CASE STUDY

Solitary Neurofibroma of the Maxillary Sinus☆

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KEYWORDS
Neurofibroma; Maxillary sinus

Abstract Neurofibromas are benign tumours of peripheral nerve tissue, frequently associated with neurofibromatosis type I. Their isolated occurrence in the maxillary sinus is rare, with only 6 cases described in the literature. We present the case of a 70-year-old woman who consulted for left hemifacial paresthesias of 10 years’ evolution. CT showed a heterogeneous lesion in the left maxillary sinus, eroding its lateral wall without signs of infiltration. Endoscopic surgery with nasalisation of the maxillary sinus was performed. The pathology was neurofibroma (positive for S-100 protein and vimentin). After 18 months, no tumour growth has been found.

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PALABRAS CLAVE
Neurofibroma; Seno maxilar

Resumen Los neurofibromas son tumores benignos del tejido neural periférico, más frecuentes asociados a neurofibromatosis tipo I. La aparición aislada en un seno maxilar es excepcional, estando descritos sólo 6 casos en la literatura. Presentamos una mujer de 70 años que consultó por parestesias hemifaciales izquierdas de 10 años de evolución. La TC mostró una lesión heterogénea en seno maxilar izquierdo que erosionaba la pared lateral del mismo sin signos de infiltración. Se intervino mediante endoscopia, nasalizando el seno maxilar. La anatomía patológica fue de neurofibroma (proteína S-100 y vimentina positivos). Tras 18 meses no hemos encontrado crecimiento tumoral.

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Introduction

Neurofibromas are benign tumours of peripheral nerves that arise from the connective tissue of their sheaths, especially the endoneurium. They are much more frequent in association with neurofibromatosis type 1, also known as von Recklinghausen’s disease, although isolated cases may also appear. Isolated neurofibromas of
the maxillary sinus are extremely rare. Through the presentation of this case, we aim to describe the clinical, histopathological and radiological characteristics of these tumours.

Clinical Case

We describe the case of a 70-year-old woman who attended consultation due to left hemifacial discomfort, with over 10 years of evolution. Endoscopic examination of the nose revealed no abnormalities. The CT scan showed a heterogeneous lesion in the left maxillary sinus eroding the lateral wall, with no signs of soft tissue infiltration (Fig. 1). The MRI scan showed an expansive lesion in the left maxillary sinus, isointense in T1 and hyperintense and heterogeneous in T2; the lesion interrupted the lateral wall, with no signs of soft tissue infiltration (Fig. 2). Faced with seemingly benign clinical and radiological symptoms, the patient was advised to undergo an endoscopic intervention for lesion biopsy and excision, within the possible limits (in addition, the patient had refused to undergo a more aggressive intervention). The intervention was carried out by nasalising the maxillary sinus (with resection of the inferior turbinate). The anatomical pathology was neurofibroma (tumour composed of a myxoid stroma and spindle cellularity, with scant, poorly defined cytoplasm, with some areas of plexiform pattern and other more collagenised ones, positive for S-100 protein and vimentin, with no signs of malignancy) (Fig. 3). The postoperative course did not have any complications. After 18 months, there was no evidence of tumour growth in the control MRI scan.

Discussion

Neurofibromas are benign tumours originating from the peripheral neural tissue, which are frequently associated with neurofibromatosis type 1, such as Von Recklinghausen’s disease, although they can occur in isolation. Nasal and paranasal neurofibromas arise from the ophthalmic and
maxillary division of the trigeminal nerve. The symptoms are non-specific and depend greatly on the exact location and extent of the injury. They are, thus, often clinically silent, reaching considerable size in this location before diagnosis.1 Trigeminal nerve tumours often produce no neurological deficit. When found within the paranasal sinuses, bone destruction is a common feature of these lesions. Neurofibromas are usually iso- or hypointense on T1-weighted images and iso- or hyperintense on T2-weighted images, with heterogeneous contrast uptake.2 Macroscopically, they are not encapsulated. Microscopically, they present immunoreactivity for S-100 protein and vimentin. Complete surgical resection is the treatment of choice.3 Monitoring is required because the lesion may recur. Solitary neurofibromas rarely become malignant. Isolated occurrence in a maxillary sinus is exceptional, with only 6 cases described in the literature reviewed (one of them, bilateral).4

Conflict of Interests

The authors have no conflicts of interest to declare.

References