European Annals of Otorhinolaryngology, Head and Neck diseases 133 (2016) 47–50

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Case report

A rare case study of a retropharyngeal neurofibroma and a brief literature review

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ARTICLE INFO

Keywords:
Neurofibromas
Neurofibromatosis
Surgical resection
CT-scan
MRI
Malignant transformation

ABSTRACT

Introduction: Space occupying lesions in the retropharyngeal space are rare.
Case report: Here, we present a rare case of a retropharyngeal neurofibroma treated surgically through an open cervical approach without any complication.
Discussion: Neurofibromas are benign, slowly growing neoplasms that could be associated or not with neurofibromatosis. They are derived from peripheral nerves. Diagnostic work-up should include CT-scan and MRI as well as a biopsy to confirm the diagnosis. Treatment of localized and diffuse neurofibromas is often surgical resection, which may require sacrifice of the nerve. Malignant transformation of these lesions, without association to NF-1, is rare and exceptionally documented in the literature.
Conclusion: We presented a rare case of solitary neurofibroma of the retropharyngeal space. Complete resection of the lesion was performed without any complication. We also presented a brief review of the literature about neurofibromas.

1. Introduction

Space occupying lesions in the retropharyngeal space are rare. Various masses have been described including angiomyoma, ganglioneuroma, malignant mesenchymoma, and neuroblastoma, but the commonest lesions are hematomas [1]. The clinical presentation is varied but most have presenting symptoms of dyspnea, dysphagia, and obstructing sleep apnoea. Here, we present a rare case of a retropharyngeal neurofibroma treated surgically. In the third section of the article, in addition to the case discussion, we also present an overview of neurofibromas clinical presentation, diagnosis and management.

2. Case report

A 64-year-old male patient was referred to our otolaryngology clinic in October 2014 for an accidental discovery of a retropharyngeal lesion during a diagnostic work-up performed for a posterior neck pain with irradiation to the occipital region. The patient had an irrelevant past medical history with no known allergies. He had a history of smoking (19 pack-years) with occasional alcohol consumption. The patient underwent CT-scan as well as a MRI. Imaging showed a retropharyngeal lesion measuring 5 × 4.5 × 2.8 cm at the level of C4, C5, and C6 vertebrae (Fig. 1). Clinical examination revealed a normal retropharyngeal mucosa with a submucosal non-pulsatile swelling in the hypopharynx. The vocal folds' mobility was preserved and there was no clinically palpable lymphadenopathy. The chest CT-scan was normal. A pharyngoscopy was performed under general anaesthesia with a biopsy through a mucosal incision that revealed a whitish-gray hard mass. Histological examination confirmed a neurofibroma (Fig. 2). One month later, resection of the lesion was performed through a left lateral cervicotomy under general anaesthesia with an orotracheal intubation (Fig. 3). The mass was bilobulated and was adherent to the intervertebral disc between C4 and C5. Complete macroscopic resection was performed without any preoperative or postoperative complication. The patient restored oral feeding two days postoperatively with no inhalation or difficulty in swallowing and was discharged from hospital four days after surgery. A CT-scan and a MRI were performed six months later and showed no signs of recurrence.

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http://dx.doi.org/10.1016/j.anorl.2015.11.003
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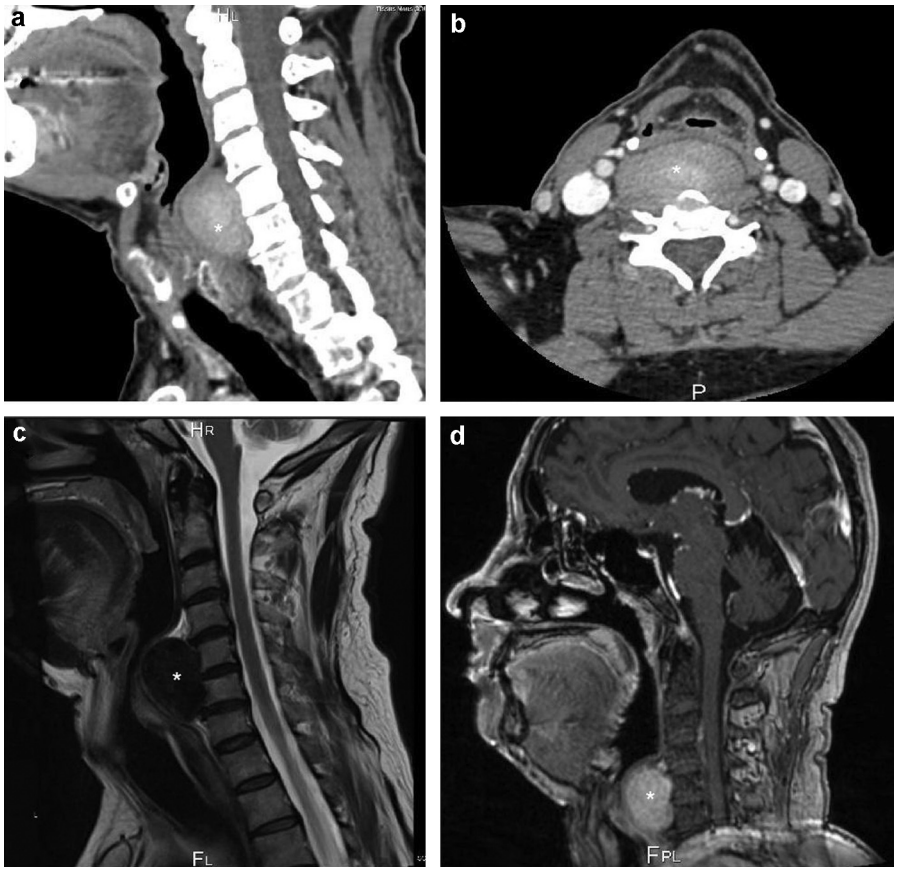

