Amelanotic melanoma

Primary amelanotic melanoma is a special subtype of Primary melanocytic neoplasm, which is especially rare.

Very few cases of amelanotic variation of primary melanoma in the CNS were reported on. General guidelines or recommendations to establish this diagnosis do not exist.

Sun et al. established intracranial and subcutaneous melanoma models using cultured malignant cells derived from amelanotic melanoma. The median survival times in a mouse model with intracranial tumors was 20 days, but a mouse model with subcutaneous tumors did not show cachexia until they were killed 28 days after inoculation with tumor cells. Histopathological analysis showed that a high karyokinesis phase and nuclear pleomorphism appeared in the intracranial model compared with the subcutaneous tumor model mice. The tumor boron concentration at 2.5 h after boronophenylalanine administration was 15.21±3.88 μg/g in an intracranial melanoma xenograft and 19.85±3.63 μg/g in a subcutaneous melanoma xenograft. Intracranial melanoma showed more malignancy and shorter survival time than did subcutaneous melanoma when the same number of tumor cells were injected, and subcutaneous and intracranial amelanotic malignant melanoma tumors are both fitted for boron neutron capture therapy [1].

Case reports

2017

Primary Amelanotic CNS Melanoma: Case Report and Literature Review [2].

2015

Ma et al. report a case of intracranial amelanotic melanoma. Preoperative assessment revealed progressive right frontal mass. The patient underwent tumor resection. The pathologic analysis reported amelanotic melanoma of intermediate grade. The further examination of the whole brain and body was negative. The familial history was also negative. The patient recovered uneventfully and went on for radiotherapy and chemotherapy. After a follow-up period of 5 months, the patient was tumor-free.

This is the second report about primary CNS amelanotic melanoma. They summarized characteristics of the primary CNS melanocytic lesions and amelanotic melanoma with review of the literature and review of cases from the department [3].

2009

A 69-year-old man presented with trigeminal neuralgia. 4 years previously he underwent tumor removal with an initial diagnosis of amelanotic malignant cutaneous melanoma; 1 year later, because of tumor recurrence, the patient underwent neck dissection, chemotherapy and radiation. Magnet resonance imaging (MRI) disclosed an enhancement of the Gasserian ganglion and tumor extension

along the mandibular and maxillar nerves of the intracranial part of the trigeminal nerve suggestive of tumor. The intraoperative macroscopic appearance of the tumor was compatible with a neurinoma. Histopathological studies proved the tumor to be a desmoplastic neurotropic melanoma (DNM) that was related to the previously treated malignant melanoma [4].

2008

Only 15 cases of intracranial amelanotic melanoma have been reported until 2008. A yellowish mass was observed in the frontal lobe. The content of the cyst consisted of old hematoma, xanthochromic fluid and necrotic tissue, was evacuated and the cyst wall was totally resected. No abnormal pigmentation was noted in the cyst wall and surrounding brain tissue. The imaging features of metastatic melanomas are distinctive due to the presence of melanin and the propensity for hemorrhage. Both hemorrhage and melanin can produce T1-weighted hyperintensity and T2-weighted signal intensity loss [5].

2006

A 63-year-old woman with diplopia and bilateral ptosis underwent brain MRI that showed a pituitary mass with signal characteristics suggestive of adenoma. Within one week she had developed nearly complete bilateral ophthalmoplegia. A repeat MRI showed extension of the mass into both cavernous sinuses. Hypophysectomy disclosed an amelanotic melanoma. Extensive search for a primary source was unsuccessful. Despite local radiation treatment, the tumor continued to grow and the patient became blind and died within several months of diagnosis. There are seven reported cases of melanoma arising primarily in the sella turcica. Two cases of metastatic melanoma to the cavernous sinuses have been reported. Amelanotic melanoma has not been reported as a cause of cavernous sinus syndrome [6].

