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UNIVERSIDADE D
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Bruno Daniel Pereira Paiva

NASAL FOSSA SCHWANNOMA COEXISTING WITH VESTIBULAR SCHWANNOMA: A CASE REPORT

Caso Clínico

Área Científica de OTORRINOLARINGOLOGIA

Trabalho realizado sob a orientação de:

Maria do Carmo Eva Miguéis[1,2]

António Carlos Eva Miguéis[1,2]

[1] Faculdade de Medicina, Universidade de Coimbra, Portugal

[2] Serviço de Otorrinolaringologia | Centro Hospitalar e Universitário de Coimbra

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RESUMO

Introdução: O Schwannoma é um dos tumores mais comuns dos que crescem na bainha dos nervos periféricos. São sobretudo solitários e esporádicos, e quando múltiplos associam-se normalmente a etiologias genéticas. A localização mais comum é a porção vestibular do VIII par craniano, raramente envolvendo a fossa nasal.

Caso Clínico: Apresenta-se o caso de um doente com um Schwannoma da fossa nasal esquerda em simultâneo com um provável Schwannoma vestibular esquerdo. Inicialmente, o doente dirigiu-se ao serviço de urgência por vertigem de início agudo, tendo sido encontrado um pólipo nasal que, após ressecação eletiva, foi classificado como um Schwannoma. Recidiva sintomática levou a que fosse feita uma TC crânio-encefálica, que demonstrou um possível Schwannoma vestibular esquerdo. O doente aguarda estudo por RM. Não existem antecedentes familiares de relevo e o doente atualmente não preenche os critérios para neurofibromatose tipo 2.

Conclusão: Caso o diagnóstico seja confirmado, este parece ser o primeiro caso relatado de um doente com um Schwannoma da fossa nasal em simultâneo com um Schwannoma vestibular unilateral. Recomenda-se que, perante um doente com diagnóstico de Schwannoma, seja tida em conta a possibilidade de poderem existir outras localizações tumorais.

Palavras-Chave: Schwannoma vestibular, Schwannoma, Pólipo nasal, Vertigem

ABSTRACT

Introduction: Schwannomas are one of the most common types of peripheral nerve sheath tumours. They are most commonly solitary and sporadic, with multiple Schwannomas being frequently associated with genetic disorders. The most common location is the vestibular portion of the VIII cranial nerve, while involvement of the nasal cavity is rare.

Case Report: We present the case of a patient with a left nasal fossa Schwannoma coexisting with a suspected left vestibular Schwannoma. He first presented with acute onset vertigo and a nasal polyp was found during the workup, which was classified as a Schwannoma after surgical resection. Symptom relapse led to a CT scan of the head being performed, which showed a potential left vestibular Schwannoma. Patient is currently awaiting confirmation by MRI. He does not meet the criteria for neurofibromatosis type 2 nor does he have any relevant family history.

Conclusion: If the vestibular Schwannoma is confirmed, we believe this is the first reported case of a unilateral vestibular Schwannoma coexisting with a nasal fossa Schwannoma in a patient who does not have neurofibromatosis. We suggest that other tumour locations should be considered when approaching a patient with Schwannoma.

Keywords: Vestibular Schwannoma, Multiple Schwannoma, Nasal Polyps, Vertigo

INTRODUCTION

Schwannomas are one of the most common types of peripheral nerve sheath tumours. They are generally benign, deriving from the Schwann cells present in peripheral nerves. [1] They are most commonly solitary and not related to any specific syndrome, with multiple Schwannomas being frequently associated with genetic disorders such as Neurofibromatosis or Schwannomatosis. [2] Symptoms can result from direct invasion of the nerve, infiltration of surrounding tissues or local mass effect, and are heavily dependent on the affected nerve and its location. [3]

Schwannomas can originate from any peripheral nerve that contains Schwann cells, but they most commonly affect the vestibular portion of the VIII cranial nerve. [1,3] Although they may be discovered incidentally when performing a neuroimaging exam, patients can also present with unilateral hearing loss, dizziness and tinnitus. [1]

Schwannomas affecting the nasal cavity are rare and review of the literature showed only one previous case where a nasal Schwannoma coexisted with a vestibular Schwannoma. [4] We present the case of a patient with a left nasal fossa Schwannoma, which has been surgically removed in the meantime, coexisting with a suspected left vestibular Schwannoma.

