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RECURRENT PLEXIFORM FACIAL NEUROFIBROMA WITH IMPACTED MANDIBULAR MOLAR (UNUSUAL CASE REPORT)

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ABSTRACT

Plexiformm neurofibromatosis is a very rare variety of neurofibromatosis type 1. It is a benign tumour originating from visceral peripheral nerves, type 1 with trigeminal nerve. There are three types of neurofibromas Localized, Diffused, Plexiform. Plexiform neurofibromas usually grow upto 5 cms but can grow continuously larger & become giant neurofibroma. It is highly infiltrative & devastating tumor with a very high recurrence rate. Here we are reporting a case of facial plexiform giant neurofibroma with impacted mandibular last molar who had undergone surgery & disease reoccurred within 6 years.

KEYWORDS: Recurrence, Giant Neurofibroma, Impacted Molar

INTRODUCTION

Neurofibromatosis is an autosomal dominant disorder which affacts skin, soft tissue nerves & bones, it is neurocutaneous affecting almost every organ of body & is of two types, type 1 & type 11.

Neurofibromatosis type 1 is an autosomal dominenet genetic syndrome caused by mutation in genes coding for neurofibromin [bhati]. There are three types of neurofibromas Localized, Diffused, Plexiform. Plexiform neurofibromas usually grow upto 5 cms but can grow continuously larger & become giant neurofibroma [1]. Giant neurofibroma is poorly defiened term & is used to describe a neurofibroma which grows to a significant size. Plexiform neurofibromas are benign tumours that originate from nerve sheath cells or subcutaneous peripheral nerves & can involve multiple fascicles. Plexiform neurofibromatosis is a relatively common but potentially devastating manifestation of neurofibromatosis type 1 (NF1). It produces very hideous deformity if the face is involved & occur in as much as 30% of patients with neurofibromatosis type 1[2]. Massive plexiform neurofibroma result in functional disability & disfigurement & occur in as much as 30% of cases of cases with type 1 neurofibromatosis. PNF present at birth and often progress during early childhood at growth rate & pattern which vary very significantly & unpredictably.

The condition can cause disfigurement by pulling down of important structures. Because of the large size of the tumors and involvement of multiple fascicles of nerves, the risk of neurological and functional destruction upon tumour resection is very high. Surgical management remains the mainstay of therapy, but in head & neck region it is limited by infiltrating nature this tomour inhert operative morbidity & high rate of growth tumor.[3] Such neurofibromas infiltrate multiple tissue planes and are thus much more difficult or impossible to resect. Most cases require repeat surgery as complete excision is generally not possible due to the infiltrating nature of these tumors[4]. Surgical intervention is thus

commonly postponed to as long as possible. PNFS have a potential for transformation into highly malignant peripheral nerve sheath tumours which occur in approximately 5% of patients [5].

MATERIALS & METHODS

A 45 yrs pld male reported to the department of oral & maxillofacial surgery indira Gandhi govt Dental college Jammu with history of tooth ach. Patient had facial disfigurement with skin folds hanging from face on left side. He had overhanging folds of loose skin from face. Patient gave past history of a small similar swelling in past which was operated 6 yrs back (Figure 1). As per history swelling recurred & increased progressively to large giant size. On Physical examination face was disfigured due to overhanging folds of skin in orbital & zygomatic region. Huge soft tissue swelling involving lft side mandible, nasolabial fold. LT, angle of mouth & LT eye drooping down, LT eye appeared smaller RT eye but Vision was not affected. There was deviation of mouth to RT side completely (Figure 2, 3, 4 frontal & lateral profile) Intraorally patient had soft tissue folds on left mandibular posterior region, with impacted mandibular last molar. These mucosal folds were soft nontender (Figure 5). OPG revealed radiolucency over LT side mandible w, on. On left side mandibular last molar was impacted & bone destruction on body, angle region extending to ramus of mandible (Figure 6) patient was not interested for treatment of fascial disfigurement & was upset due to recurrence of lesion at much larger size than before. AS per patient he was living happily without any pychologic embarrassment & had no functional or neurologic problems even before surgery He had got his surgery done for cosmetic or esthetic reasons only but unfortunately lesion recurred to much bigger size than before RT side patient had grossly decade mandibular first molar with pain, for which patient had come to hospital. Extraction of first molar was done under LA.

