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PRIMARY MELANOMA OF MECKEL'S CAVE

Case report

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ABSTRACT - We present a case of trigeminal neuralgia with cranial normal magnetic resonance image (MRI) and computed tomography. The pain was not relieved by carbamazepine and microvascular decompression surgery was done. After two months the pain was similar to the condition before surgery. At this time, MRI showed an expansive lesion in Meckel's cave that was treated with radical resection by extra-dural approach. The pathologic examination revealed a primary melanoma. The follow-up after six months did not show abnormalities.

KEY WORDS: primary melanoma, central nervous system, Meckel's cave, carbamazepine.

Melanoma primário do cavo de Meckel: relato de caso

RESUMO - Apresentamos um caso de neuralgia do trigêmeo com investigação radiológica de ressonância magnética (RM) e tomografia computadorizada apresentando resultado normal. A dor não apresentou alívio com carbamazepina, sendo indicado descompressão microvascular do trigêmio. Passados dois meses, o paciente queixava-se de dor com intensidade similar à do pré-operatório. Nova RM mostrou lesão expansiva no cavo de Meckel, a qual foi tratada cirurgicamente por abordagem extra-dural. O exame anatomopatológico foi compatível com melanoma primário. O seguimento radiológico, após seis meses da cirurgia, não apresentou anormalidades.

PALAVRAS-CHAVE: melanoma primário, sistema nervoso central, cavo de Meckel, carbamazepina.

The central nervous system (CNS) is host to a wide variety of melanocytic neoplasms including the common metastatic malignant melanoma and the relatively benign, as well as overtly malignant, primary neoplasms of leptomeningeal melanocyte[1]. Metastases of malignant melanomas in the CNS are fairly common; their incidence is approximately 5-10% of all tumors metastatizing to the CNS. In contrast, primary melanomas of the CNS are a rare occurrence[2,3]. Trigeminal neuralgia is often the first complaint for a tumor of the Gasserian ganglion. Tumors of the fifth nerve in general are extremely rare and comprise 0.2 % of all primary and secondary intracranial neoplasms[4]. Melanomas of the fifth nerve are even rarer[5,6].

We report a case of trigeminal neuralgia that was not relieved by carbamazepine and microvascular decompression. The expansive lesion was detected in Meckel´s cave only after a second magnetic resonance image (MRI) and the histopathological examination showed a primary melanoma.

CASE

A 55-year old man, smoker, complains of right trigeminal neuralgia that began 4 months ago. The neurological examination was normal and treatment was instituted with carbamazepine 600mg a day. The neurological investigation was performed by cranial MRI and computed tomography (CT). After 30 days the patient returned without relief of the facial pain and with a normal neuroradiological examination (Fig. 1A). Microvascular decompression was indicated with displacement of the vessel impinging on nerve. The pain relief was temporary. After 60 days the pain was similar to the conditions before surgery and carbamazepine dose was increased to 1000 mg a day CT showed only the area of surgical manipulation (Fig. 1B). Despite the increased carbamazepine dose, the pain was more intense and frequent. Another MRI showed an expansive lesion in the right Meckel's cave, hyperintense in T1 with homogenous contrast (Fig. 2A, B). Radical surgical excision was performed by extradural

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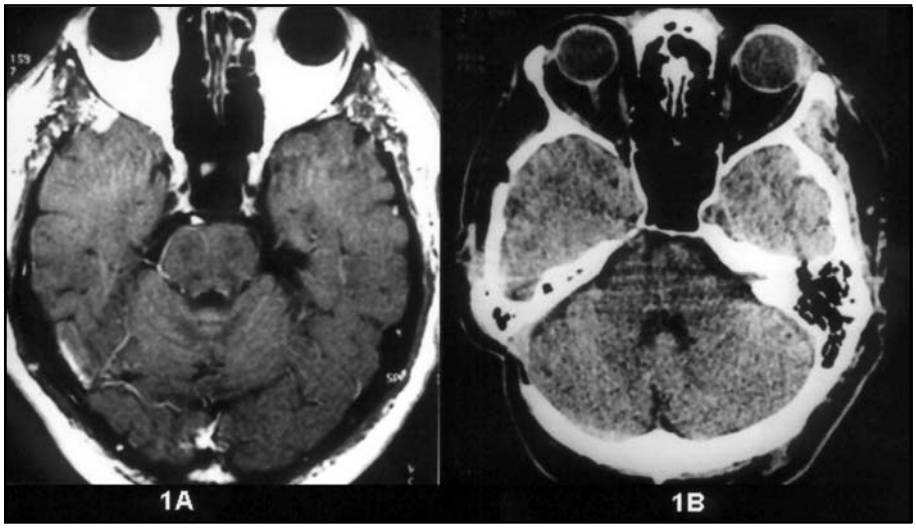

Fig 1. Normal neuroradiological investigation. Post contrast T1-weighted axial MRI scan (1A) and axial CT (1B). The CT was done after the microvascular decompression surgery.

