

颅骨浆细胞肿瘤 1 例

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【摘要】 浆细胞瘤又称骨髓瘤,来源于骨髓内的浆细胞。颅骨浆细胞肿瘤只占髓外浆细胞肿瘤的极少部分,属临床罕见疾病。本文报道 1 例颅骨将细胞肿瘤,62 岁女性,因头部包块伴疼痛不适 6 个月余入院。额部头皮包块大小约 1.0 cm×2.0 cm,质地韧,界限不清,轻微压痛。颅脑 CT 平扫发现颅骨多发骨质破坏并额部软组织结节。颅脑 MRI 平扫+增强发现颅骨多发骨质破坏区并局部软组织结节形成。术前考虑嗜酸性肉芽肿。完善术前准备后,予以颅骨病损切除+颅骨成形术治疗,全切除额部病灶,术后病理检查证实为浆细胞肿瘤。术后规律行达雷妥尤单抗靶向治疗+伊沙佐米化疗,术后 16 个月复查颅脑 MRI 平扫未发现肿瘤复发。术后 31 个月电话随访显示,病人的右下肢偶有疼痛,病人可拄拐行走。总之,颅骨浆细胞肿瘤极为少见,影像学检查可为手术治疗提供定位诊断,但病理检查仍是诊断金标准,术后结合病理检查结果进行放化疗,可改善病人预后。

【关键词】 颅骨肿瘤;浆细胞瘤;骨髓瘤;显微手术;疗效
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Plasma cell tumor of the skull: a case report

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【Abstract】 Plasmacytoma, also known as myeloma, is derived from plasma cells in the bone marrow. Skull plasma cell tumors account for only a small part of extramedullary plasma cell tumors, which is a rare disease in clinic. This paper reported a case of skull plasma cell tumor, a 62-year-old female, who was admitted to hospital due to head mass accompanied by pain and discomfort for more than 6 months. The size of the frontal scalp mass was about 1.0 cm×2.0 cm, with tough texture, unclear boundary, and slight tenderness. The CT found multiple bone destruction of the skull and frontal soft tissue nodules. The plain scan+contrast-enhanced MR images found multiple bone destruction areas of the skull and local soft tissue nodules. Eosinophilic granuloma was considered before operation. After completing preoperative preparation, skull lesion resection+cranioplasty was performed, and the total resection of the frontal lesions was confirmed by postoperative pathological examination as plasma cell tumor. After the regular treatment of daratumumab targeted therapy+ixazomib chemotherapy, no tumor recurrence was found in the reexamination of plain scan MR images 16 months after the operation. A telephone follow-up at 31 months postoperatively showed that the patient had occasional pain in the right lower limb and could walk with a crutch. In summary, skull plasma cell tumors are extremely rare, imaging examination can provide localization diagnosis for surgical treatment, but pathological examination is still the gold standard for diagnosis, postoperative radiotherapy and chemotherapy combined with pathological examination results can improve the prognosis of patients.

【Key words】 Skull neoplasms; Plasma cell tumor; Plasmacytoma; Myeloma; Microsurgery; Efficacy

1 病例资料

62 岁女性,因发现头部包块伴疼痛进行性加重 6 个月于 2020 年 11 月 6 日入院。起初,症状为无明显诱因发现头部包块,不可移动,伴疼痛不适,有压痛,发作时无明显加重或缓解因素,疼痛可忍耐,可自行缓解,未予重视。后来,上述症状逐渐加重,遂来我院就诊。入院体格检查:神志清楚,GCS 评分 15 分;额部处可见一突出于头皮包块,大小约 1.0 cm×2.0 cm,质地韧,界限不清,轻微压痛。颅脑 CT 平扫发现颅骨

多发骨质破坏并额部软组织结节(图 1A、1B)。颅脑 MRI 平扫+增强显示颅骨多发骨质破坏区并局部软组织结节形成(图 1C~F)。术前考虑嗜酸性肉芽肿。完善术前准备后,行颅骨病损切除+颅骨成形术。术中见额骨病灶膨胀性生长,质地韧,血供丰富,角突处病灶可见周围骨质板障侵蚀,病灶向内侵蚀进入硬膜,予以人工脑膜修补破损硬膜,钛网修补颅骨缺损。术后病理结果:(颅骨病损组织)肿瘤;免疫组化结果:肿瘤细胞呈 CD38(+),CD56(+),CD138(局灶+),MUM1(弱+),CD10(局灶+),λ(+),κ(-),CD20(-),CD79α(-),CK(-),Vim(+),Ki-67(约 30~40%+);病理结果结合形态学及免疫组化结果,诊断为:(颅骨病损组织)浆细胞肿瘤。术后复查颅脑 CT(图 1G、1H)见额部颅骨病损已切除。术后原病灶部位疼痛消失。术后规律行“达雷妥尤单抗靶向治疗+伊沙佐米化疗”,术后 16 个月复查颅脑 MRI 平扫未见肿瘤复发、软组织结节仍存在(图 1I)。术后电话随访 31 个

