CASE STUDY

Merkel Cell Carcinoma of the Ethmoid Sinus

Carcinoma de células de Merkel etmoidal

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Clinical Case

This is the case of a 36-year-old male with a history of nasal surgery 2 years previously for sinonasal polyps. He complained of left nasal obstruction and progressive ipsilateral otalgia of 2 months evolution. On anterior rhinoscopy he presented with left septal deviation in Cottle areas ii–iii, with abundant mucopurulent rhinorrhea and a lesion of polypoid appearance originating in the middle meatus (Fig. 1); there were no changes in the rest of the ENT and cervical examination.

Tomographic study revealed density of soft tissue of rounded appearance in the left ethmoid sinus, well-delimited, with no signs of bone lysis, and a hydroaerial level in the right maxillary and sphenoid sinuses.

Endoscopic surgery was performed to the paranasal sinuses with excisional biopsy of the growth originating in the left ethmoid sinus, with defined edges, smooth and pink surface of indurated consistency and friable on palpation. Sphenomaxillary sinusitis was discovered secondary to meatal obstruction.

The preliminary histopathological diagnosis with H&E stain was of an undifferentiated, small, blue cell neoplasm. The immunohistochemical diagnosis was of a poorly differentiated malignant neoplasm of epithelial origin with cytokeratin 20 expression (low molecular weight with paranuclear pattern), cytokeratin 7 negative and positive coexpression of neuroendocrine markers (specific neuronal enolase and chromogranin A) compatible with Merkel cell carcinoma (Fig. 2). Grimelius stain was performed to confirm the case. The PET-CT scan reported i, ii and iii cervical adenomegalies, with increased metabolic activity. The patient initially received 6 cycles of chemotherapy, each 3 day cycle consisted of 150 mg cisplatin on the first day and 150 mg etoposide on the following 2 days. He was given 35 sessions of 50 Gy radiotherapy fractionated into 25 initial sessions and a 15 Gy overdose fractionated into 10 subsequent sessions. Thirty minutes prior to each radiotherapy session 500 mg of amifostine IV was applied as radioprotector.

Figure 1  Polypoid appearance of Merkel cell carcinoma.

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Merkel cell carcinoma

Follow-up at one year consisted of nasal fibroendoscopy, magnetic resonance imaging and PET-CT scan, and the latter reported no data of activity.

Currently the patient is being monitored, is in generally good condition and has no sinonasal symptoms or signs of a recurrence of the cancer.

Discussion

Merkel cell carcinoma is a rare and aggressive neuroendocrine tumour, which accounts for less than 1% of all skin tumours; 90% of the patients are male, and only 5% of cases of merkeloma present in people under 50; its pathogenesis has not been clearly studied. Merkel cell polyomavirus has been recently associated with carcinoma, suggesting oncogenesis induced by the virus; it is also associated with immunodeficiencies, UV radiation and exposure to arsenic.

Clinically it presents as an erythematous or purple growth which is elastic and smooth in exposed areas of the skin. Lesions smaller than 2 cm tend to be asymptomatique. The patient presented obstructive nasal symptoms and otalgia, which corresponds to most sinonasal growths. 30% of the patients present with metastatic disease at the time of diagnosis.

Diagnosis is histopathological and with H&E stain shows small, rounded blue cells with hyperchromatic nuclei, and immunohistochemistry shows positivity for chromogranin A, specific neuronal enolase and cytokeratin 20. This tumour metastasises in up to 50% of the cases, the most frequent being the skin, lymph nodes, liver, bone, lungs and brain.

Survival at 5 years varies between 30% and 75% depending on the stage; there is 45% recurrence over an average of 9 months. There are still no universally accepted treatment protocols for this illness; the majority recommend wide local excision followed by radiotherapy of 4500–6000 cGy for 5 or 6 weeks. This tumour is chemosensitive and represents an option for patients with advanced disease and recurrence of disease.

Conclusion

This disease is rare and particularly rare in the paranasal sinuses. The clinical presentation of the case and its similarity to other common diseases of the nose and paranasal sinuses (such as sinonasal polyposis), require that it be considered and included as a further differential diagnosis in polypoidal and neoplastic rhinosinus disease. Immunohistochemical study is essential for accurate diagnosis.

Conflict of Interest

The authors have no conflict of interests to declare.

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References