DISCUSSIONS

Plexiform neurofibroma is a rare type of generalized neurofibromatosis, which occur due to overgrowth of neural tissue in the subcutaneous fat or deeper in the body. These are considerd as hamartomas rather than typical tumour [67]. PNF are benign peripheral nerve sheath tumours, often involving trigeminal or upper cervical nerves[8]. They are diffused & elongated fibromas usually seen in only 5-10 % of patients with NF1[9]. They originate from nerve sheath cells & can involve multiple fascicles PNF is a rare type of generalized neurofibromatosis which occurs due to overgrowth of neural tissue in subcutaneous tissue. The diffuse & soft nature of PNF is often compared with “a bag of worms” [7]. The condition is autosomally dominant, with variable penetration & presents as multiple nodules of varios sizes which are firm non tender, often associated with café au lait spots & spindle deformities [10]. Café ul lait spots are pigmented macules that may vary from light brown to dark color with irregular borders, can appear anywhere on face but are less common on face[4]in the present case there were no spots.

PNF can occur anywhere along a nerve & may appear on the face [7] [11] orbit, globe [12] or spinal cord & frequently involve fifth ninth & tenth cranial nerves[13]The condition can be quite disfiguring as in our case. Symtoms can be ranging from discomfort to extreme pain, neurologic deficits & psycologic disturbences because of abnormal anatomy [7] where as in our case there was no pain, irritation, discomfort neurologic deficit etc. The tumour can cause intracranial pressure, seizure, or cranial nerve abnormalities De Bella K et a [14] & national institute of health concensus [15] reported that many patient with NF1 (25-40%) often have below average intelligence & 5-10 % have mental retardation where as the present patient had normal I, q or intelligence & was mentally fit. Although skin is most predominantly involved stomach kidneys, urinary bladder, heart, larynx are also affected. In head & neck region scalp, cheek neck oral cavity [4].

Bone involvement may be due to both external resoption & internal osteolytic defects. External resoption may be due to pressure applied on the bone by the neurofibroma [16]. As is in our case, radiographically bone resoption is seen in body & ramus region. Skeletal involvement is seen in 40% of the cases scoliosis being most common feature [17] TMJ may be involved, oral cavity is involved in 66-72% of the cases, there is lengthening of fungiform papilla in 50% of the cases with PNF. In our patient tongue was normal, In 25%0f PNF cases oral neurofibromas are seen, hard or soft tissue of oral cavity can get affected.

Most common lesions are seen on tongue [4, 5, 9, 12, 13 from 13yrs old] in our case gingival is involved, which is very uncommon location. Shapiro et al reported that gum involvement in PNF1 is 5 %[18]. Localized oral neurofibroma usually appear as asymptomatic nodules covered by normal mucosa [4,7,9,12] as in our case. However when adjacent cranial nerves, they can affect the motor functions of facial, hypoglossal nerve, or sensitivity of trigeminal nerve.[4,7,9] Gingival neurofibromas can cause malposition of teeth or impaction[4, 12] like the present case & also may show facial disfigurement due to hyperplasia of mandible or maxilla, malar bone & TMJ. Facial PNF may also cause facial assymmtry as in our case.

Radiologic findings include widening of mandibular cannal, mandibular foramen & mental foramen [12]. Neurofibromas are rarely intraosseous & show unilocular well defined radiolucencies. [9 10]. In our case there was nodular intraoral nodules, of nomal mucosa color, tongue was not involved, bone destruction on body, angle & ramus of mandible with impacted last left mandibular molar, which is not reported till now. Neurofibromas usually grow slowly without pain & can be symptomatic at birth or through time. Most cases require repeat surgery as complete excision generally is not possible due to the infiltrative natures of these tumors [3]. Such neurofibromas infiltrate multiple tissue planes & are thus much more difficult & impossible to resect[4]. Partial or total resection of these lesions can be treatment of choice to solve aesthetic or functional problems. Total resection with 1 cm nomal margin is treatment of choice for accessible & small tumours [8]. But it is advisable to wait until growth is complete to prevent recurrence [7, 9, 12, 13]

Clinical management for the PNF requires a multidisciplinary approach. However current treatment options for PNF are limited to surgical intervention. Resection is performed when tumour is severly disfiguring or severly compromises functionality.[19]. Complete resection is often difficult because of extensive & infiltrative nature of the lesion[20]. Needle et al [3] analyzed the largest series of surgically managed PNFS & demonstrated that 54% recurred within a 10 years period, with the greatest risk of recurrence found in lesion involving the head & neck region. Resection & debulking of invasive PNF is however associated with high rate of recurrence.