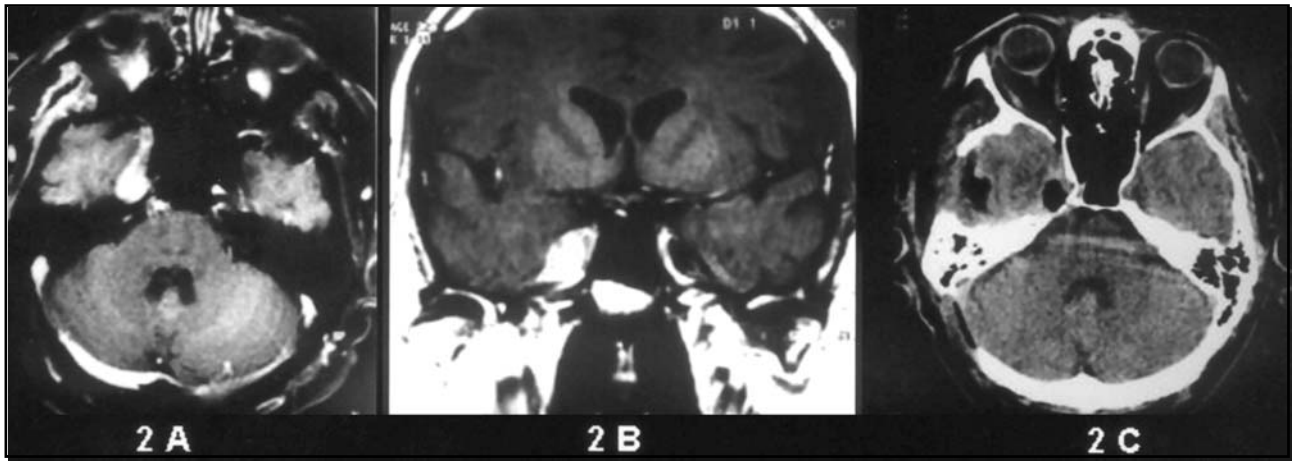

Fig 2. Post contrast T1-weighted axial (2A) and coronal (2B) MRI scans showing expansive lesion at the right Meckels cave and the radical excision after surgery demonstrated by axial CT (2C).

