Meckel's cave primary lymphoma

Diagnosis

The diagnosis of lymphoma should be considered for lesions affecting Meckel's cave in high-risk immunocompromised patients. The presence of an apparent dural tail in an otherwise typical schwannoma is the distinguishing characteristic of a lymphoma. The absence of hyperostosis helps differentiate it from a meningioma [1].

Treatment

The preferred surgical strategy is biopsy for diagnosis and then radiotherapy and chemotherapy rather than major cranial base surgery for total resection. [2].

Management of lesions involving Meckel's cave can represent a challenge for neurosurgeons, because of the deep-seated location and the surrounding complex neurovascular structures. Very small lesions arising from MC are generally asymptomatic and radiological follow-up with head MRI and PET-CT is sufficient to control these lesions. In rare cases, the rapid increase in the size of lesions and the alteration of the neurologic status make early histological characterization mandatory in the plethora of lesions arising from Meckel's cave; a very small percentage is represented by central nervous system lymphomas. Primary diffuse large B cell lymphoma is the most commonly found. Aggressive surgery, in case of suspicious Meckel's cave lesions, is strongly discouraged because this procedure may increase the risk of postoperative deficit and provides no survival benefit compared with biopsy alone [3].

Outcome

Extra-axial primary CNS lymphoma, considered rare, mainly arises in the white matter of the brain. Though the tumor responds well to radiation and chemotherapy, the prognosis of primary CNS lymphoma remains poor.

Its prognosis appears to be identical to that of other intracranial lymphomas [4]

Case reports

The aim of a paper of Paglia et a. was to report a very rare case of primary Meckel's cave diffuse large B-cell lymphoma (only seven cases were described in the literature) and standardize an operative algorithm to avoid the risks of an incorrect surgical conduct [5].

A 65-year-old man was examined at another hospital for unilateral facial pain. Carbamazepine was

prescribed, but his symptoms did not improve. Magnetic resonance imaging (MRI) revealed swelling of the trigeminal nerve and a mass lesion in Meckel's cave. The patient was referred to our hospital at this point. Gadolinium-enhanced MRI and F18-Fluorodeoxyglucose-position emission tomography suggested a likely malignant tumour and a biopsy was performed. Histopathological examination showed diffuse a large B cell lymphoma. The patient was treated with high-dose methotrexate (HD-MTX) and radiotherapy. Despite responding well to initial treatment, the patient relapsed, with lymphoma observed throughout the body. He died of pneumonia 18 months after the initial diagnosis. Facial pain is a symptom that is commonly managed in general practice. If symptoms do not improve, repeated imaging studies, including contrast MRI, is warranted. This is the first reported case of primary neurolymphomatosis (NL) of the trigeminal nerve associated with facial pain alone. Furthermore, HD-MTX and radiotherapy may be considered for the management of primary NL of a cranial nerve [6)].

Ang et al. described an atypical man with diffuse large B cell lymphoma localized to the sphenoid wing and adjacent cavernous sinus, initially presenting with isolated ipsilateral facial pain mimicking trigeminal neuralgia due to invasion of Meckel's cave but subsequently progressing to intra-axial extension and having synchronous features of systemic lymphoma. Primary central nervous system lymphoma is uncommon, accounting for approximately 2% of all primary intracranial tumors, but its incidence has been steadily increasing in some groups. It usually arises in the periventricular cerebral white matter, and reports of lymphoma in extra-axial regions are rare. This man highlights the importance of maintaining lymphoma in the differential diagnosis of tumors of the skull base presenting with trigeminal neuralgia-like symptoms [7)].

A 52-year-old man with a history of malignant lymphoma of the cecum presented with lancinating facial pain in the left. Magnetic resonance imaging (MRI) revealed a tumor in the Meckel's cave extending along the trigeminal nerve. The tumor was partially removed via left retrosigmoid lateral suboccipital craniotomy. Histological examination showed findings consistent with diffuse large B cell lymphoma, which was later confirmed to be metastatic lesion from the cecal lesion. Postoperative chemotherapy with cyclophosphamide, high dose, cytarabine, steroid (dexamethasone), etoposide, and Rituximab (CHASER) followed by whole brain irradiation (30 Gy) resulted in complete remission. Although facial pain persisted, the patient's general condition remained favorable and he did not experience recurrence over the 51-month follow-up period. Histological confirmation and awareness of malignant lymphoma are very important to determine the therapeutic strategy and to avoid misdiagnosis or delayed diagnosis. Long-term survival of patients with metastatic malignant lymphoma in the Meckel's cave extending along the trigeminal nerve was very rare. In addition, metastatic malignant lymphoma in the extra-axial and peripheral nervous tissue might be different from primary central nervous system lymphoma in the white matter, since the efficacy of chemotherapeutic agents against malignant lymphomas in the extra-axial regions is not attenuated by the blood brain barrier. [8)].

Kinoshita et al. reported a case of primary lymphoma of Meckel's cave mimicking a trigeminal schwannoma radiographically, which achieved complete remission through use of rapid high-dose MTX therapy and radiation therapy.

