## Solitary parotid plexiform neurofibroma - diagnostic difficulty in a clinically unsuspected case

Sir,

A 15-year-old girl presented to the OPD with the complaints of a painless, progressively increasing swelling present at the angle of right jaw for the last two years. Patient did not have any cutaneous, ocular or skeletal manifestations of neurofibromatosis-1 (NF-1). None of the family member had any stigmata of NF-1. Her physical examination revealed a large, lobulated, firm, non tender swelling at the angle of mandible. Examination of the oral cavity revealed vague fullness in the floor of mouth with healthy oral mucosa with House-Brackman grade III ipsilateral facial palsy. There was no palpable cervical lymphadenopathy. Clinical impression of a benign parotid tumor was made.

To ascertain the nature of the lesion, CT scan was performed which showed an irregular heterogeneously enhancing, encapsulated mass lesion involving both the lobes of right parotid gland extending into the available tissue spaces with no evidence of underlying bone erosion [Figure 1]. On MRI, the lesion showed areas of low to intermediate signal intensity on T1W sequence and high signal intensity on T2W sequence with heterogeneous enhancement [Figure 2]. Mass was biopsied and

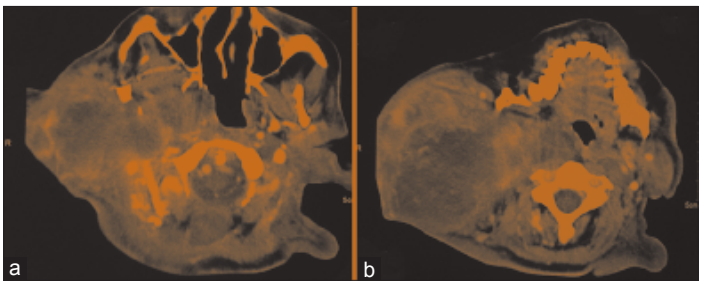


**Figure 1: (a,b) CT scan shows a large well defined, encapsulated, heterogeneously enhancing soft tissue density lesion in the region of the right parotid involving both the lobes, extending into parapharyngeal and retropharyngeal space upto pharyngeal mucosal space**

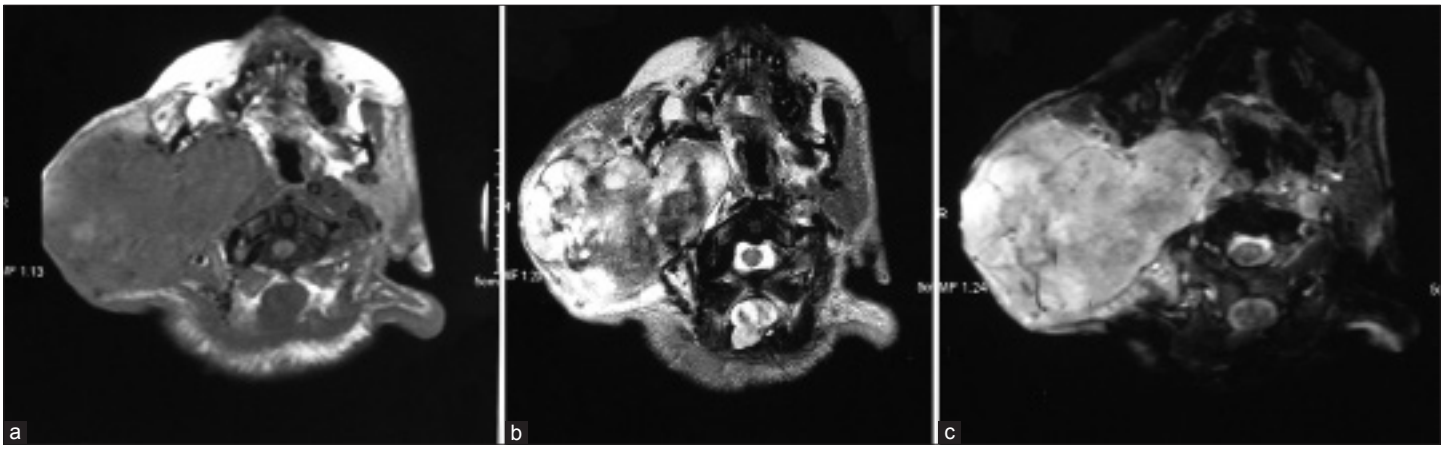


**Figure 2: (a) T1W MR sequence shows a well defined, encapsulated, heterogeneously enhancing soft tissue lesion of the right parotid involving both the lobes, extending medially upto pharyngeal mucosal space displacing adjacent vessels and bones. (b) T2W sequence shows heterogeneously hyperintense mass lesion. (c) GRE sequence shows no evidence of blooming artifact suggesting absence of any bleed or calcification**

histopathological examination revealed a benign lesion composed of proliferating fibroblast with few nerve fibres in a background of myxoidstroma consistent with plexiform neurofibroma.

After pre-operative counseling about the risk of facial nerve damage, total parotidectomy with en-bloc resection of the mass was performed. Per-operatively the mass was well encapsulated, red-brown colored and was found adhered to the thickened facial nerve, which was inevitably severed. Post-operatively, complete facial nerve palsy was noted and patient was advised physiotherapy with continuous follow-up.

Neurofibroma originating from the intraparotid facial nerve is very rare. Till date, in literature primary intraparotid facial nerve involvement by a plexiform neurofibroma is reported in only 9 cases,[1] of these only one patient did not have any stigmata of NF-1 or any family history.[2] Neurofibroma can be solitary or occur in association with NF-1. The solitary neurofibroma, by definition occurs in patients who do not have NF-1. There are no characteristic symptoms of parotid neurofibroma; however, pain, tenderness, facial spasm or paralysis may occur.

Neurofibromas and Schwannomas are the most common neurogenic tumors found in head and neck, common sites of involvement being the vagus nerve, ventral and dorsal cervical roots and sympathetic trunk. Neurofibromas are unencapsulated nerve sheath tumors composed of schwann cells, perineural cells and fibroblasts in a collagenous and mucoid matrix,[3] they are histologically benign and classified as WHO grade-I tumor. Neurofibromas and schwannomas cannot be completely differentiated on the basis of sectional imaging, in which case a clinical history of neurofibromatosis helps. The main role of imaging and MRI is in defining the extent of the disease process which aids in surgical planning.[4]

The rarity of the parotid plexiform neurofibroma has limited the clinical, radiological and histopathological description of the tumor. The features seen in the reported cases including the present case shows that they are locally infiltrative benign tumors that tend to grow slowly. Although they may grow any time, growth spurts are particularly seen in early childhood and during puberty or pregnancy.

Treatment modality depends upon the size of the lesion and status of facial nerve. Some authors suggest serial follow-up by CT and electroneurography while others prefer early surgical repair.[5] Larger tumors need resection of the nerve and cable graft by a peripheral nerve.

Thus, solitary parotid plexiform neurofibroma is a rare parotid neoplasm requiring a high index of suspicion to diagnose it preoperatively. Imaging features although suggestive are not definite for diagnosis which requires histopathological examination.

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