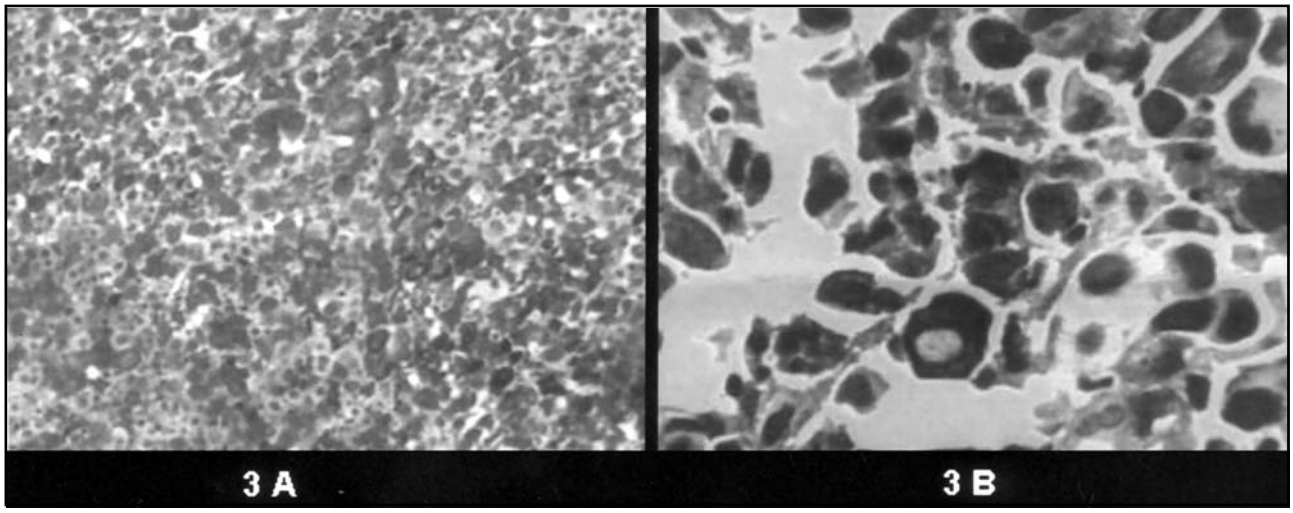

Fig 3. Malignant tumor with cellular pleomorphism and the presence of melanocytic cells. Photomicrographs illustrating histological features. H & E, original magnification, X 100 (3A) and X 400 (3B).

approach (Fig. 2C). After surgery there was right facial anesthesia and the trigeminal pain was relieved. The histopathological exam revealed a malignant tumor with cellular pleomorphism and the presence of melanocytic cells (Fig. 3). Skin was normal at inspection. Radiological examinations of the chest and abdomen failed to detect any other lesions. The 9-month radiological follow-up did not detect tumor recurrence.

DISCUSSION

Primary melanocytic tumors occur both as diffuse leptomeningeal proliferations and discrete expansive lesions. These tumors consist of a spectrum ranging from well-differentiated melanocytomas to melanoma[1]. Patients of all ages are affected. Diffuse lesions, which usually affect children, are occasionally associated with multiple or extensive pigmented lesions of the skin. This complex is known as neurocutaneous melanosis[1].

Melanoma can arise anywhere melanoblasts are found, and it has been suggested that dural melanoma originates from heterotrophic dural melanocytes. In the cranialspinal axis, melanocytes are most frequent in the leptomeninges that occur predominantly around the ventral and lateral aspects of the spinal cord, the ventral aspect of the brain, cerebral peduncles, and sylvian fissure[7,-10]. While some degree of leptomeningeal melanosis occurs in up to 85% of the population, heterotrophic melanin detected in the dura is rare[11]. Reports of primary dural melanoma are rare[12-18] and tumors of the fifth nerve in general are extremely rare and comprise 0.2 % of all primary and secondary intra-cranial neoplasms[4]. Melanomas of the fifth nerve are even rarer[5,6]. In von Recklinghausen's disease, primary malignant melanomas have been described on the skin as well as in the uvea[19- 21], iris[22] and meninges[23-25]; none have been reported to involve the fifth nerve as far as we know.

An interesting observation was that carbamazepine associated with microvascular decompression did not relieve the trigeminal pain. This should call attention to the fact that it is necessary to persist with the radiological investigation.

Meningeomas and schwannomas of the region of Meckel's cave are not uncommon. Their CT appearance may be similar to ours except that, on plain CT scan, melanomas usually appear hyperdense[26].

Primary malignant melanomas of the CNS vary in cytologic features much like those at other sites. Mitosis ranges from scant to numerous. Necrosis may be extensive, producing perivascular pseudopapillae. Transition to a diffuse growth pattern, satellite nodule formation in surrounding meninges and irregular invasion of spinal cord substance is common in melanoma[1]. In immunohistochemistry, melanocytic tumors are vimentin-reactive but lack epithelial markers. Staining for S-100 protein varies, but is often strong. Though not melanoma-specific, the HMB-45 stain is usually positive[1].

It appears that primary dural melanoma follows a less aggressive course, patients surviving longer than those with leptomeningeal melanoma[18,27]. A review on leptomeningeal melanoma showed a median postoperative survival of 1 month and an overall 2-year survival of only 15%[28]. Patients treated surgically for primary dural melanoma fare somewhat better, with disease-free intervals ranging from 9 to 18 months[12,13,18]. Adjuvant radiotherapy was not prescribed because of a paucity of evidence to suggest any benefit[13,29,30].

Our case is a rare example of a primary melanocytic tumor of the fifth cranial nerve. We considered the melanoma in our case as primary because, there was no evidence of any other melanoma on the skin or in the eye, there were no associated giant hairy nevi and imaging screening failed to reveal any extra-cranial malignancy.

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