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A solitary huge neurofibroma of the soft palate

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Abstract
Neurofibroma of the soft palate, an extremely rare tumour, is probably not yet reported in the literature. We report the first case of an isolated neurofibroma of the soft palate not associated with von Recklinghausen's disease (VRD), which is also, probably, the first reported case of neurofibroma of the soft palate. The tumour was completely removed from the soft palate after performing tracheostomy, necessitated due to difficulties in intubation.

Key words: Palate, Soft; Neurofibroma

Introduction

Tumours of the peripheral nerve are commonly benign and include schwanommas and neurofibromas. Neurofibromas may occur either multiply or solitarily. When solitary (one or two lesions), they are spontaneous tumours without any internal manifestations; if three or more are present, it is termed as neurofibromatosis.[1] In brief, neurofibromatosis (von Recklinghausen's disease, VRD) is an autosomal dominantly inherited syndrome. It is classified into Type 1 (classical neurofibromatosis with prominent cutaneous manifestation), Type 2 (central or bilateral acoustic neuromas), Type 3 (mixed form), Type 4 (variant form resembles Type 2 but with numerous cutaneous neurofibroma), Type 5 (segmental, dermatomal NF). Type 6 (no neurofibroma, only *café au lait* spots) and Type 7 (late onset form).[2] The subtypes of neurofibroma are localized, plexiform and diffuse. Localized neurofibroma is solitary, polypoid or nodular, mostly cutaneous, and is usually not associated with neurofibromatosis. It is circumscribed but is an unencapsulated lesion with varied appearance.[3] Plexiform neurofibroma is composed of one or more adjoining nerves that have been transformed into neurofibromas, and sometimes, may become very large to form a tortuous mass leading to considerable disfigurement. Plexiform neurofibroma is considered virtually pathognomic of neurofibromatosis. Diffuse neurofibroma appears as a plaque-like area of subcutaneous thickening.[3,4]

Neurofibromas are usually associated with VRD and are multiple in number, but may also occur as a solitary tumour.[5] A review of the literature showed the occurrence of plexiform neurofibroma of the oral cavity and oropharynx as a rare manifestation.[6] Crozier[7] reported a patient with neurofibroma of the tongue and VRD, who developed severe upper airway obstruction during induction of anaesthesia. Her mouth opened adequately but intubation via both oral and nasal routes failed, as there was a large mass at the base of the tongue. Emergency cricothyroidectomy was performed as oxygen saturation dropped to 35 per cent. Isolated laryngeal neurofibroma not associated with VRD is even more unusual, and such a tumour has been removed by microlaryngoscopy.[8] In this paper, we report an isolated huge neurofibroma of the soft palate; most probably the first reported case.

Case report

A 50-year-old woman presented with the history of a gradually enlarging mass inside her mouth for two years at the otolaryngology clinic. Initially, it was small and nodular but was gradually increasing in size, resulting in difficulties in swallowing and speech. The swelling was otherwise painless. The patient often suffered from breathlessness during sleep.

Examination revealed a large polypoid mass (approximately 5 cm × 4 cm × 4 cm) arising from the right side of the soft palate and occupying most of the oral cavity. The overlying mucosa was intact; the mass was non-tender, firm, non-compressible and non-pulsatile with restricted mobility. The neck glands were not palpable and there was no swelling in other parts of the body. Systemic examination did not reveal any significant abnormality. The provisional diagnosis was minor salivary gland tumour.

After pre-operative investigations, the results of which were within the normal limits, excision of the mass was planned. However, tracheostomy was imperative as attempted oral and nasal intubation by a consultant anaesthetist was not successful, due to the huge mass inside the oral cavity; oxygen saturation dropped to 70 per cent and bradycardia developed. These changes resolved within a short time after administration of 100 per cent oxygen through the tracheostomy tube. Once the patient was stable, a curved incision was made on the mucosa of the soft palate anterior to the base of the tumour. The incision was further deepened through the submucosa, and meticulous dissection around the tumour was continued with gentle traction. The entire mass was removed from the soft palate with a rim of normal tissue around the tumour, resulting in a surgical defect. The bleeding was controlled by electro cautery, and the wound was repaired in layers. The patient was fed through a nasogastric tube for seven days; during this period, oral hygiene was maintained by frequent mouthwash with diluted hydrogen

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peroxide solution. Healing was satisfactory. There was no distortion of faucial arch and no alteration of velopharyngeal competence.

