

. 个案报道 .

高龄第四脑室原发性黑色素瘤 1 例

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【摘要】颅内原发性黑色素瘤是一种罕见的神经外科疾病,可发生在颅内任何位置,但第四脑室内极其罕见。病人的预后因肿瘤分化程度、位置及切除范围不同而存在差异,因此,术前准确的诊断、术中尽可能完整切除肿瘤并辅以术后放疗是改善病人预后的关键。本文报道 1 例 81 岁病人,术前头颅 CT 及增强 MRI 检查均显示第四脑室肿瘤,完善术前准备后,在显微镜下完全切除肿瘤,术中发现枕部皮肤、枕骨内板、脑膜及皮层均明显黑染,术后病理证实原发性黑色素瘤。虽然病人高龄,但术后临床症状消失,日常生活能自理,术后 10 个月影像学检查未见肿瘤复发,也未发现肿瘤远处转移。这提示高龄原发性脑室黑色素瘤,严格把握手术适应证,充分术前准备,可以考虑手术治疗。

【关键词】原发性脑室肿瘤;黑色素瘤;第四脑室;显微手术;高龄

【文章编号】1009-153X(2024)10-0635-03

【文献标志码】B

【中国图书资料分类号】R 739.41; R 651.1¹

A case of primary melanoma in the fourth ventricle in an elderly patient

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【Abstract】Intracranial primary melanoma is a rare tumor, yet it is extremely rare in the fourth ventricle. The prognosis of patients varies due to differences in tumor differentiation degree, location, and extent of resection. Hence, accurate preoperative diagnosis, complete tumor resection during the operation as much as possible, and supplemented with postoperative radiotherapy are the keys to improving the prognosis of patients. This paper reports a case of an 81-year-old patient. Both preoperative head CT and enhanced MRI examinations indicated a tumor in the fourth ventricle. After completing preoperative preparations, the tumor was completely resected under the microscope. During the operation, it was discovered that the occipital skin, the inner plate of the occipital bone, the meninges, and the cortex were all conspicuously blackened. Postoperative pathology confirmed primary melanoma. Although the patient was of advanced age, the clinical symptoms disappeared after the operation, and he was able to take care of himself in daily life. No tumor recurrence was found in imaging examinations 10 months after the operation, nor was distant metastasis detected. This indicates that for elderly patients with primary ventricular melanoma, by strictly grasping the surgical indications and conducting adequate preoperative preparations, surgical treatment can be considered.

【Key words】Primary ventricular tumors; Melanoma; The fourth ventricle; Microsurgery; Elderly patients

颅内黑色素瘤是临床上一种少见的神经外科疾病,大多数由颅外黑色素瘤转移引起,原发性颅内黑色素瘤罕见^[1]。本文报道 1 例 81 岁第四脑室原发性黑色素瘤,在显微镜下完全切除肿瘤,术后病理证实黑色素瘤。

1 病例资料

81 岁男性,因头痛 4 个月伴行走不稳 2 周于 2021 年 7 月 2 日入院。入院前 4 个月,无明显诱因出现头痛,部位不固定,呈持续性钝痛,时常伴有恶心,休息后未见好转;无肢体无力,无大小便失禁,无呕吐,当时未重视,未正规检查及治疗。两周前,出现行走不稳,当地医院头颅 CT 检查示第四脑室类圆形稍高密度影,室管膜瘤、脑膜瘤不排除。为求进一步治疗,来我院就诊。入院体格检查:枕外隆突处皮肤呈黑色,大小约 2 cm×4 cm(图 1F),未见其他阳性体征。行头颅 MRI 检查显示:第四脑室团块状异常信号,病灶 T₁ 像表现为高信号, T₂ 像表现为低信号;肿瘤边界清晰、形态不规则,增

