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A Huge Plexiform Neurofibroma in a 20 Months Old Child: Case Report

20 Aylık Çocukta Dev Pleksiform Nörofibrom: Olgu Sunumu

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ABSTRACT

Plexiform neurofibroma is a rare, benign, peripheric nerve tumor. In the literature, there has been very few reports of it in the childhood period, especially with huge size which is not resectable totally. We herein presented a 20-months old male patient with plexiform neurofibroma. The tumor overfilled in every possible space in the head and neck region and it was not available for total resection. Therefore, an initial conservative-supportive treatment plan was considered. The aim of the present case report was to discuss the management of a massive pediatric head and neck tumor.

Keywords
Plexiform neurofibroma; pediatric; head and neck tumor; treatment; tracheotomy

ÖZET

Pleksiform nörofibrom nadir görülen, iyi huylu, periferik sinir tümörüdür. Literatürde, çocukluk çağında, özellikle de total olarak rezeke edilemeyecek ölçüde dev boyutlarda olan çok az olguya rastlanmaktadır. Bu olgu sunumunda pleksiform nörofibromlu 20 aylık bir erkek hasta sunulmuştur. Tümör baş ve boyun bölgesinin bütün olası boşluklarını doldurmaktaydı ve cerrahi olarak total rezeksiyona izin vermeyecek büyüklükteydi. Bu nedenle, ilk aşamada konservatif ve destekleyici bir tedavi planı düşünüldü. Bu olgu sunumunun amacı dev boyutlu bir pediatrik baş boyun tümörüne yaklaşımın tartışılmasıdır.

Anahtar Sözcükler
Pleksiform nörofibrom; pediatrik; baş boyun tümörleri; tedavi; trakeotomi

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INTRODUCTION

Plexiform neurofibroma is a benign, peripheric nerve tumor which originates from Schwann cells. Mostly, it is found as a component of Neurofibromatosis Type 1 (NF-1). It does not metastasize but local recurrence is very often, Male/Female ratio is 3:2 and it is most commonly found in head and neck region (%14-37).[1] Malignant transformation is rare (%10-15) and mostly seen in NF-1 patients who have big and deeply seated tumors.[1] The main treatment is surgery. In literature, reports of plexiform neurofibromas are rare and majority of the reported cases are young adults. We herein presented a very young child who had a huge-sized plexiform neurofibroma that was not resectable totally.

CASE REPORT

20-months old male patient referred to our clinic with dyspnea, snoring and a diffuse mass occupying both sides of the neck which enlarged over the previous 2 months. He was born after 8 months of gestation by Caeserian section due to intrauterine exitus of his twin. When he was 3 months old, a biopsy was taken from the mass under his tongue and it was reported as nerve sheath mixoma. It regressed spontaneously without therapy but when he was 6 months old his family noticed small masses in his neck which grew progressively. In the physical examination, there was a diffuse, solid mass which overfilled all neck zones bilaterally (Figure 1). The rest of the otolaryngologic and systemic examination was normal. We did not detect any cranial nerve paralysis or neurological deficits. Any intraabdominal lymph nodes or hepato-splenomegaly were ruled out by abdominal ultrasonography. Tracheotomy was performed in order to secure the airway and relieve dyspnea. Another biopsy was taken transcervically which was reported as plexiform neurofibroma (Figure 2a, 2b). Cervical MRI revealed a solid mass without definite borders bilaterally at neck region which overfilled the parotis, submandibuler and sublingual spaces, reaching

