CASE STUDY

Nasopharyngeal Melanoma: A Rare Disorder

El melanoma nasofaríngeo: una entidad infrecuente

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Clinical Case

We present the case of a 66-year-old patient without relevant history, who was admitted due to loss of consciousness, tonic-clonic movements and somnolence. The initial physical examination revealed no cardio-respiratory alterations or neurological deficits. Complementary tests did not show haematological or electrocardiographic involvement and the chest X-ray was normal. The cranial computed tomography (CT) scan revealed areas of right vasogenic cerebral oedema and a 14-mm nodule in the left frontal lobe, both compatible with cerebral metastases.

We began searching for the primary tumour (pulmonary > renal > melanoma > breast > colorectal) with a CT of the thorax and abdomen. The CT revealed 2 small, 10-mm hypodense nodules in the hepatic segments VII and II, suggestive of metastasis. A cranial magnetic resonance (MR) confirmed the presence of bilateral cerebral metastases, the largest of some 2 cm in the right hemisphere, with similar images in the right occipital lobe (0.5 cm) and the left frontoparietal (1 cm) (Fig. 1A and B). Nasofibrolaryngoscopy detected an irregular, non-exophytic whitish tumour outgrowth of the left tube rim (Fig. 2A), with an anatomo-pathological result of nasopharyngeal mucosal melanoma (MM) (Fig. 2B–D). We presented the case in the head and neck oncology committee, which decided on chemoradiotherapy treatment at palliative dosages. The patient died 2 months after diagnosis.

Discussion

Mucosal melanoma is a rare entity (0.5%-2% of the aerodigestive tract melanomas) that has been poorly studied to date, which makes early diagnosis difficult. In our case, an uptake lesion of the left tube rim went undetected on the brain MR. This was probably due to the fact that there was no extension towards neighbouring regions, as occurs in other radiological patterns for MM (Fig. 1C), and that the most frequent options in differential diagnosis include haemorrhagic processes, fungal infections, mucoceles or fatty lesions. Assessment of chest extension is essential. Techniques such as PET-CAT scan are sometimes used to rule out the existence of further distant metastases. In contrast to the cutaneous types, MM is not related to sunlight exposure and it often behaves more aggressively. There is a peak of incidence between the 4th and 7th decades of life and its most frequent location in the head and neck is nasal. Treatment consists of surgery, with concomitant radiotherapy (RT) and chemotherapy (CT), and prognosis depends on its extension.

All head and neck MM cases should be followed up for 5 years, given the high rate of persistence/recurrence. For Díaz Molina et al. in the 17 patients diagnosed, the most frequent symptom of presentation was epistaxis (77%). The surgical technique of choice was paralateronasal rhinotomy (10 of the 12 operated on) over endoscopic surgery; 3
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Figure 1 Cranial NMR. 1A: axial slice. Bilateral cerebral metastases, the largest of some 2 cm in the right hemisphere. 1B: sagittal slice. Left frontoparietal metastasis. 1C: axial slice. Hyper-uptake in left tube rim.

refused treatment, 1 was treated with CT, 1 with CT + RT and 4 were given complementary immunotherapy (3 with Bacille Calmette–Guerin (BCG) vaccine and 1 with interferon alpha-2-beta. Survival at 5 years was 35.7%, with the main cause of therapeutic failure being local recurrences and metastases. McLean et al. in a series of 30 cases that differentiated rhinosinusal tumours (22) from oral (8), did not find any statistically significant differences in survival by location; they reported a mean 5-year survival rate of 17 months for the oral tumours and of 12 months for the rhinosinusal. Mendenhall emphasises in his study that the optimum treatment is surgery. Despite this, 50% of the tumours resected tend to present recurrences, a percentage that decreases is RT is associated. According to the Slavicek series, the most frequent locations are the lateral wall of the nasal cavity > septum > maxillary sinus > ethmoid block > orbital-ethmoid complex > nasopharynx > lacrimal sac > tonsils > hypopharynx. Of the 19 patients in that series,

Figure 2 Nasofibrolaryngoscopy and histopathological detail. 2A: nasofibroscopy image of mucosal melanoma over left tube rim. 2B: histopathological image (HE staining) of mucosal melanoma. 2C: histopathological image (HMB-45 staining) of mucosal melanoma. 2D: histopathological image (S100 staining) of mucosal melanoma.
were treated surgically, 7 with RT, 1 using CT and the remaining patient died before initiating any treatment. Of the 10 operated on, 4 received complementary RT and 3, CT. From all of these, 9 cases had recurrences, of which 5 were operated on and 4 treated with CT-RT. Mean survival was from 2 to 22 months, with the 3-year survival rate being 41%.

Although immunotherapy is currently effective only in a small group of patients, the new treatments for MM focus on this direction. Classically, the use of BCG directly on the tumour has been the most widely used immuno-suppression method. At a later date, interferon came to represent another alternative for treatment of Stage II/III MM. At present, drugs such as vemurafenib \(^{11}\) (BRAF\(^{V600E}\) inhibitor) or ipilimumab (a monoclonal anti-CTLA-4 antibody) are being unveiled as novel, promising therapies that serve as resources for new research paths.\(^{12}\)

### Conclusion

Head and neck MM is a particularly rare tumour, it is difficult to diagnose and it has a late clinical presentation in its metastatic form. Nasofibroscopy study is crucial for its diagnosis. Treatment for this type of tumours should be carried out through a committee of experts that approach the best therapeutic option from different perspectives, with the best option still being surgery.

### Conflict of Interests

The authors have no conflicts of interests to declare.

### References