强后明显均匀强化(图 1A~C),较大层面大小约 3.3 cm×3.1 cm×2.9 cm,邻近延髓、小脑受压、移位,周围血管未见明显受侵;SWI 序列示肿瘤内未见明显出血;各脑室、脑池稍扩大。2021 年 7 月 11 日全麻下经后正中入路手术切除第四脑室肿瘤。取枕部正中切口,上至枕外隆突上 2 cm,下至第二颈椎棘突。全层切开头皮,分离颈下肌群至颅骨,乳突撑开器撑开切口,暴露枕骨鳞部、寰椎后弓。磨钻钻孔一枚,铣刀形成大小约 4 cm×4 cm 骨窗,见颅骨内板呈黑色(图 1D),硬膜张力高,硬脑膜及蛛网膜呈黑色(图 1E)。打开枕骨大孔 2 cm 后,植入手术显微镜,“Y”形打开硬膜,见蛛网膜及蛛网膜下腔亦成黑色(图 1E);打开枕大池,进一步充分释放脑脊液,探查见肿瘤位于小脑蚓部前第四脑室内,呈黑色,质地较软,与周围脑皮质边界清楚,肿瘤内有囊变,可见少量褐色囊性组织,脑干及小脑未见明显侵犯。延肿瘤边界分离肿瘤,分块切除,显微镜下全切除肿瘤,后组颅神经、脑干及椎基底动脉保护完好。术区彻底止血,剪除部分黑染硬膜,人工硬脑膜严密修补硬膜,弃除骨瓣,逐层关颅。术后病理结果示:(枕部头皮)梭形细胞痣,黑染硬脑膜、黑染颅骨均未见肿瘤组织;第四脑室肿瘤细胞呈梭形,核仁深染,细胞内可见大量黑色素

doi:10.13798/j.issn.1009-153X.2024.10.015

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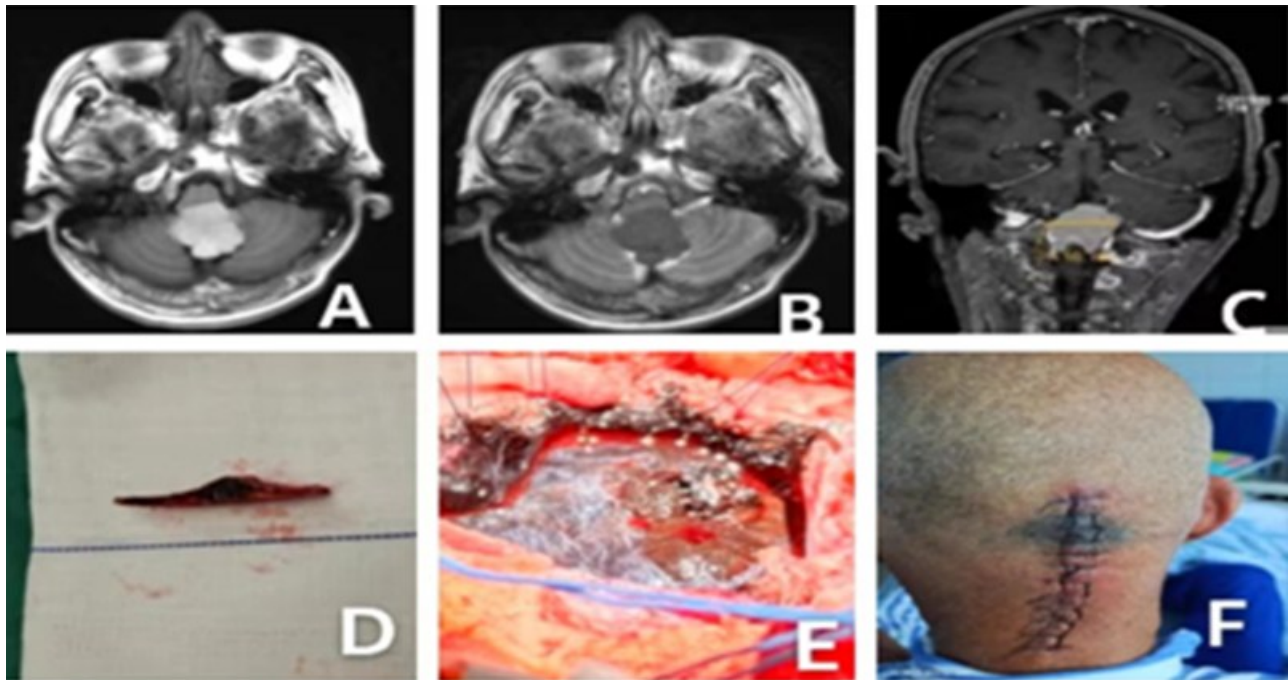


图1 第四脑室原发性黑色素瘤术前影像、术中所见

A~C. 术前MRI显示病灶T1像表现为高信号,T2像表现为低信号,增强后明显均匀强化;D. 术中切除的黑染的颅骨;E. 术中显微镜下观察发现硬脑膜、蛛网膜及表面皮层均被黑染;F. 术中观察枕部皮肤黑染,大小约2 cm×4 cm

Figure 1 Preoperative images and intraoperative findings of a patient with primary melanoma in the fourth ventricle

A~C: Preoperative MRI reveals that the lesion exhibits a high signal on T₁-weighted images and a low signal on T₂-weighted images, presenting obvious homogeneous enhancement after contrast administration. D: The inner plate of the skull resected intraoperatively shows distinct black staining. E: Intraoperative microscopic observation indicates that the dura mater, arachnoid, and the surface cortex are all significantly black-stained. F: During the operation, a black-stained area approximately 2 cm×4 cm is observed on the occipital skin.

