to manage since no effective therapies for malignant soft tissue sarcomas at present & delay in diagnosis as small portion of large pre-existing tumor shows malignant change. The mainstay of treatment of plexiform neurofibromas and of MPNST is wide local surgical excision. Neurofibromas are complex tumors composed of axonal processes, Schwann cells, fibroblasts, perineural cells, and mast cells. Histology characterized by a proliferation of Schwann cells in the nerve sheath across the length of nerve, involving multiple fascicles and also can involve multiple nerves. These convoluted masses described as “bag of worms” appearance [1,2]. Plexiform neurofibromas are locally invasive, non-metastating and categorized by their location. Head, neck and face tumors are most common, followed by lesions of spine, extremities, mediastinum and abdomen. Neurofibromas occasionally cause pain or loss of function due to nerve compression. Surgical management of plexiform neurofibroma is challenging due to infiltrating nature of these tumors, operative morbidity and tendency of recurrence [3,4]. We report a case of right fore arm plexiform neurofibroma in 32 years male with specific emphasis on its clinical, MRI and histopathological features.

Case History

32 years old male presented to the department of General surgery with chief complaint of swelling on the right forearm on lateral aspect since last 14 years. It started as a small swelling which gradually increased to present size now extending to right wrist. No history of regression in size or rapid growth. No history of similar lesion in family. On general examination, it revealed presence of brownish white café au-lait spots over trunk measuring sub-centimetric to 1.5 cms in diameter. No other swelling all over the body. On local examination, a diffuse swelling present over lateral aspect of right forearm extending to wrist measuring 15 cms x 6 cms x 4 cms in size with indistinct margins in a subcutaneous plane. It was mobile but was adherent to skin at few places. It was soft in consistency. Based on history and clinical findings a provisional diagnosis of neurofibromatosis type 1 was made. MRI scan was done which revealed a large lobulated mass in the right forearm extending to right wrist but restricted to subcutaneous plane, facial planes free of tumor invasion. (Figures 1 and 2). Patient was then operated for wide local excision of tumor with primary closure of defect with Romovac drain in subcutaneous plane [5]. Post operative course was uneventful and the drain was removed on 7th Post operative day [POD] and sutures removed on 10th POD. The histopathological examination of excised tumor revealed section composed of bundles of nerve fibres arranged in concentric manner with Schwann cells and fibroblasts with wide separation of small nerve fascicles suggestive of plexiform neurofibroma (Figures 3).

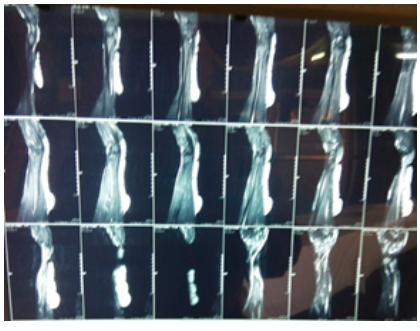

Figure-1 MRI Showing the Neurofibroma

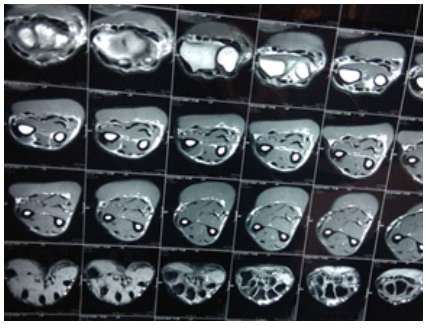

Figure-2 Cut Sections of MRI showing a better understanding of the tissue planes.

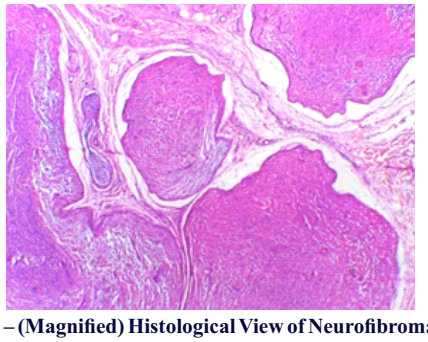

Figure 3 – (Magnified) Histological View of Neurofibroma

Discussion-

Neurofibromatosis is the common neurocutaneus syndrome [6]. Has two genetic variants type1 and type2. NF-1 occurs in 1 in 4000 births. The gene is located on chromosome 17 which codes for the tumor suppressor protein neurofibromin. Incidence of PNs is around 25-30% of NF-1 patients. These tumors arise in various regions including trunk, limbs, head and neck. They can cause dysfunction like cosmetic abnormalities, pain and functional deficits. Two types of neurofibromas have been recognized 1) Diffuse type and 2) Nodular type [7]. They can arise anywhere along the nerve and have poorly defined margins. They frequently involve cranial nerves and upper cervical nerves. The cranial nerves commonly involve in PNs are trigeminal, glossopharyngeal and vagus nerves. Neurofibromas are composed of Schwann cells, fibroblasts, mast cells and vascular components. They consist of poorly organized mixture of nerve fibrils with interlacing of nerve tissue. Though surgery is the main stay of treatment for solitary neurofibromas, in case of plexiform neurofibromas this is not a cure, because of the invasive nature and location of the tumor, which prevents complete resection. Moreover surgery is not undertaken unless the tumor becomes symptomatic or extensive disfigurement occurs. Plexiform neurofibromas can transform to malignant nerve sheath tumor [8] hence surgical excision is advised.

Conclusion-

Plexiform neurofibroma causes pain, disfigurement and may turn rarely to malignancy this makes difficult to predict the prognosis of the disease. Its progressive nature proceeds towards the worst condition of the disease requiring long term follow up of patients.

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