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Case Report

A Rare Cause of Cervical Swelling: Solitary Neurofibroma

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Abstract

Neurofibroma in the head and neck region is a rare entity and is difficult to diagnose both clinical and radiological. It presents as solitary slowly progressing tumor or as a part of neurofibromatosis disease. We, hereby, report a case of 27-year-old female admitted for a left sided lateral neck swelling, for which the surgical indication was aesthetical impairment. The pathological study was in the favor of neurofibroma. Multiple neurofibroma in the head and neck region may occur in the skin as part of neurofibromatosis and as solitary lesions in the region of the neck. Even they are rare, they should be considered as a differential diagnosis of a neck tumor. Surgery remains the gold standard in the treatment of these types of tumors. It is essential that the surgeon keep in mind the possibility of these tumors as a differential diagnosis of lateral neck swellings.

Key words: Cervical mass, Head and neck, Neurofibroma

INTRODUCTION

A neurofibroma is a benign tumor of peripheral nervous system arising from nerve sheath. Skin (cutaneous neurofibroma) and peripheral nerves (solitary neurofibroma) are common site of its occurrence. These can present sporadically or in association with neurofibromatosis type 1 (NF1), which is an autosomal dominant genetically inherited disease.[1] Neurofibromas arise from Schwann cells which are non-myelinating. These cells exhibit biallelic inactivation of NF1 gene that codes for the protein neurofibromin.[2] The clinical presentation is not specific. Its clinical presentation can be confused with a vascular tumor or malformation, a skin or connective tissue tumor, benign, or malignant.[3] Neurofibroma in the head and neck region is not common. Here, we present one rare case of neurofibroma in the left cervical region.

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CASE DESCRIPTION

A 27-year-old female patient presented to ENT outpatient department with a history of swelling in the left lateral aspect of the neck for the past 10 years. The patient had irrelevant medical history with no known allergies. The swelling was gradually progressive with no associated history of pain. On local examination, there was 8 cm × 4 cm, firm in consistency, non-tender, and non-pulsatile mass in the left lateral aspect of the neck. CECT scan of the neck shows a well-defined thin-walled hypodense lesion measuring 6.3 cm × 8 cm × 10 cm (AP*TR*SI) with numerous internal septae in the left lateral aspect of the neck, medially, and deep to the sternocleidomastoid extending opposite C2 to D3 vertebral levels. Medially, it was displacing the left lobe of thyroid and abutting the left lateral wall of esophagus; however, the fat planes were maintained. Anteromedially, it was also displacing the CCA and ECA and further insinuating into the prevertebral space causing asymmetric bulge in the pharyngeal wall with narrowing of the left pyriform sinus [Figure 1].

Fine-needle aspiration cytology of the mass was undiagnostic and showed blood only. Surgical exploration was planned and the patient was taken for surgery under general anesthesia. Intraoperatively, the mass was grey white

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in color, encapsulated, and glistening measuring 10 cm × 6 cm × 5 cm [Figures 2 and 3]. On the cut surface, it appears mucoid and relatively avascular.

Histopathological examination report showed the tumor mass composed of carrot shredded appearance of stroma with spindle-shaped wavy hyperchromatic nuclei suggestive of benign mesenchymal tumor possibly neurofibroma [Figure 4]. The post-operative period was uneventful. The suture of the wound was removed on the 7th post-operative day and the patient was discharged in satisfactorily condition.

DISCUSSION

A neurofibroma in the head and neck region is very rare. Haller first discovered the carotid corpuscle in 1742. Rodier introduced the term "neuroma" for tumors of the peripheral nerves in 1803.[4] Neurofibromas are slowly progressive benign tumors. These tumors can cause compression on adjacent vital structures or interfere with normal physiology.[5] The presentation of plexiform neurofibroma is uncommon, usually presenting at birth or in the 1st years of life.[6] Neurofibromas usually occur in viscera, solitary cervical presentation is rare. Solitary neurofibromas in cervical region usually present as slowly progressive painless swelling in neck without any other symptoms. Hence, these tumors are commonly ignored by patients, as in our study, neck mass was ignored by the patient for 10 years. Clinical and radiological diagnosis is difficult in these patients due to no specific symptoms and radiological specific features pertaining to this tumor and rarity of this tumor in this location.[7] In our case, specific diagnosis of neurofibroma was not established clinically or radiologically. Recent advances in immunohistochemistry allow increasingly precise diagnosis.[8,9] The majority of neurofibromas are benign, but malignant transformation may occur.[10] Most of these tumors can be quite disfiguring