沉着;免疫组化染色:瘤细胞CK(pan)(-)、S-100(+)、HMB-45(+)、Melan-A(A103)(+)、Vimentin(+)、SOX10(+)、GFAP(+)、Ki-67(+,约80%),考虑恶性黑色素瘤。

2 讨论

在早期胚胎发育过程中,黑色素细胞主要位于神经嵴,随后迁移至皮肤、眼和内耳,部分至软脑膜;因此,黑色素瘤好发于皮肤。颅内原发性黑色素瘤少见,占全身黑色素瘤的1%,占颅内肿瘤的比例<1/1000^[2]。颅内原发性黑色素瘤的年龄、性别及预后,文献报道各不相同。Suranagi等^[3]认为中老年人多见,男性居多。汪卫建等^[4]认为中年人且女性多见。有学者认为颅内原发性黑色素瘤恶性度较低,预后较好^[5];也有学者认为其恶性度极高并且预后极差^[6]。据文献报道,合并皮肤太田痣者颅内原发性黑色素瘤的可能性增加^[6,7]。这与本例病人的临床表现高度吻合。尽管如此,我们在术前仍未能考虑到此种罕见疾病,直至术中打开骨瓣,看见黑染的颅骨内板及黑染的硬脑膜,才意识到黑色素瘤的诊断可能。

颅内黑色素瘤CT的典型表现为高或等密度肿块并明显强化,密度高低与黑色素含量正相关。MRI信号特点取决于

肿瘤内黑色素细胞的含量及是否发生瘤内出血,当肿瘤内黑色素含量超过10%时,可出现典型的T₁WI高信号,T₂WI低信号。

该病可通过组织病理学检查、免疫组化、电镜检查来明确诊断。恶性黑色素瘤抗体(human melanoma black-45, HMB-45)和S-100蛋白均阳性表达,但神经上皮细胞标记物(如CK)表达缺失,可以排除上皮源性肿瘤。这一点有助于低黑色素型肿瘤的诊断。本文病例HMB-45、S-100均阳性,CK(-),符合恶性黑色素瘤的诊断标准。

原发性中枢神经系统黑色素瘤的最佳治疗方法是完全切除肿瘤。由于肿瘤可沿蛛网膜下腔及脑沟裂蔓延,增加肿瘤完全切除的难度,即使完全切除,也有局部复发可能^[8]。术后辅助放疗能否降低复发率或肿瘤延长复发时间,缺乏相关报道,但肿瘤复发可行二次手术切除。

原发性中枢神经系统恶性黑色素瘤的预后优于转移性黑色素瘤,术后生存期可达9~12年^[9]。尽管本文病例肿瘤恶性程度较高,Ki-67高达80%,且术后未行放疗,但术后头痛消失,日常生活状态良好,术后10个月影像学复查未见肿瘤复发且未发现远处转移。分析原因,可能与术中完整的肿瘤切除而解除了占位引起的局部压迫症状,使脑积水得到缓

解,从而改善了病人生活质量。

【利益冲突声明】:本文不存在任何利益冲突。

【作者贡献声明】:张义彪、常奎、徐敬斌实施手术;王鑫、肖珂、高亚峰查阅资料;张义彪撰写论文。

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(2022-05-18 收稿,2024-04-29 修回)



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综上所述,椎动脉远端发育异常伴基底动脉开窗畸形合并相关动脉瘤临床罕见,诊断上推荐颅脑 DSA,治疗上应注意保护分支血管、尽量保护开窗环;椎动脉发育不良使用支架辅助栓塞治疗动脉瘤可能增加缺血事件发生,应注意预防。

【利益冲突声明】:本文不存在任何利益冲突。

【作者贡献声明】:刘建武收集病例资料、撰写论文、修改论文;何婷、况莹收集病例资料;朱健明、陈志华修改论文。

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(2022-03-23 收稿,2024-01-22 修回)