CASE REPORT

A 51-year old male comes to the emergency room with acute onset vertigo. He has hypertension, but no other medical or surgical history. There is no relevant family history. His only medication is Irbesartan. He denies smoking or drinking alcohol frequently. Physical exam is unremarkable. To rule out a stroke as the cause of the symptoms, a CT scan of the head and neck is performed, which shows no evidence of ischemic or haemorrhagic events. However, a polyp is described in the left nasal fossa and the patient is referred to the Otorhinolaryngology outpatient consultation. He is prescribed Betahistine 16mg, to be taken twice a day.

The patient is seen about 5 months later, complaining of left nasal obstruction, hyposmia and rhinorrhoea. He denies having any episodes of epistaxis, headache, facial pain or oedema. As part of the routine head and neck examination of a patient complaining of nasal obstruction, [5] an anterior rhinoscopy is performed, showing a polypoid lesion in the left nasal fossa, exteriorized from the middle meatus. The patient is then scheduled for polypectomy through endoscopic surgery. Presurgical CT-scan is shown in Figure 1.

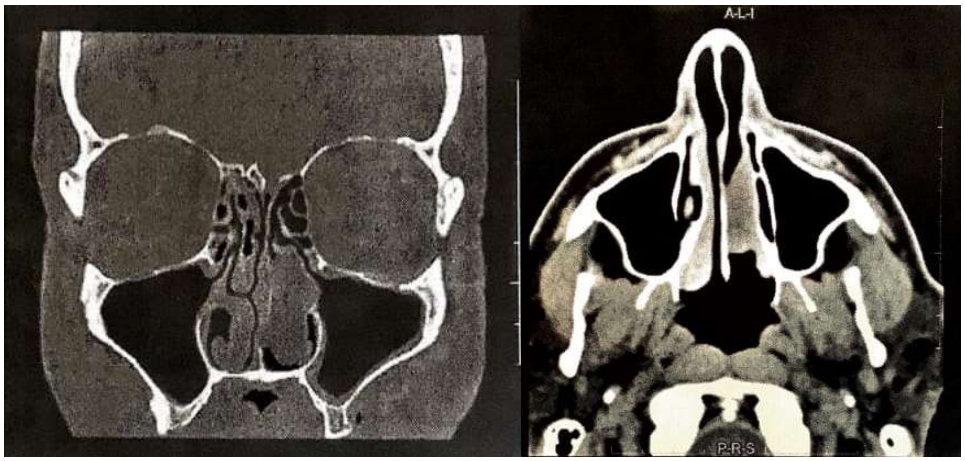

FIGURE 1: Paranasal sinuses CT-scan, showing a voluminous polyp in the left nasal fossa.

The surgery is performed 7 months later without complications. A 3,4x2,5x0,9cm polyp is removed and sent for histopathological analysis, which reveals a non-capsulated lesion, alternating between two different patterns: a cellular dense area, arranged in fascicles, showing palisading of nuclei and nuclei-free zones between them, consistent with Verocay bodies, and a hypocellular area, where loose tissue and oedema predominate (Figure 2a).

Cells are strongly immunoreactive to S100 (Figure 2b) but not to smooth muscle actin, which aids the final diagnosis of left nasal fossa Schwannoma.

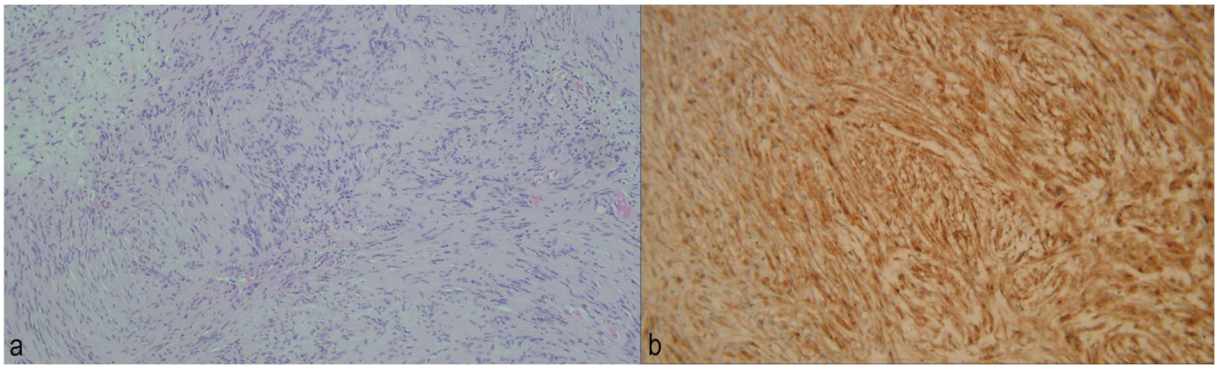

FIGURE 2: a. Histology slide of the polyp, showing two alternating patterns: a cellular area with Verocay bodies and a hypocellular area with loose tissue and oedema. **b.** S100 antibody staining showing strongly immunoreactive cells.