The excised tumour was an irregular multilobulated mass with a grayish white glistening surface. Histological examination showed a benign spindle cell tumour with wavy fibrillar materials. It consisted of elongated spindle cells, with wavy nuclei and poorly defined pale eosinophilic cytoplasm. There were also numerous short spindle cells and many small nerve fibres that were typical of localized neurofibroma (Figure 1).

Subsequently, the patient was meticulously examined and investigated for other stigmata of VRD. Family history showed that no members of the family suffered from neurofibromatosis. Ophthalmological examination showed no Lisch nodule in the iris. Pure tone audiometry and impedence audiometry tests were normal. CT scan of the head and neck and ultrasound of the abdomen were normal. There was no recurrence of tumour at nearly one and a half years follow-up.

Discussion

Yamada *et al.*[9] reported a seven-month-old male infant admitted with respiratory distress due to a huge sublingual mass. Multiple *café-au-lait* spots were also detected all over his body. A review of the family history revealed that his father had VRD. The infant died of respiratory failure; autopsy showed plexiform neurofibromatous nodule and extensive plexiform neurofibromas of vagus, recurrent larygneal and phrenic nerves. Recently, Sharma and Srinivasan[6] reported an isolated plexiform neurofibroma of the oropharynx as a rare manifestation of VRD; the neurofibroma was succesfully excised. Tsutsumi *et al.*[10] also presented a solitary plexiform neurofibroma of the submandibular gland. A rare case of neurofibroma of the cervical portion of the vagus nerve was presented by Galli *et al.*[11] where the tumour pushed medially the left lateral wall of the naso-, oro- and hypo-pharynx and the ipsilateral tonsil. The neurofibroma was removed through left cervical approach.

Our extensive search of the literature revealed that neurofibroma of the oropharyngeal region is very rare. To our knowledge, an isolated huge neurofibroma arising from soft palate without any other manifestations of VRD has not been reported. We report the first case of an isolated neurofibroma of the soft palate not associated with VRD probably the first reported case of neurofibroma of the soft palate. The tumour was very large and occupied most of the oral cavity. Histologically, it was a localized neurofibroma and there were no other evidences of neurofibromatosis. The treatment of choice for palatal tumours is to remove them by excising the portion of palate to which it is attached; this applies whether the tumour is situated in the hard or soft palate. Enucleation of any salivary tumour is liable to leave behind residual neoplastic areas and, in addition, approximately half of palatal tumours are malignant.[12] Because of the rarity of neurofibroma of soft palate along with absence of other manifestations of VRD, our provisional diagnosis was different ie. minor salivary gland tumour. Operative removal of this tumour imposed two types of technical difficulties: anaesthetic and surgical. Although in this case, we did not encounter any major surgical difficulties, there were potential chances of extensive damage to the muscles of the soft palate, leading to nasal regurgitation and difficulties in speech. However, the intubation failure was a real one and could have been fatal. Therefore, preparedness for tracheostomy was mandatory in such a situation.

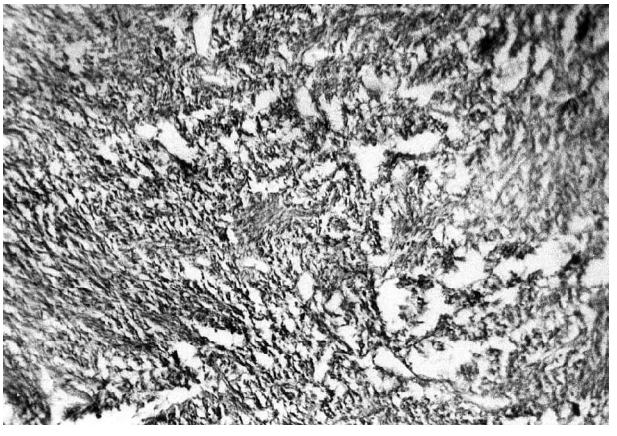

FIG. 1

Tumour composed of elongated spindle cells, with wavy nuclei and poorly defined pale eosinophilic cytoplasm. There were also numerous short spindle cells and many small nerve fibres suggestive of neurofibroma. H & E; ×100.

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