The patient, a 55-year-old Japanese male, presented left trigeminal neuralgia. Magnetic resonance

imaging (MRI) revealed a mass lesion in the left side of Meckel's cave, with extension into the cerebellopontine angle and the infratemporal fossa through the foramen ovale, suggesting trigeminal schwannoma. However, the patient suffered radiologically inexplicable progressive cranial nerve palsy, which suggested malignant disease. MRI and CSF disclosed malignant tumor dissemination; biopsy revealed malignant lymphoma. The treatment, composed of the rapid infusion of high-dose MTX and whole brain and spine radiation, resulted in complete remission.

This case, which included atypical presentation of malignant lymphoma, illustrates the importance of including malignant lymphoma in the differential diagnosis of CP-angle and Meckel's cave tumor. The results also confirmed the usefulness of combined rapid high-dose MTX therapy and radiation. [9].

Wakamoto et al. reported a rare primary intracranial malignant lymphoma that spread along the trigeminal nerve through the skull base foramen. The patient was a 50-year-old woman, who was diagnosed as having a primary intracranial malignant lymphoma in the right temporal lobe and had undergone an operation and radiation 5 years previously. The tumor was reduced in size and no recurrent tumor could be detected for 5 years. The patient complained of left face swelling and CT scan revealed a large mass in the pterygopalatine fossa. MRI revealed the recurrent tumor in the left Meckel's cave with extension into the cavernous sinus. The tumor extended through the foramen ovale into the pterygopalatine fossa, through the superior orbital fissure into the orbital cavity and through the infraorbital fossa into the face subcutaneously. Biopsy of the subcutaneous tumor was carried out and the pathological diagnosis was malignant lymphoma, B cell type, which was identical with the initial tumor. MRI revealed the enlarged trigeminal nerve and 3D-CT revealed the enlargement of the infraorbital fossa and the foramen ovale. We suspected that primary intracranial malignant lymphoma had recurred in the left Meckel's cave and the tumor had spread along with the peripheral three divisions of the trigeminal nerve. Perineural spreading along the trigeminal nerve passing through the skull base in patients with nasopharyngeal carcinoma is not rare, but this rarely occurs in the case of intracranial tumors [10].

Abdel Aziz et al. reported a case of primary lymphoma of Meckel's cave. The ability of lymphoma to mimic a trigeminal schwannoma, both clinically and radiographically, resulted in misdiagnosis and flawed surgical strategy. They discussed the characteristics of a Meckel's cave lymphoma on magnetic resonance images, the predisposing medical conditions that should cause the neurosurgeon to add lymphoma to the normal differential diagnosis, and appropriate management strategies.

A 40-year-old African-American woman presented with a 5-month history of progressive facial numbness and pain in all three divisions of the left trigeminal nerve. Magnetic resonance imaging revealed a mass in the left side of Meckel's cave, with extension into the lateral compartment of the cavernous sinus, without encasement of the internal carotid artery, through the foramen rotundum into the posterior aspect of the maxillary sinus, and through the foramen ovale into the pterygopalatine fossa. The diagnosis, based on clinical history and radiographic imaging, was schwannoma of Meckel's cave. The patient had a history of systemic lupus erythematosus that had been treated with intermittent steroid therapy.

The surgical approach selected was a frontotemporal craniotomy with orbitozygomatic osteotomy and anterior petrosectomy. The lesion was totally excised, although the gross intraoperative appearance of the lesion was inconsistent with the preoperative diagnosis, and the pathological examination was unable to establish a histological diagnosis on the basis of frozen sections. Histological diagnosis was

confirmed on permanent section after surgery as B-cell lymphoma. Evaluation for other primary sites produced negative results. The patient was then treated with cyclophosphamide (Cytotoxan; Bristol-Myers Oncology, Princeton, NJ), doxorubicin (Adriamycin; Pharmacia & Upjohn, Kalamazoo, MI), vincristine, and prednisone chemotherapy every 3 weeks for six cycles and then by radiation therapy to the affected area.

The diagnosis of lymphoma should be considered for lesions affecting Meckel's cave in high-risk immunocompromised patients. The presence of an apparent dural tail in an otherwise typical schwannoma is the distinguishing characteristic of a lymphoma. The absence of hyperostosis helps differentiate it from a meningioma. At this point, the preferred surgical strategy is biopsy for diagnosis and then radiotherapy and chemotherapy rather than major cranial base surgery for total resection. [11)].

Artico et al. presented a rare case of Meckel's cavity lymphoma. Only two other cases of identical localization were presented in the literature. The symptoms consisted of sensorimotor impairment of the Vth nerve associated with slight exophthalmos. C.T. scan showed a hyperdense lesion in Meckel's cavity. After total surgical removal, histological analysis diagnosed a B-lymphocyte non-Hodgkin's lymphoma. The patient received both radiotherapy and chemotherapy and at one year follow up, the clinical course was good. The lesion had no clinical or radiological specificity. Its prognosis appears to be identical to that of other intracranial lymphomas [12)]

References

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