The patient is seen a few days later for postoperative follow-up. He maintains complaints of vertigo. Tone audiometry, tympanogram and videonystagmography are all normal, with dynamic posturography showing a heavy visual dependency.

Two weeks later, the patient returns to the emergency room with complaints of dizziness and difficulty walking. During the physical exam, the patient presents a broad-based gait and is unable to walk with one foot directly in front of the other. Another CT scan of the head and chest is requested, which reveals an expansive lesion in the left internal auditory meatus, extending to the cerebellopontine angle, with less than 1cm in size (Figure 3). The findings are consistent with a vestibular Schwannoma and further studies through magnetic resonance imaging are suggested, which the patient is currently awaiting, along with an evaluation by the Neurosurgery department.

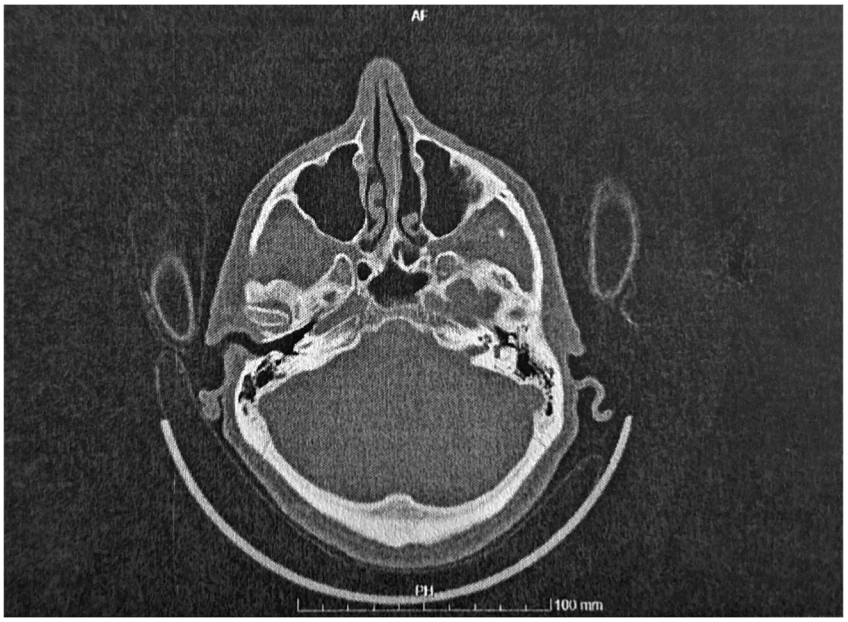

FIGURE 3: Head CT scan, without contrast, performed in an acute episode of dizziness and gait abnormalities, showing an expansive lesion in the left internal auditory meatus extending to the cerebellopontine angle, with less than 1cm in size. Performed after left nasal fossa polypectomy.

DISCUSSION

It seems likely that the first episode of vertigo was already caused by a growing vestibular Schwannoma, even though no evidence of it was reported in the initial CT scan. Currently, the preferred method of diagnosis when suspecting of a vestibular Schwannoma is gadolinium-enhanced magnetic resonance imaging (MRI). [6] When performed, it should show an enhancing lesion by the internal auditory canal extending into the cerebellopontine angle. [1] While the CT scan findings of this patient were suggestive of vestibular Schwannoma, it is still important that an MRI is performed to confirm the diagnosis and assess the best individualized approach. Therefore, the absence of an MRI poses a limitation to the discussion of this case.

Out of the genetic disorders known to cause multiple Schwannomas, only Neurofibromatosis type 2 (NF2) is associated with vestibular Schwannomas, often bilateral, which constitutes a hallmark of NF2. [7] About 50% of patients with NF2 do not have family history of the disease, having acquired a de novo mutation. [7] The reported patient does not currently meet the Manchester criteria for NF2. [8] However, he will if in the future he develops one of meningioma, cataract, glioma, neurofibroma, cerebral calcification or Schwannoma. [8] Therefore, adequate follow-up should ensue.

