.个案报道.

颅-鼻-眶非霍奇金B细胞淋巴瘤1例

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【摘要】颅-鼻-眶非霍奇金淋巴瘤为原发性中枢神经系统淋巴瘤，属结外非霍奇金淋巴瘤的一种，临床罕见，约占所有颅内恶性肿瘤1.6%，占结外淋巴瘤的1%~2%。该病恶性程度极高，手术旨在明确病理及减轻肿瘤占位效应，主要以放化疗为主，但总体预后较差。本文报道1例颅-鼻-眶非霍奇金B细胞淋巴瘤，66岁女性，行手术活检，术后病理证实为非霍奇金B细胞淋巴瘤，术后随访1个月死亡。总之，颅-鼻-眶非霍奇金大B细胞淋巴瘤临床较少见，多被误诊为鼻咽癌、眼眶肿瘤及球后视神经炎等，多数行病理检查可得到明确诊断。该病恶性程度极高，单纯依靠手术并不能明显延长生存期，主要以放化疗为主，有利于局部病灶的控制，但总体预后较差。所以正确早期诊断该病非常重要。

【关键词】颅-鼻-眶肿瘤；非霍奇金淋巴瘤；治疗；预后

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Cranial-nasal-orbital extranodal marginal zone B-cell lymphoma: a case report

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【**Abstract**】Cranio-nasal-orbital non-Hodgkin's lymphoma, as a form of primary central nervous system lymphoma and a subtype of extranodal non-Hodgkin's lymphoma, is clinically rare, accounting for approximately 1.6% of all intracranial malignant tumors and 1%~2% of extranodal lymphomas. The malignancy of this disease is extremely high. The aim of surgery is to clarify the pathology and alleviate the space-occupying effect of the tumor. The main treatment modalities are radiotherapy and chemotherapy, yet the overall prognosis is unfavorable. This paper presents a case of cranio-nasal-orbital non-Hodgkin's B-cell lymphoma in a 66-year-old female. A surgical biopsy was performed, and the postoperative pathology confirmed non-Hodgkin's B-cell lymphoma. The patient died one month after the surgery. In conclusion, cranio-nasal-orbital non-Hodgkin's large B-cell lymphoma is relatively rare in clinical settings and is often misdiagnosed as nasopharyngeal carcinoma, orbital tumors, or retrobulbar optic neuritis, among others. A clear diagnosis can mostly be achieved through pathological examinations. The malignancy of this disease is extremely high. Relying solely on surgery does not significantly prolong the survival period. The main treatments are radiotherapy and chemotherapy, which are conducive to the control of local lesions, but the overall prognosis is poor. Hence, accurate and early diagnosis of this disease is of paramount importance.

【**Key words**】Cranio-nasal-orbital tumors; Non-Hodgkin's lymphoma; Treatment; Prognosis

1 病例资料

66岁女性，因突发视物模糊不清半个月于2021年2月21日入院。入院体格检查：左额部可扪及头皮肿物，质韧，触之无活动；双侧瞳孔不等大、不等圆，右侧直径2.5 mm，对光反射存在，右眼粗测视力正常；左侧瞳孔直径5 mm，直接对光反射消失，间接对光反射存在，左眼视力无光感，左眼睑下垂，左眼球活动受限。头颅CT扫描示颅-鼻-眶内肿瘤破坏骨质(图1A、1B)。头颅MRI平扫示颅-鼻-眶内肿瘤呈不均匀混杂信号，大小约0.6 cm×1.0 cm×1.2 cm；增强扫描后环内呈不强化，大小约1.4 cm×0.9 cm×0.6 cm，左侧视神经受压下移(图1C~E)。术前诊断颅-鼻-眶恶性肿瘤。2021年2月25日在局麻下行经鼻内穿刺活检术，肿物呈灰白样。术后病理显示示：(鼻腔肿物)非霍奇金大B细胞淋巴瘤，结合免疫组化结果，倾向弥漫大B细胞淋巴瘤；免疫组化：CK(-)，LCA(+)，CD20(+)，CD79α(+)，CD3(-)，CD5(-)，CD21(-)，Bcl-2(+)约95%，Bcl-6(+)约50%，CD10(-)，c-myc(+)约40%，MUM-1(+)，Ki-67(+)约90%。术后随访1个月死亡。

2 讨 论

颅-鼻-眶非霍奇金淋巴瘤为原发性中枢神经系统淋巴瘤，属结外非霍奇金淋巴瘤的一种，临床罕见，约占所有颅内恶性肿瘤1.6%，占结外淋巴瘤1%~2%[1]。该病无明显性别差异，发病年龄高峰在40~60岁。该病多以首诊部位就诊，大多就诊于眼科及耳鼻喉科，而非神经外科。其临床表现缺乏特异性，主要为视力、视野障碍、鼻塞、颅内压增高症状等，多因侵袭的部位不同而出现不同症状。

大B细胞淋巴瘤因细胞核大且不规则多形，呈弥漫性浸润增长，部分甚至完全破坏正常淋巴结或结外结构，常累及大脑及其它软组织在内的结外部分，原发部位可以发生在任何部位[2]。非霍奇金大B细胞淋巴瘤CT影像多无特异性，主要表现为相应部位占位及骨质破坏；头颅MRI T_1WI多呈低信号，T_2WI多呈不均匀相对低信号，增强后病灶呈明显均匀强化，边界清楚，DWI弥散受限，呈高信号[3]。本文病例术前头