Fig. 1. Imaging of the retropharyngeal neurofibroma (star). CT-scan sagittal section (a) and axial section (b) with contrast injection showing a prevertebral lesion with low contrast enhancement. T2-weighed MRI sagittal slice (c) showing the same lesion with low signal intensity. The lesion shows enhancement after gadolinium injection on T1-weighed MRI sagittal slice (d).

3. Discussion

Neurofibromas most frequently affect patients who are 20–30 years old, and they have no sex predilection. There are three subtypes of neurofibroma: localized, diffuse, and plexiform. Localized neurofibromas are most likely to arise from cutaneous nerves, with the occasional involvement of deep-seated nerves. These neoplasms are benign, slowly growing, and relatively circumscribed, but are not encapsulated. They consist of Schwann cells and perineural cells, and have varying amounts of mature collagen [2–4]. Localized neurofibromas tend to be large. In the setting of neurofibromatosis type 1 (NF-1), they tend to be multiple and more commonly deep in location. In 60%–90% of cases, like in our case report, they occur in patients who do not have NF-1. In such cases, they are termed solitary neurofibroma [5]. Plexiform neurofibroma is said to be indicative of NF-1, even though it may be the only manifestation of the disease [2]. Plexiform neurofibromas are pathognomonic for this entity, usually involving a long segment of a major nerve trunk and extending into the nerve branches. Diffuse neurofibroma is an uncommon subtype of neurofibroma. It has been reported to occur most commonly among children and young adults, typically involving the skin and subcutaneous tissues of the head and neck. Unlike other types of neurofibroma, which have a mass-like pattern of growth, diffuse neurofibroma is a poorly defined lesion that spreads along connective tissue septa and surrounds rather than destroys adjacent normal structures [4]. All three types of neurofibromas (localized, diffuse, and plexiform) can be associated with NF-1 [2,3]

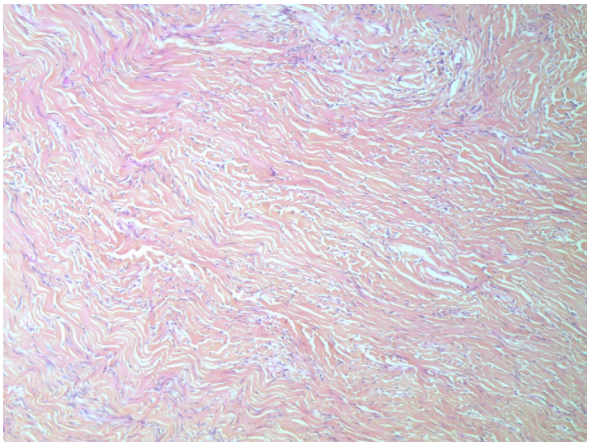

Fig. 2. Pathological analysis of the retropharyngeal neurofibroma. Spindle-shaped cells arranged in loose bundles within a matrix rich in collagen and mucopolysaccarides (Haematoxylin-eosin staining, original magnification: 10 × objective lenses).

On radiological imaging, solitary neurofibroma manifests as a well-delineated smooth margin predominantly solid fusiform mass [3,6,7]. On CT-scan, neurofibroma appears hypodense relative to muscle and usually shows little or no contrast enhancement. On MRI, neurofibromas exhibit low to intermediate signal intensity on T1-weighted images and high signal intensity on T2-weighted images, with a heterogeneous or a homogeneous pattern. Inhomogeneous contrast enhancement is seen in two thirds of cases after administration of contrast agent [2,8].

At gross examination, localized neurofibroma represents a mass with an entering and an exiting nerve [2]. In our case, we did not find