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A Rare Localization of Solitary Neurofibroma: A Case Report

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Abstract **Case Report**

Neurofibromas are benign connective tissue tumors that develop mainly at the expense of the endoneurium of peripheral nerves. The head and neck region is one of the sites of neurogenic tumors such as neurofibromas. Usually part of a neurofibromatosis, solitary localization in the naso-sinus cavities is rare, estimated at 4% of head and neck localizations. We report the case of an 8-year-old child hospitalized in our department for a nasal tumor, radiographic Images revealed a nasoethmoidal tumor with right orbital extension and bilateral temporoparietal schizencephaly with open cleft. Despite its rarity, solitary neurofibroma must be taken into account in the differential diagnosis of a unilateral benign tumour of the naso-sinus cavities.

Keywords: Neurofibromas, nasal, tumor, schizencephaly.

INTRODUCTION

Neurofibroma is a benign connective tissue tumor that develops mainly at the expense of the endoneurium of peripheral nerves. The head and neck region is one of the sites of neurogenic tumors such as neurofibromas. Usually part of a neurofibromatosis, solitary localization in the naso-sinus cavities is rare, accounting for an estimated 4% of head and neck localizations. Neurofibromas are thought to develop from the 1st and 2nd dividing branches of the trigeminal nerve, destined for different structures. Despite its rarity, solitary neurofibroma must be taken into account in the differential diagnosis of a unilateral benign tumour of the naso-sinus cavities.

CASE REPORT

We report a social case of an 8-year-old child abandoned by his parents, who was hospitalized in our department for a nasal tumor evolving very progressively since birth. The child presented a psychomotor retardation with absence of walking and language, a radiological extension assessment including IMR and CT scan revealed a naso-ethmoidal tumor with right orbital extension, and a bilateral temporoparietal schizencephaly with an open cleft (Figure 1 & 2).

An excision of the tumor was performed by external nasal approach and histological examination confirmed the diagnosis of neurofibroma.

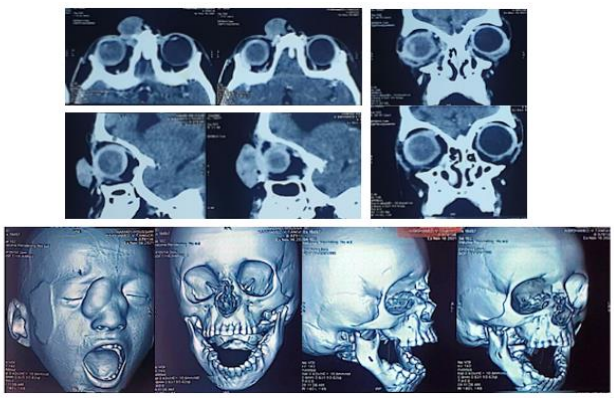

Figure 1: CT scan images of the neurofibroma nasal tumor in axial, coronal, sagittal and 3D sections

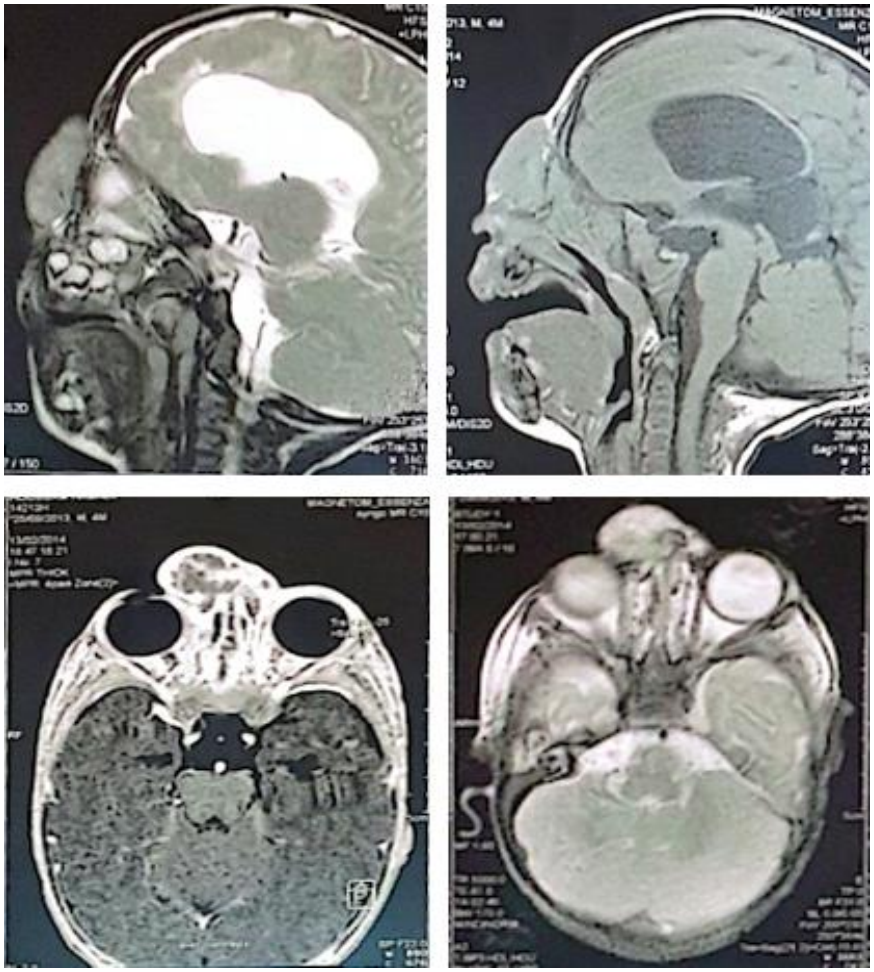

Figure 2: IMR images revealing the tumor and the temporoparietal schizencephaly

DISCUSSION

A neurofibroma is a benign tumor arising from the connective tissue of peripheral nerves, particularly the endoneurium. Neurofibromas usually occur as part of typeI neurofibromatosis, but can also be isolated, in which case they are referred to as solitary neurofibromas. Solitary localization in the naso-sinus cavities is rare (less than 4% of cervico-facial localizations) [1, 2], with the turbinates or maxillary sinus regularly reported as the preferred site [2, 3]. Neurofibromas arise from the 1st and 2nd dividing branches of the trigeminal nerve, destined for various structures in the naso-sinus cavities, notably the septum, turbinates and maxillary sinus. There is no predilection for gender, with an average age at presentation of 46 years. Patients typically present a massive lesion evolving for 2 to 3 years often associated with obstruction, epistaxis or pain. Tumors have an average diameter of 3.1 cm, suggesting that they are fairly slow-growing in light of the number of months of symptoms. The majority of lesions are unilateral, although bilateral tumors can occasionally be observed [4, 5]. Nuclear magnetic resonance imaging with gadolinium injection shows a heterogeneous contrast pattern suggestive of neurofibromas [6, 7]. Preoperative biopsy is often difficult to interpret, apart from immunohistochemistry, demonstrating immunoreactivity to protein S 100. Surgical removal with anatomopathological examination of the entire surgical specimen is therefore necessary. Therapeutically, complete surgical excision is the treatment of choice for neurofibromas [8, 6], as recurrence is possible even if its frequency remains low [8]. Also, cases of malignant degeneration have been described in the literature [9, 10]. The possibility of recurrence and malignant degeneration implying in all cases prolonged surveillance of patients.

CONCLUSION

Neurofibromas are rare benign tumors of the peripheral nerve sheath of the nasosinus tract. It carries a risk of recurrence and degeneration, and requires prolonged clinical and radiological surveillance.

Conflicts of Interest: No.

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