1992

A thirty-four-year-old man was admitted to our hospital because of the disturbed visual acuity and pain on the eye movement of the right eye. He had prominent right eye and CT-scan and MRI of the brain disclosed a tumor which could be obviously distinguished from the extraocular muscles, optic nerve and the bulb of eye in the retrobulbar region. On operation we identified dark-red solid tumor which was 3.0cm in diameter, and diagnosed it malignant melanoma pathologically. Because postoperative study detected amelanotic melanoma in the white patch on the right upper extremity, this right orbital tumor was considered to be the metastases of it from the right upper extremity. Metastatic malignant melanoma of the skin to the orbit is very rare, while most of the eye-associated malignant melanoma originates from uveal tract, special choroid, and conjunctiva. This case was the 26th case of these in the world and the first case in Japan, furthermore the 4th case in the world whose first symptoms were caused by the orbital metastases [7].

1991

Primary intracranial amelanotic melanoma was verified at autopsy in a 38-year-old male. Correct diagnosis of amelanotic melanoma needs electron microscopy or immunohistochemistry, since

Masson staining is negative due to the absence of melanin pigment. We adopted the following criteria for clinical use: macroscopically not dark and microscopically negative for Masson staining, but ultrastructurally various melanoma types present. Although the clinical profile of this case is consistent with melanotic melanoma, the more detailed features of primary intracranial amelanotic melanoma require future study [8].

1)
Sun T, Li Y, Wu T, Xie X, Chen G, Wei Y, Li B, Zhou Y, Du Z. Comparative analysis of pathology and boronophenylalanine uptake in experimental orthotopic and heterotopic amelanotic melanoma. Melanoma Res. 2014 Aug;24(4):315-21. doi: 10.1097/CMR.0000000000000086. PubMed PMID: 24915302.
2)
Mayer S, Mauer UM, Mathieu R, Hackenbroch C, Knupfer J, Schulz C. Primary Amelanotic CNS Melanoma: Case Report and Literature Review. J Neurol Surg A Cent Eur Neurosurg. 2017 Jul 27. doi: 10.1055/s-0037-1604326. [Epub ahead of print] PubMed PMID: 28750450.
3)
Ma J, Zhang Z, Li S, Chen X, Wang S. Intracranial amelanotic melanoma: a case report with literature review. World J Surg Oncol. 2015 May 12;13:182. doi: 10.1186/s12957-015-0600-z. Review. PubMed PMID: 25963017; PubMed Central PMCID: PMC4436165.
4)
Hashemi M, Stark A, Hugo H, Mehdorn M. Intracranial trigeminal nerve metastases of a desmoplastic neurotropic melanoma: case report. Cent Eur Neurosurg. 2009 May;70(2):91-4. doi: 10.1055/s-0028-1082065. Epub 2009 May 25. PubMed PMID: 19711263.
5)
Cemil B, Emmez H, Oztanir N, Tokgoz N, Dogulu F. A cystic amelanotic melanoma metastases to the brain: case report. Neurocirugia (Astur). 2008 Aug;19(4):365-7. PubMed PMID: 18726049.
6)
Jacob S, Pye E, Hbahbih M, Messios N, Rajabally YA. Rapidly progressive bilateral ophthalmoplegia and enlarging sellar mass caused by amelanotic melanoma. J Neuroophthalmol. 2006 Mar;26(1):49-50. PubMed PMID: 16518168.
7)
Toyoda H, Fukui K, Okabe H, Kitoh A, Iguchi I, Kanematsu I. [A case of malignant melanoma with orbital metastases which caused the first symptoms]. No To Shinkei. 1992 Oct;44(10):929-33. Review. Japanese. PubMed PMID: 1285995.
8)
Seki Y, Ohara K, Aiba T, Unakami M, Hara M. Primary intracranial amelanotic melanoma–report of an autopsy case. Neurol Med Chir (Tokyo). 1991 Dec;31(12):773-6. PubMed PMID: 1726225.