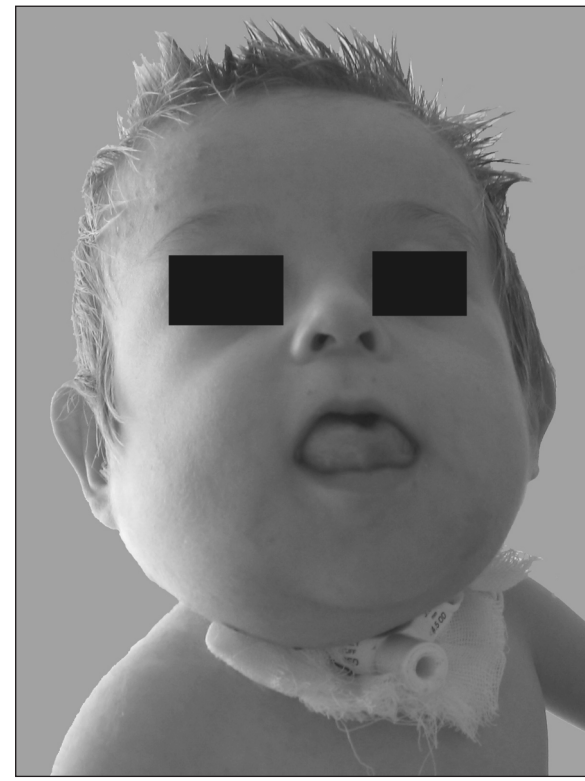

Figure 1. Anterior view of the patient after tracheotomy.

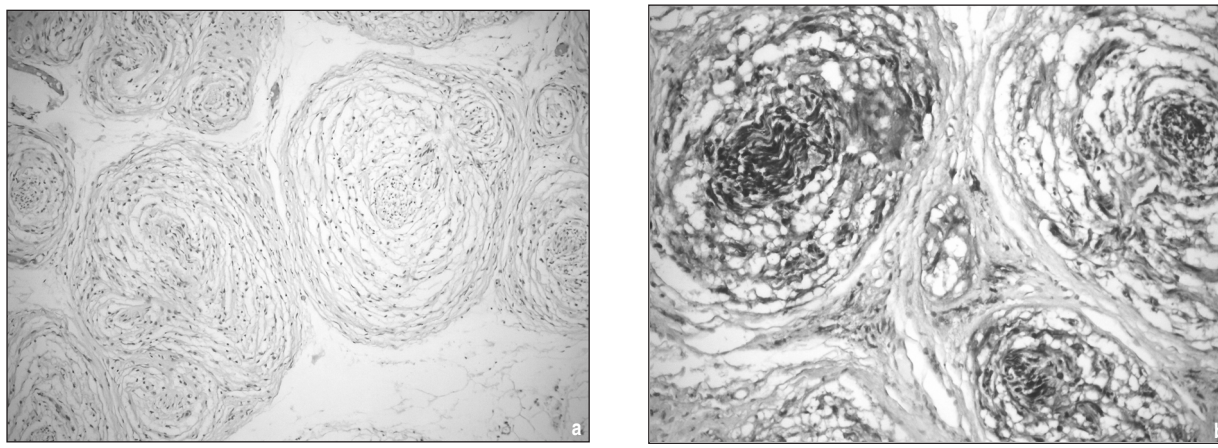

Figure 2. a) Plexiform neurofibroma with tortuous enlargement of the nerves (HE x200), **b)** Positive S-100 staining of the neural sheath (S-100 x 200).

to tonsilla pharyngica, right side of the root of tongue and tip of tongue, and obliterated the nasopharynx (Figure 3a, 3b). Moreover, it occupied the prevasculer space at the entrance of the thorax. The diagnosis of NF-1 was ruled out as he did not have Lisch nodules, freckling in the axillary regions, cafe au lait macules and bone deformities. It was decided to postpone surgical treatment and he was put under follow-up with 3 months intervals. A written consent was obtained from the parents for use of medical data and photographs of the patient.

