CASE REPORT

Plexiform Neurofibroma of Face: A Challenge for Plastic Surgeons

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ABSTRACT

Plexiform neurofibroma is a benign tumour of peripheral nerves and connective tissue. They develop most often in patients with neurofibromatosis type 1 (NF1) and often grow continuously. Removal of plexiform neurofibromas is usually unsatisfactory because the network-like growth of these tumours often involves multiple nerve fascicles and other adjacent tissues. We present an interesting case of Plexiform Neurofibroma involving right half of the face. Surgical excision and debulking of the tissues were performed alongwith the correction of orbital skeleton and facial contours.

INTRODUCTION

Plexiform neurofibromas (PNF) are benign tumours originating from subcutaneous or visceral peripheral nerves and involves multiple fascicles. These tumours are either present at birth or develop within the first years of life. PNF occur almost exclusively in patients with neurofibromatosis type 1 (NF1), an autosomal dominant disorder caused by the genetic alteration of the tumour suppressor gene, NF1.[1] These lesions usually progress to adulthood, affecting skin, soft tissue, nerves and bone to varying degrees.[2] PNF can arise in various parts of body and may lead to severe clinical sequelae.[2] The course of the disease is unpredictable but the patients commonly seek medical care after a disfiguring mass and various facial soft tissue deformities appearing with or without dysplastic bony defects of the facial skeleton.[3] When extensive invasion of the facial soft tissue occurs, it is nearly impossible to radically excise all the involved facial soft tissues including the facial nerve.[3] Tumour rests may re-grow after the operation. The growth rates and patterns vary largely and are not predictable. Magnetic resonance imaging (MRI) enables distinction among 3 growth patterns in PNF: superficial, displacing and invasive.[4] For superficial PNF, total or sub-total resection may be possible. But invasive PNF infiltrates multiple tissue planes and cannot be completely resected without functional disturbances.[5] Plastic surgeons can be faced with facial deformity caused by deep or wide extensive involvement of plexiform neurofibroma on the craniofacial structures resulting in a debilitated social life for the patient.[6]

A rare case of plexiform neurofibroma with extensive facial involvement is presented.

CASE REPORT

A 26 years old woman visited the Plastic Surgery Clinic because of a disfiguring neurofibroma involving right half of the face, thereby affecting

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cheek, orbit, nose and lips (Fig. 1). Clinical examination of facial nerve paralysis was not possible because of the nature and extent of the involvement. Her baseline investigations were normal. X-rays of the facial skeleton revealed involvement of the underlying bony skeleton, i.e., outer orbital wall. A staged correction was planned to reconstruct the facial deformity. First of all, a diluted solution of adrenaline was injected carefully in the whole tissue. Excision of the mass was performed as far as possible. Complete excision was not practicable due to the wide spread and involvement of the underlying facial muscles. Enough tissue was left with the skin to avoid any necrosis. Faciocutaneous flaps were trimmed at the end of the procedure. Mass was also resected from the right nostril. Facial nerve was not found during the operation. Suspension of the angle of the mouth was performed. Resection of the lid and periorbital tissue was also performed. Debulking of the lid was combined with lateral canthopexy. Patient received blood transfusions during and after the operation. The drains were placed which were removed on 5-7 postoperative days. Compression dressing was applied. Postoperative course was satisfactory (Fig.2). There was no skin necrosis. The stitches were removed on the10th postoperative day. The patient was discharged after two weeks. Regular follow-up was done. The second stage was performed after 6 months in which the bony skeleton of the orbital wall was debulked and reconstructed. Healing was satisfactory with good postoperative result (Fig.3). Further suspension of the angle of the mouth with palmaris tendon graft was planned for the next stage of reconstruction.

DISCUSSION

Plexiform Neurofibroma is a major facial hamartoma and one of the most devastating, destructive, and debilitating disease involving the skin, muscle, mucosa and skeletal system.[3] Although benign in histological appearance, these lesions can be clinically malignant in their deforming and inexcisable growth. Facial drooping and other deformities frequently recur despite partial excision, leading to unsatisfactory function and poor aesthetic appearance. Total removal of facial neurofibroma and perfect correction of the facial soft tissue by conventional means or a single operation is impossible because of the invasion of superficial and deep facial structures, destruction of normal histological layers, and regrowth. Resection is further complicated by the absence of any encapsulating structure and, thus, any intermingling with normal facial layers from the skin, superficial fascia, muscles, nerves to, possibly, the cranial cavity.[1,7] Various factors such as patient's age, extent and location of the tumour have been discussed to correlate with surgical outcomes and post-operative tumour regrowth. Similarly loss of elasticity or integrity of the remaining soft tissue, regrowth or recurrence of a progressive mass, loss of skeletal and soft tissue support, and soft tissue detachment from skeletal structure by tumour infiltration, contribute to worsen the facial deformities.[7]

When viewed histologically, diffuse proliferation of compactly arranged spindle-shaped fibroblasts might infiltrate the surrounding soft tissues including the dermis, subcutaneous tissue, and blood vessels. Preexisting collagen bundles in the dermis and subcutaneous tissue forming a loose fibrous tissue separate the fibrous myxomatous dermis and skin. These changes cause soft tissue detachment from skeletal support and are aggravated by facial gravitational forces.[7,8]

Various techniques have been described including the netting operation in which Teflon mesh was used to suspend the inelastic drooping facial soft tissue [5,8,9] Similarly suspension of the tissue to the underlying bone with Mitek anchors may give more prolonged retention.[2] Plexiform neurofibroma is a

4. Mautner VF, Hartmann M, Kluwe L, Friedrich RE, Funsterer C. MRI growth patterns of plexiform neurofibromatosis in patients with neurofibromatosis type 1. Neuroradiology 2006; 48:160-5.

5. Friedrich RE. Schmelzle R. Hartmann M, Mautner VF. Subtotal and total resection of superficial Plexiform neurofibromas of face and neck: four case reports. J Craniomaxillofac Surg 2005; 33:55-60.

6. Serletis D, Parkin P, Bouffet E, Shroff M, Drake JM, Rukta JT. Massive Plexiform neurofibromas in childhood: natural history and management issues. J Neurosurg 2007; 106:363-7

7. Jackson IT. Neurofibromatosis of the skull base. Clin Plast Surg 1995; 22:513-16.

8. Park BY, Hong JP, Lee WJ: Netting operation to control neurofibroma of the face. Plast Reconstr Surg 2002; 109:1228 – 36.

9. Marchac D, Britto JA. Remodelling the upper eyelid in the management of orbitopalpebral neurofibromatosis. Br J Plast Surg 2005; 58:944-56.

10. Poyhonen M, Niemela S, Herva R. Risk of malignancy and death in neurofibromatosis. Arch Pathol Lab Med 1997; 121:139-43.