Management of vestibular Schwannoma should be individualized, focusing on controlling the tumour and minimizing functional deficit. [9] In the last few decades, less invasive approaches have become increasingly popular, either by watchful waiting or by performing stereotactic radiosurgery. [10] Studies suggest that watchful waiting shows the best overall outcomes in patients whose tumours will not grow significatively, while maintaining a high functional level, which includes the elderly and those with significant comorbidities. [10,11] In general, though, patients with small to medium-sized vestibular Schwannomas show the best results after an early diagnosis and going under Gamma Knife radiosurgery, achieving tumour control in over 97% of cases with a very low morbidity. [10,12] Patients are also very likely to experience symptom relief, specially those with vertigo or balance disorders. [12]

CONCLUSION

If the patient does not go on to meet the criteria for NF2, we believe we are reporting the first case of a unilateral vestibular Schwannoma coexisting with a nasal fossa Schwannoma in a patient who does not have neurofibromatosis. This case suggests that during the workup of a patient with Schwannoma, obtaining a detailed clinical history and integrating with the findings of the physical exam is of particular relevance, as other tumour locations should be considered even if no relevant family history or genetic predisposition appears to exist.

REFERENCES

1. Jameson JL, Fauci AS, Kasper DL, Hauser SL, Longo DL, Loscalzo J. Harrison's Principles of Internal Medicine. 20th Edition. McGraw Hill Higher Education. 2018.
2. Whitaker SB. Oral and maxillofacial pathology. 4th Edition. Vol. 131, Journal of the American Dental Association (1939). W.B. Saunders Co.; 2000. 718 p.
3. Schoen FJ, Mitchell RN. Robbins & Cotran Pathologic Basis of Disease. Elsevier Inc. 2015. 570 p.
4. Lee EJ, Kim MK, Kang MS, Kim KS. Nasal tip schwannoma coexisting with vestibular schwannoma. J Craniofac Surg. 2016;27(7):E626–7.
5. Johnston DR, Rosen MR. Anterior Rhinoscopy and Nasal Endoscopy in the Diagnosis of Sinonasal Disease. In: Diseases of the Sinuses. Springer New York; 2014. p. 277–93.
6. Dawes PJD, Mehta D, Arullendran P. Screening for vestibular schwannoma: magnetic resonance imaging findings and management. J Laryngol Otol. 2000;114(8):584–8.
7. Asthagiri AR, Parry DM, Butman JA, Kim HJ, Tsilou ET, Zhuang Z, et al. Neurofibromatosis type 2. Vol. 373, The Lancet. 2009. p. 1974–86.
8. Evans DGR, Baser ME, O'Reilly B, Rowe J, Gleeson M, Saeed S, et al. Management of the patient and family with neurofibromatosis 2: A consensus conference statement. In: British Journal of Neurosurgery. Br J Neurosurg; 2005. p. 5–12.
9. Kaul V, Cosetti MK. Management of Vestibular Schwannoma (Including NF2): Facial Nerve Considerations. Vol. 51, Otolaryngologic Clinics of North America. W.B. Saunders; 2018. p. 1193–212.
10. Kondziolka D, Mousavi SH, Kano H, Flickinger JC, Lunsford LD. The newly diagnosed vestibular schwannoma: Radiosurgery, resection, or observation? Neurosurg Focus. 2012 Sep;33(3).
11. Wolbers JG, Dallenga AHG, Romero AM, Van Linge A. What intervention is best practice for vestibular schwannomas? A systematic review of controlled studies. Vol. 3, BMJ Open. BMJ Publishing Group; 2013.
12. Boari N, Bailo M, Gagliardi F, Franzin A, Gemma M, del Vecchio A, et al. Gamma Knife radiosurgery for vestibular schwannoma: clinical results at long-term follow-up in a series of 379 patients. J Neurosurg. 2014 Dec 1;121:123–42.

ATTACHMENTS

Attachment 1: Informed Consent Form

Formulário de consentimento

Para o consentimento de um(a) doente para a publicação de imagens e/ou informação sobre este/esta.

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Relação com o(a) doente (se o(a) doente não assinar este formulário): _____

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Eu _____ *[NOME COMPLETO EM LETRA DE IMPRENSA]* dou o meu consentimento para que o Material sobre mim/o(a) doente apareça numa publicação.

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(5) O texto do artigo será editado para verificação de estilo, gramática e consistência antes da publicação.

(6) Eu/o(a) doente não receberei/á qualquer benefício financeiro com a publicação do artigo.

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Se assinar em nome do(a) doente, indique o motivo pelo qual o(a) doente não pode dar o consentimento (por exemplo, o(a) doente faleceu, tem menos de 18 anos ou tem défice cognitivo ou intelectual).

_____ *Data:* _____

☐ *Se estiver a assinar para uma família ou outro grupo, assinale a caixa para confirmar que todos os membros relevantes da família ou grupo foram informados.*

Dados da pessoa que explicou e administrou o formulário ao(à) doente ou seu representante (por exemplo, o autor correspondente ou outra pessoa que tenha autoridade para obter o consentimento).

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Data: _____