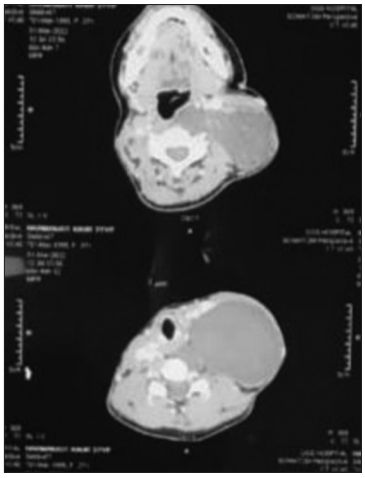

Figure 1: Axial contrast-enhanced computes tomography scan showing the location of tumor in the left lateral aspect of the neck

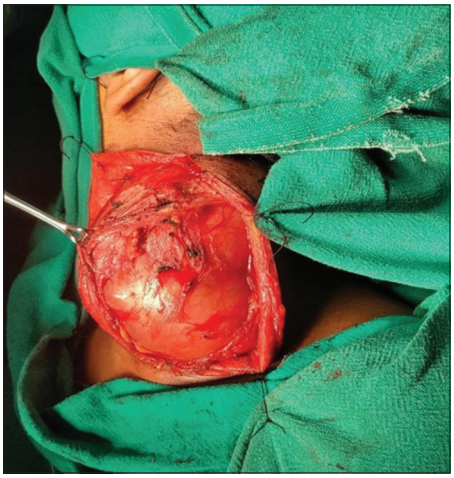

Figure 2: Surgical exploration of the tumor intraoperatively

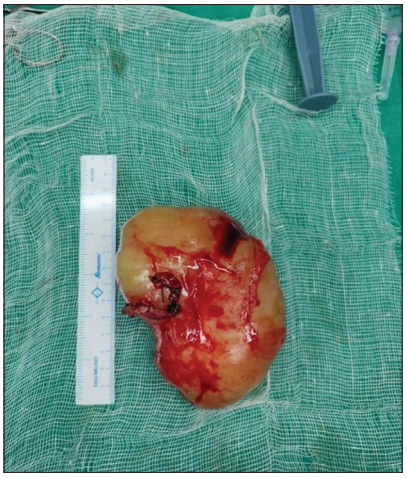

Figure 3: Excision of the mass

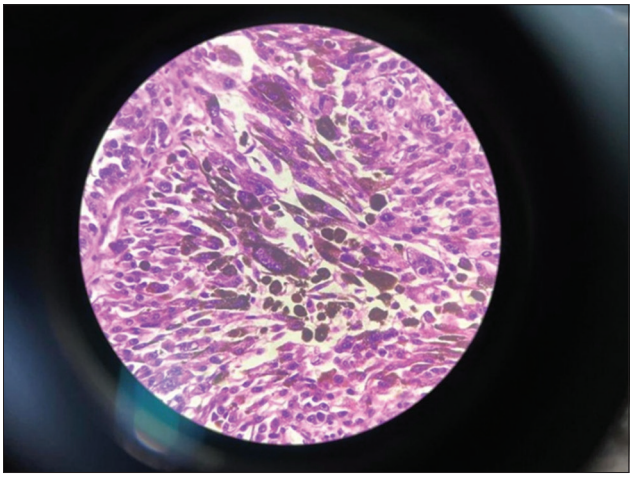

Figure 4: Histopathological slide image showing carrot shredded appearance of stroma with spindle-shaped wavy hyperchromatic nuclei

and are rarely known to cause symptoms ranging from minor discomfort to extreme pain. In our case, the only complaint patient was presenting was the disfigurement. The consistency of the lesion is like a "bag of worms" due to the presence of soft areas interspersed with firm nodular areas. These lesions sometimes are vascular in nature. At histological analysis, a localized and solitary neurofibroma is composed of interlacing fascicles of wavy, elongated cells that often contain abundant amounts of collagen. Rarely, myxoid areas and degenerative regions could be found in neurofibromas. Diffuse neurofibroma contains very uniform, prominent fibrillary collagen. Both localized and diffuse neurofibromas are positive for S-100 protein at immunohistochemical analysis, but this is not a stable finding.[11,12] The treatment of localized and diffuse neurofibromas (not associated with NF-1) is often surgical excision. Sudden increase in size of a previously diagnosed neurofibroma should be viewed with great suspicion of malignant transformation and must be considered for immediate biopsy. The estimated prevalence of malignant transformation varies from 2% to 29%, but they are not well documented in the literature.[13] In our case, tumor was slowly progressing over 10 years, so we did complete excision and sent for histopathological examination.

CONCLUSION

Neurofibroma in the head and neck region is a very rare clinical entity, but it should be considered in the differential diagnosis of head and neck swellings. Multiple neurofibroma in the head and neck region may occur in the skin as part of neurofibromatosis and as solitary lesions in the region of the neck. They are rare tumors with low risk of malignant transformation. If local conditions and the general condition of the patient allow surgery, complete resection of the lesion remains the gold standard in the treatment of these locally invasive tumors.

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We confirm that this manuscript has not been published elsewhere and is not under consideration by another journal.

All the authors have approved the manuscript and agreed to the submission.

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