Case Report

Embryonal rhabdomyosarcoma of the paranasal sinuses: a case report and review of literature

Pei-Xue Wu1*, Yan-Fang Liang2*, Jin-Cheng Zeng3, Jian-Bo Ruan2, Dong-Ping Kang2, Can Chen2, Tao Zeng2, Qiu-Liang Wu4, Wei-Hua Xu5

1Department of Clinical Medicine, The Fourth Affiliated Hospital of Nanchang University, Nanchang, Jiangxi 330003, China; 2Department of Pathology, Taiping People’s Hospital of Dongguan, The Fifth People’s Hospital of Dongguan, Dongguan 523905, China; 3Guangdong Provincial Key Laboratory of Medical Molecular Diagnostics, Guangdong Medical College, Dongguan 523808, China; 4Department of Pathology, Sun Yat-Sen University Cancer Center, Guanzhou 510060, China; 5Department of Otolaryngology, Affiliated Hospital of Guandong Medical College, Zhanjiang 524023, China. *Equal contributors.

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Abstract: Embryonal rhabdomyosarcoma (ERMS) is a rare malignancy with a poor outcome. In this article, we describe a case of ERMS in the paranasal sinuses from a 60-year-old male patient. ERMS derived from the paranasal sinuses is extremely rare. The diagnosis of ERMS must be based on histological findings and immunohistochemical findings. In this case, microscopic observation showed tumor cells were arranged in flocked sheets, cord-like and acinar-like by hyperplastic fibrous tissue. And ERMS tissues were immunopositive for myogenin, desmin, MSA, CD56, vimentin, CD99, Syn and Ki-67 (40%+), and immunonegative for CK, EMA, LCA, GFAP, NSE, S-100, HMB-45 and Melan-A. Here, the patient was treated with multimodal therapy including endoscopic surgery, chemotherapy and radiation, but the patient’s postoperative recovery is not too smooth.

Keywords: Embryonal rhabdomyosarcoma, paranasal sinuses, immunohistochemistry

Introduction

Rhabdomyosarcoma (RMS) is a malignant tumor derived from the mesenchymal cells, with varying degrees of striated muscle cell differentiation and chromosomal or genetic imbalances [1, 2]. There are two principal types: alveolar RMS (ARMS) and embryonal RMS (ERMS). ERMS is a rare malignancy arising from undifferentiated mesoderm, and appears most frequently in children and adolescents, and more commonly occurs in men than women [3-5]. Although the head and neck, genitourinary system (bladder, prostate and vagina) and limbs are the most common site of ERMS, the ERMS derived from the paranasal sinuses is extremely rare, especially in the elderly [5-8]. Here, we report an ERMS of the paranasal sinuses in 60-year-old man presented as reddish tumors on the bilateral ethmoid and right sphenoid, discharging yellow sticky pus on the right frontal sinus.

Case report

A 60-year-old man had a symptom of bilateral nasal congestion nearly 20 days, right eye prominent with pain and diplopia (especially, when looking down) 1 week, smell disappeared, and having an intermittent headache from the right side of head. Specialist examination showed tenderness (+) on the right maxillary, frontal and ethmoid area, and several enlarged, rigid and flexible lymph nodes with medium quality, approximately 3 cm in diameter, on the right submandibular. Reddish tumors on the bilateral ethmoid and right sphenoid, and yellow sticky pus secretions on the right frontal sinus were seen in the operative. Patient with endoscopic surgery on the paranasal sinuses were carried out.

Gross findings

A mass of pale or gray mixed red crushed tissue measured 3 cm × 2.5 cm × 0.6 cm in volume were taken out on the right paranasal sinuses. And, a mass of pale or gray mixed red crushed tissue measured 2.5 cm × 2.5 cm × 0.7 cm in volume were taken out from left ethmoid (Figure 1A). The patient was diagnosed as having a tumor in the paranasal sinuses.
Histopathological findings

The tumor tissues were stained with hematoxylin and eosin (Figure 1B and 1C). Microscopic observation showed tumor cells were arranged in flocked sheets, cord-like and acinar-like by hyperplastic fibrous tissue. A visible focal necrosis was easy to see on the tumor tissues.
Partly spindle or oval tumor cells displayed moderately pleomorphic nuclei with unintelligible cytoplasm but activity mitosis. Other fusiform, ribbon or tadpole-like tumor cells displayed abundant amounts of eosinophilic cytoplasm.

**Immunohistochemical findings**

Tumor cells were positive for myogenin, desmin, MSA, CD56, vimentin, CD99, Syn and Ki-67 (40%) (Figure 2), and negative for CK, EMA, LCA, GFAP, NSE, S-100, HMB-45 and Melan-A (Figure 3).

**Discussion**

It is important to distinguish ERMS from olfactory neuroblastoma (ONB), Ewing’s sarcoma/peripheral primitive neuroectodermal tumor (PNET), malignant melanoma (MM) and small cell carcinoma (SACC). ONB is a malignant tumor usually occurs in human upper nasal cavity, appears most frequently in children under 5-year-old [8]. Histological analysis indicates that small round tumor cells often forming rosettes separated by a fibrovascular stroma [9]. PNET belongs to malignant tumors and is composed of small round, tightly packed tumor cells of a neuroectodermal origin. Immunohistochemical analysis indicates that PNET tumor tissues were immunopositive for CD99, but immunonegative for myogenic markers like myogenin [10]. MM is a malignancy associated with a high mortality rate, also commonly occurs in the nasal cavity and paranasal sinuses [11]. Immunohistochemical analysis is useful to distinguish ERMS from MM in nasal cavity and paranasal sinuses, because MM tumor tissues were immunopositive for S-100 and HMB-45, and immunonegative for Myogenin and desmin [11]. Although SCC of the paranasal sinuses is extremely rare, the discrimination of ERMS from SCC is essential. SCC appears most frequently in the elderly. Immunohistochemical analysis shows SCC tumor tissues were immunopositive for epithelial markers like EMA [12]. In this case, microscopic observation showed tumor cells were arranged in flocked sheets, cord-like and acinar-like by hyperplastic fibrous tissue. And ERMS tissues were immunopositive for Myogenin (+), desmin (+), SMA (+), CD56 (+), Vimentin (+), CD99 (+), Syn (+) and Ki-67 (40%+), and immunonegative for CK (-), EMA (-), LCA (-), GFAP (-), NSE (-), S-100 (-), HMB-45 (-) and Melan-A (-), so the diagnosis and distinguish of ERMS must base on histological findings and immunohistochemical findings.

RMS is the most common soft-tissue malignancy, the 5-year failure-free survival rate for this malignancy is approximately 70% [13]. ERMS in the sphenoid or ethmoid sinus is a rare tumor that can respond well to radiation and chemotherapy [14]. However, endoscopic surgery of the ERMS is indicated after chemotherapy or radiation. Here, the patients were treated with multimodal therapy including endoscopic surgery, chemotherapy and radiation, but the patient’s postoperative recovery is not too smooth.

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**Disclosure of conflict of interest**

None.

**Address correspondence to:** Dr. Wei-Hua Xu, Department of Otolaryngology, Affiliated Hospital of Guangdong Medical College, Zhanjiang 524023, China. E-mail: xwhua302@tom.com

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