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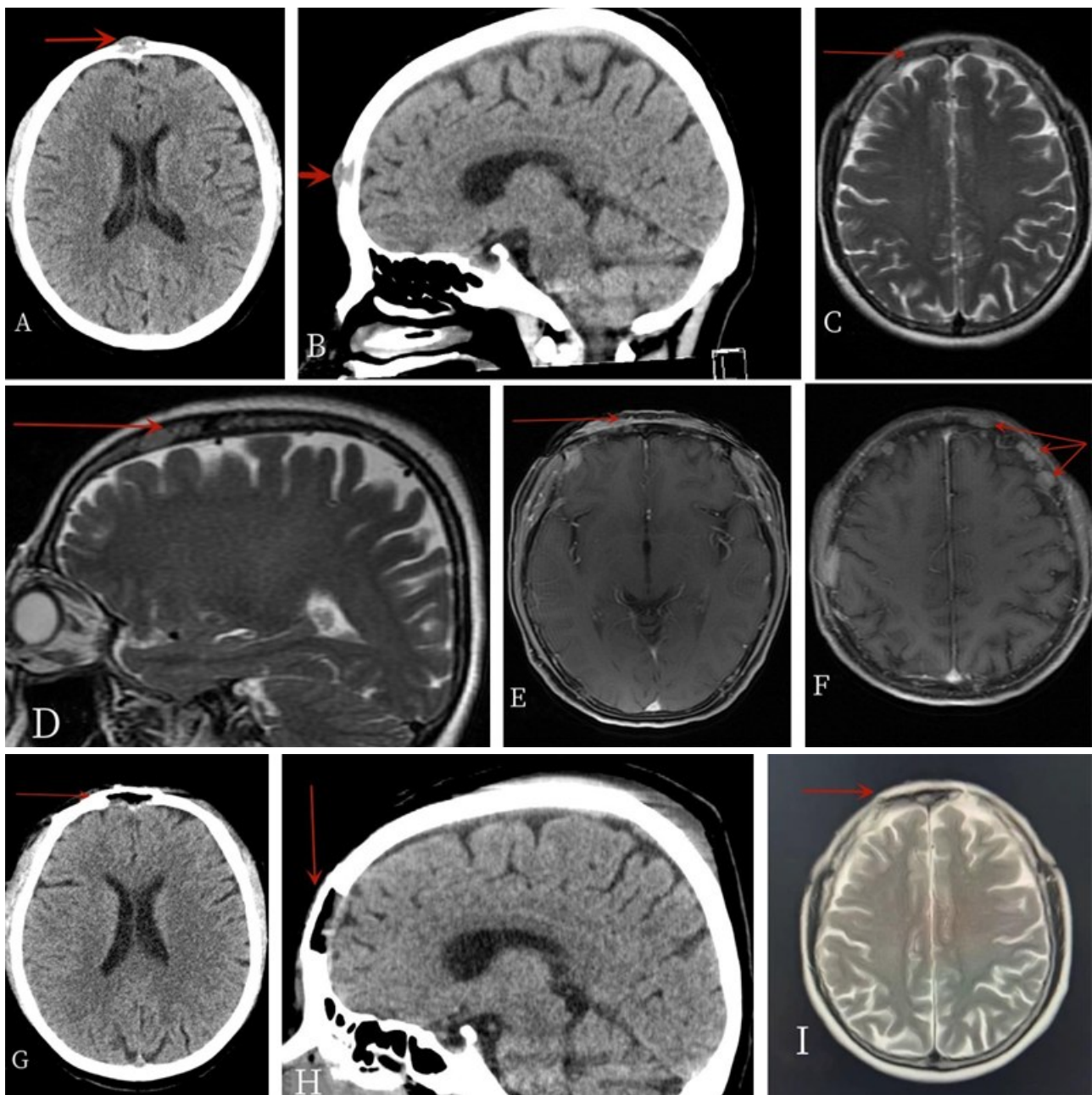