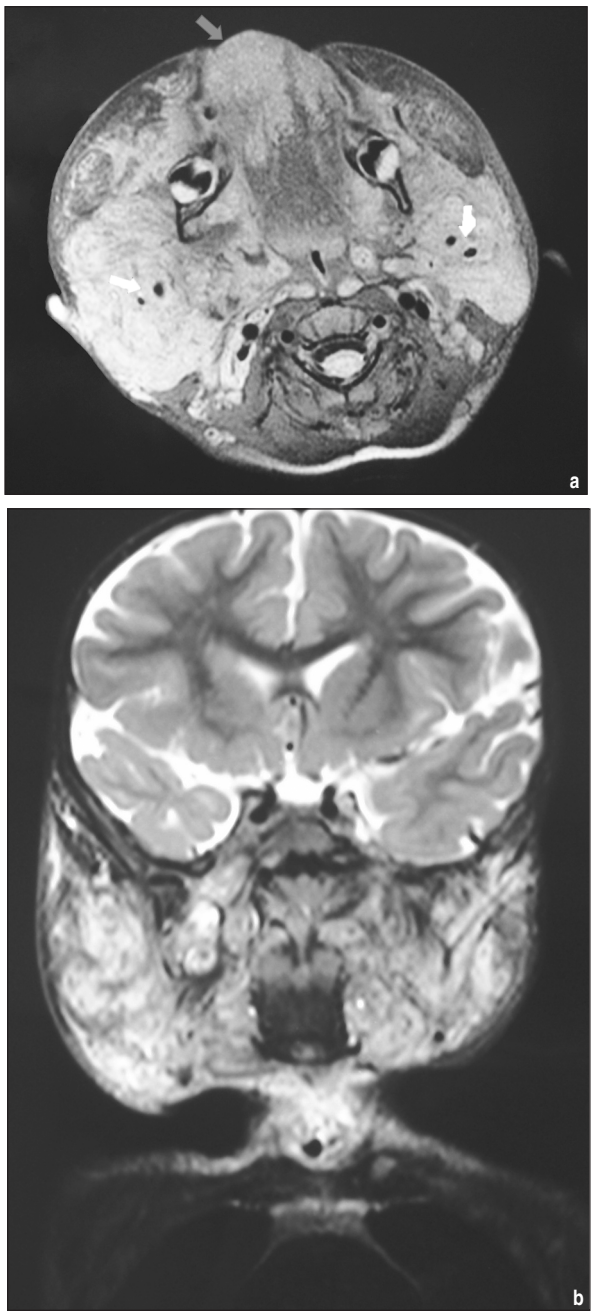

Figure 3. a) The view of the mass in axial neck MRI. Black arrows show carotid arteries which were surrounded by mass and white arrow shows tongue invasion, **b)** The view of the mass in coronal neck MRI. Invasion of all spaces of head and neck area is visible.

DISCUSSION

Plexiform neurofibroma is one of the diagnostic criteria of NF-1, but it can also be detected in patients who do not have NF-1. Although, it is a benign tumor, it often shows local recurrence. Malignant transformation is rare, however, it can cause organ dysfunctions, morbidity, decreased quality of life and aesthetic problems related to its localization and size.

The diagnosis of NF-1 depends on clinical examination namely, cafe au lait macules, Lisch nodules, freckling in the axillary regions and neurofibromas. Detection of NF-1 gene at the long arm of chromosome 17 which encodes neurofibromin protein would support the diagnosis.[2]

In the differential diagnosis, the most common causes of pediatric head and neck masses should be excluded. Lymphomas are one of the most common causes of mass in pediatric patients at head and neck region. Presence of systemic symptoms may help in differential diagnosis, lymphpomas usually involve only lymph node regions but such a diffuse involvement does not exclude the diagnosis. Rhabdomyosarcoma is an other common pediatric tumor that should be considered in the differential diagnosis. Of the congenital lesions, lymphangiomas and hemangiomas may present with a diffuse mass in head and neck region, however, both lymphangiomas and hemangiomas are usually soft and in hemangiomas red discolorisation may happen if dermal tissues are involved. Definitive diagnosis of the type of tumor depends on pathological examination. Tortuous enlargement of the neural tissue and positive staining with S-100 are diagnostic for the neurofibromas. In the differential diagnosis of NF-1, tuberosclerosis may be excluded by its macules being hypopigmented. Malignant forms are ruled out by low mitotic index in KI-67 staining and hypocellularity of the tumor. Clinically, a sudden enlargement of the mass or emergence of a cranial nerve paralysis during the follow-up should alert the physician for malignant transformation.

Main treatment is surgery because it is radio-resistant and not very sensitive to chemotherapeutics because of its low rate of cell proliferation.[3] On the other hand many authors think that surgery is very difficult in that

kind of tumors and it often recurs and grows faster than before, following surgery.[4,5] In the previous studies it was reported that recurrence was less than 40% after subtotal resection, whereas, it was less than 20% after total resection. Recurrence rate is especially much higher in patients under 10 years old.[3] Therefore, many surgeons are against surgery especially in children, if tumor is localized at head and neck region.[6]

The present case was unique for having such a big tumor in such a young age. We decided not to operate on this patient at the moment, because total excision was not possible and partial resection had a high risk for more aggressive course. Therefore, we offered only supportive treatment including tracheotomy until he grew older, and drawbacks related to young age ceased. By then, even a palliative surgery might offer success.

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