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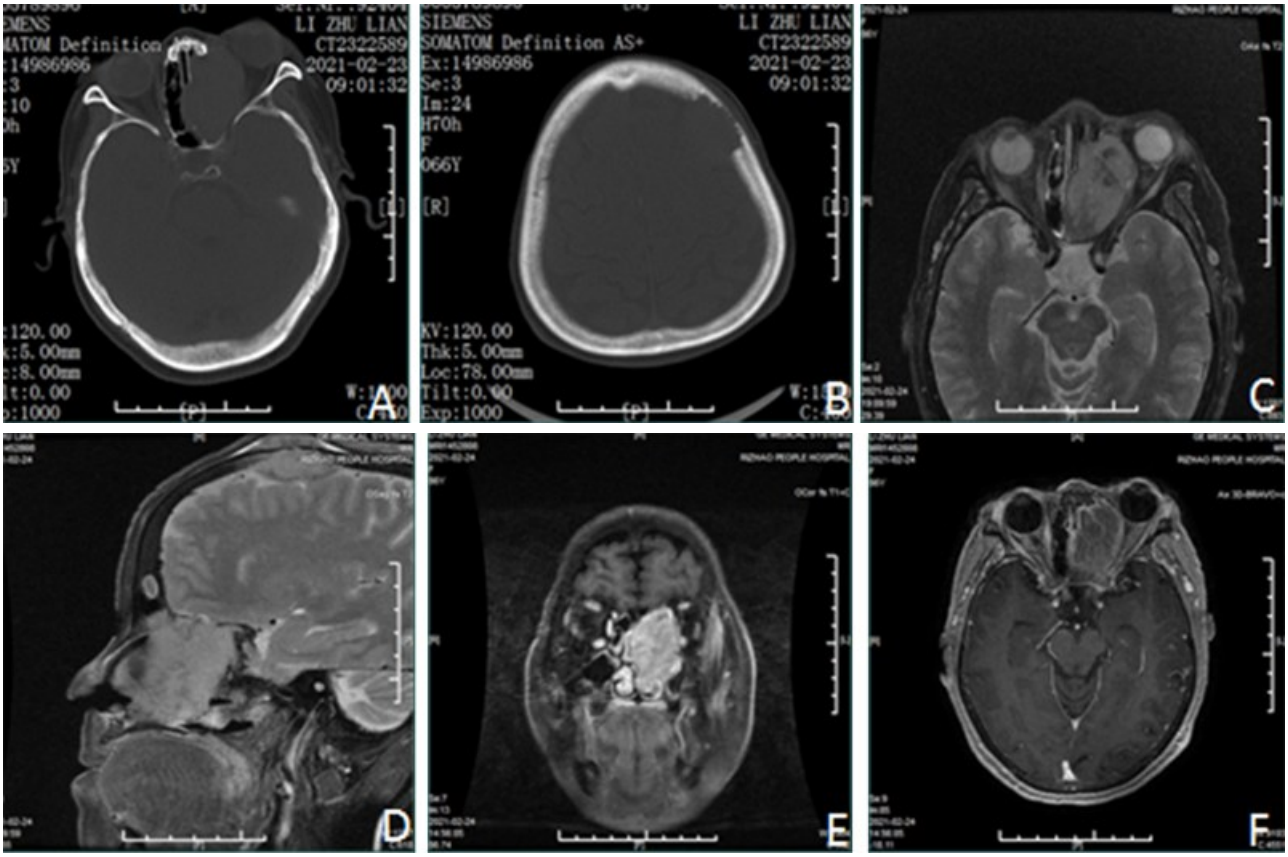

图1 颅−鼻−眶非霍奇金B细胞淋巴瘤影像

A、B. 头部CT显示颅−鼻−眶内肿瘤破坏骨质；C、D. MRI平扫显示颅−鼻−眶内肿瘤呈不均匀混杂信号；E、F. MRI增强扫描显示病灶强化较明显

Figure 1 Images of a patient with cranio−nasal−orbital Non−Hodgkin´s B−cell lymphoma

A−B: Head CT reveals bone destruction by the tumor within the cranio−nasal−orbital region. C−D: Plain MRI scan shows that the tumor in the cranio−nasal−orbital region presents heterogeneous and mixed signals. E−F: Enhanced MRI scan indicates a relatively obvious enhancement of the lesion.

颅MRI T_1WI呈低信号，T_2WI呈不均匀信号，强化后呈均匀强化，边界尚清楚。

总之，颅−鼻−眶非霍奇金大B细胞淋巴瘤临床较少见，多被误诊为鼻咽癌、眼眶肿瘤及球后视神经炎等。多数行病理检查可得到明确诊断。本文病例入院诊断眼眶肿瘤，术后经病理证实为非霍奇金大B细胞淋巴瘤。该病恶性程度极高，单纯依靠手术并不能明显延长生存期，多数学者认为非霍奇金大B细胞淋巴瘤治疗主要以放化疗为主，有利于局部病灶的控制，但总体预后较差。本文病人随访1个月死亡，所以正确早期诊断该病非常重要。

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