图 1 1 例额部颅骨浆细胞肿瘤手术前后影像表现

A、B. 术前颅脑 CT 平扫见额部颅骨破坏(红色 ↑ 示);C、D. 术前颅脑 MRI 平扫可见颅骨多发破坏并软组织结节形成(红色 ↑ 示);E、F. 术前颅脑 MRI 增强可见颅骨多发骨质破坏及多发软组织结节形成(红色 ↑ 示);G、H. 术后复查颅脑 CT 可见颅骨病损病灶切除,病损处颅骨予钛网修补(红色 ↑ 示);I. 术后 16 个月复查颅脑 MRI 平扫见软组织结节仍存在(红色 ↑ 示)

Figure 1 Imaging features of a case of frontal skull plasma cell tumor

A-B: Preoperative CT images showed frontal skull destruction (shown in red ↑). C-D: Preoperative plain scan MR images showed multiple skull destruction and soft tissue nodule formation (shown in red ↑). E-F: Preoperative contrast-enhanced MR images showed multiple skull bone destruction and multiple soft tissue nodule formation (shown in red ↑). G-H: Postoperative CT images showed skull lesion resection, and the skull lesion was repaired with titanium mesh (shown in red ↑). I: Postoperative plain MR images showed soft tissue nodule still existing 16 months after surgery (shown in red ↑).

月,右下肢偶有疼痛,可拄拐行走。

2 讨论

浆细胞肿瘤好发于血液系统^[1],以颅骨病变为首发症状临床少见^[2,3]。颅骨浆细胞肿瘤是浆细胞肿瘤在颅骨生长的表现,颅骨呈溶骨性破坏,若肿瘤压迫硬脑膜及脑组织、产生占位效应,则可出现神经系统症状。颅骨浆细胞肿瘤在骨质破坏后,被软组织填充,软组织肿块及骨质破坏区界限往往比较清楚,MRI 增强可见肿块信号明显强化,信号常均匀一致^[4]。本文病例的影像学表现与此一致。

颅骨浆细胞肿瘤临床少见,临床表现及影像学表现特异性不高,术前易误诊^[5],常误诊为慢性硬膜外机化血肿、脑膜瘤、嗜酸性肉芽肿、骨转移瘤、骨肉瘤等。病理学上看来,多数浆细胞肿瘤呈弥漫性分布、并具有较典型的浆细胞分化特点^[6]。Singh 等^[7]报道免疫组化染色 CD138 阳性是诊断浆细胞瘤的可靠标志物。为进一步了解病变与颅骨、硬膜及脑组织关系,术前颅脑 MRI 平扫及增强为必不可少的检查,可清楚显示肿瘤的大小、形态以及对周围组织压迫等,为手术提供正确的影像学定位。

本文病例主要表现为头部包块,伴有压痛;颅脑 CT 及 MRI 检查见颅骨多发骨质破坏并软组织结节形成,软组织结节明显强化,不排除嗜酸性肉芽肿。影像学结果与术后病理不符,影像学考虑嗜酸性肉芽肿通常会表现为颅骨骨质破坏伴软组织肿块,MRI 呈长 T₁、长 T₂ 信号,极易与该病混淆,只有依靠术后病理检查明确诊断。因浆细胞肿瘤血供丰富,可侵蚀颅骨及硬膜,手术出血量大,应特别注意控制出血。本文病例由于止血得当,手术失血约 50 ml。

对于颅骨浆细胞肿瘤,颅脑影像学检查可清晰显示颅骨破坏程度、浸润范围及肿瘤与临近组织的关系,有助于降低手术风险、提高手术安全性。目前,手术尽早切除病变,可有效避免神经功能损害加重。浆细胞肿瘤分型不同,治疗及预后亦不同,蛋白酶体抑制剂硼替佐米应用于多发性骨髓瘤,明显延长生存期^[8,9]。本文病例术后化疗使用伊沙佐米,亦为蛋白酶体抑制剂,随访 16 个月未见肿瘤复发。

总之,颅骨浆细胞肿瘤临床较少,术前难以准确诊断,最终诊断仍依靠病理检查。术前影像学检查可明确病灶及毗邻关系,为选择治疗方式提供参考。充分的术前准备,可有

效降低手术风险。术中控制出血,全切除肿块,术后辅以个体化局部放疗和化疗,可改善病人预后。

【利益冲突声明】:本文不存在任何利益冲突。
【作者贡献声明】:万豪杰负责病例资料采集,撰写论文;韩锋负责论文修订;杨华、杨明、罗登建、黄睿、陈伟参与文章讨论,提出修改意见。

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