In one pediatric series, resection developed recurrence in 20% & incomplete had a recurrence in upto 45%of cases [3]. One of the limiting factor is vascularity of these lesions & abnormal tendency to bleed. These tumours bleed profusely during surgery because of friable nature of neo vessels. Surgical management remains the mainstay of treatment for these tumours but functional disturbances are almost inevitable while resecting tumours involving the head & neck region[21] No chemotherapeutic agent has been yet identified that reduces the size of these tumours [22]. In our case recurrence of tumour took place after 6 yrs of initial resection, tumour grew aggressively, with more functional & cosmetic deformity. In our case recurrence took place after 6 years of resection which caused much Facial disfifutrment than before.

CONCLUSIONS

Neurofibromas infiltratrate multiple tissue planes & Due to the infiltrative nature of these tumours especially in

head & neck region are much more difficult or impossible to resect. Extensive surgical procedures have to be weighed against functional disturbences that are almost inevitable while resecting invasive neurofibromas in head & neck. Surgery should only be undertaken after explaining the patients functional disability after extensive resection of facial tissues, postoperatively & high rate of recurrence. Due consideration should be given to the possible psycologic problems & ragular counseling sessions should be given to the patients.

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APPENDICES

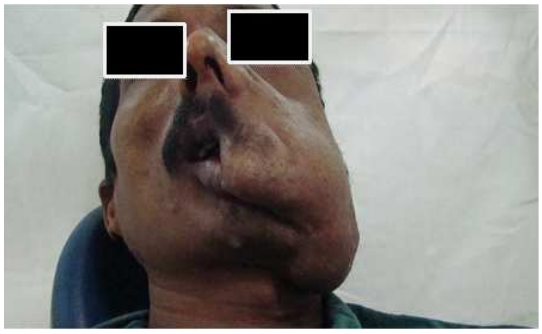

Figure 1: Scar Mark of Previous Surgery

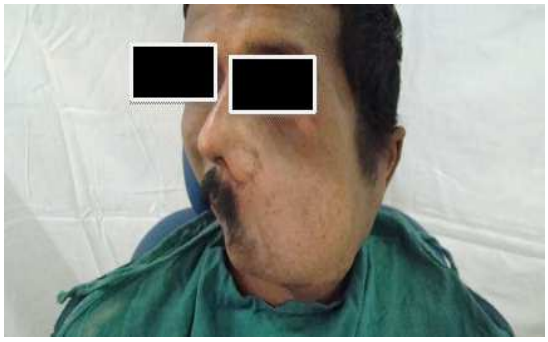

Figure 2: Facial Plexiform Neurofibroma (Lateral Profile)

Figure 3: Front Profile

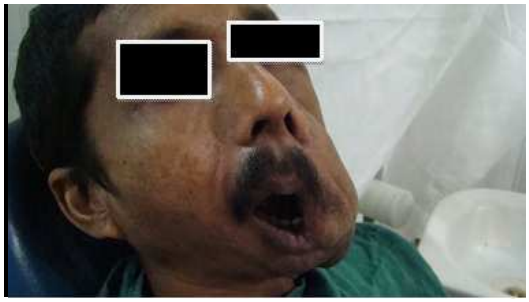

Figure 4: Deviation of Facial Structures to RT Side

Figure 5: Intra Oral Mucosal Nodules

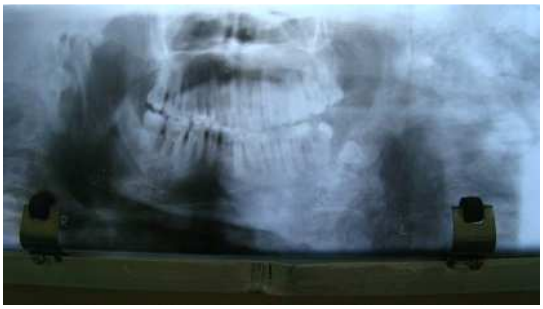

Figure 6: Bone Destruction LT Ramus Angle & Body

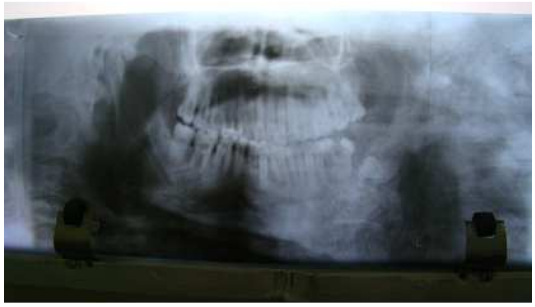

Figure 7: Impacted Mandibular Third Molar & Missing 2nd Molar