

Case Report

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

Pei-Xue Wu^{1*}, Yan-Fang Liang^{2*}, Jin-Cheng Zeng³, Jian-Bo Ruan², Dong-Ping Kang², Can Chen², Tao Zeng³, 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, Guangzhou 510060, China; ⁵Department of Otolaryngology, Affiliated Hospital of Guangdong Medical College, Zhanjiang 524023, China. *Equal contributors.

Received June 13, 2014; Accepted July 10, 2014; Epub August 15, 2014; Published August 30, 2014

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.

Embryonal rhabdomyosarcoma of the paranasal sinuses

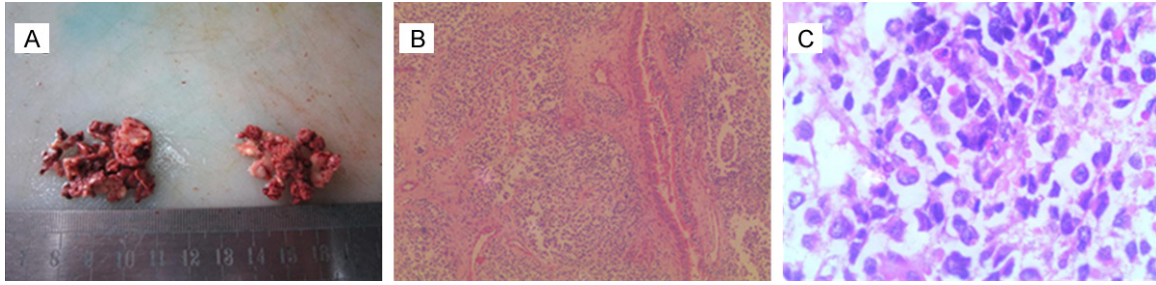


Figure 1. The ERMS tissues were stained with hematoxylin and eosin. A: A mass of pale or gray mixed red crushed tissue from the right paranasal sinuses and a mass of pale or gray mixed red crushed tissue from left ethmoid. B: Tumor cells were arranged in flocked sheets, cord-like and acinar-like by hyperplastic fibrous tissue (40 ×). C: 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 (100 ×).

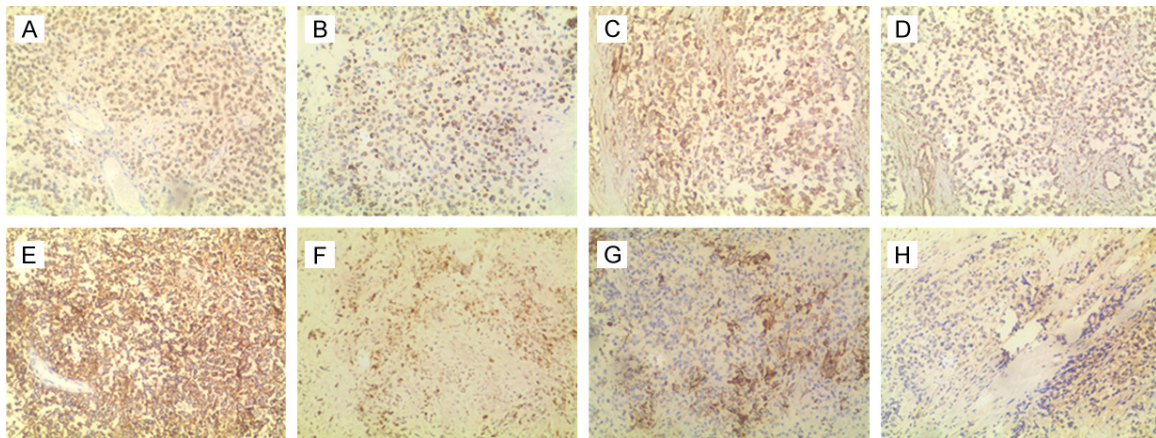


Figure 2. ERMS tissues were stained with immunohistochemistry. The tissues were immunopositive for myogenin (A), desmin (B), MSA (C), CD56 (D), vimentin (E), CD99 (F), Syn (G) and Ki-67 (H).

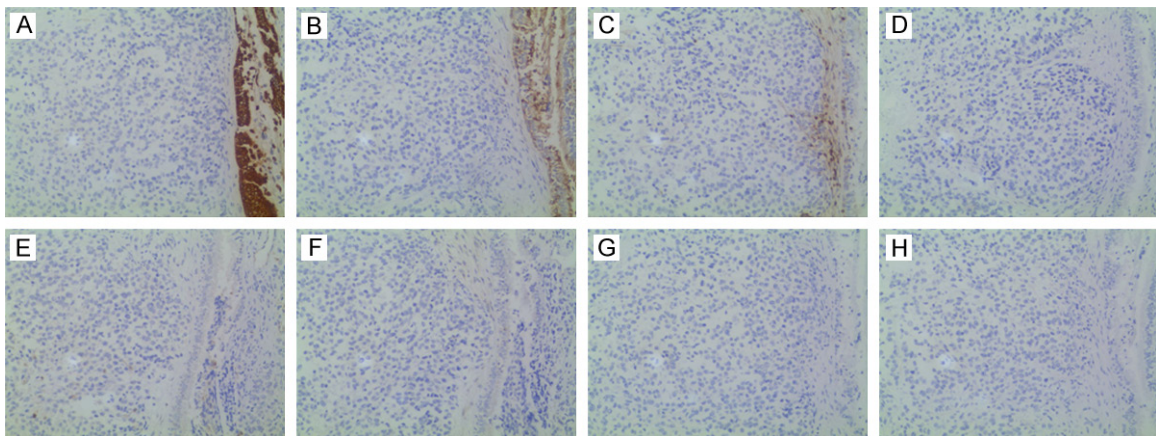


Figure 3. ERMS tissues were stained with immunohistochemistry. The tissues were immunonegative for CK (A), EMA (B), LCA (C), GFAP (D), NSE (E), S-100 (F), HMB-45 (G) and Melan-A (H).

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.

Embryonal rhabdomyosarcoma of the paranasal sinuses

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 (SCC). 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.

Acknowledgements

This work was supported by National Natural Science Foundation of China (No. 81201831), Dr Startup project of Science Research Foundation of GDMC (No. B2011021) and the Science and Technology Project of Dongguan (20131051010006).

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

References

- [1] Linardic CM, Downie DL, Qualman S, Bentley RC, Counter CM. Genetic modeling of human rhabdomyosarcoma. *Cancer Res* 2005; 65: 4490-4495.
- [2] Liu C, Li D, Hu J, Jiang J, Zhang W, Chen Y, Cui X, Qi Y, Zou H, Zhang W, Li F. Chromosomal and genetic imbalances in Chinese patients with rhabdomyosarcoma detected by high-resolution array comparative genomic hybridization. *Int J Clin Exp Pathol* 2014; 7: 690-698.
- [3] Maurya OP, Patel R, Thakur V, Singh R, Kumar M. Embryonal rhabdomyosarcoma of orbit—a case report. *Indian J Ophthalmol* 1990; 38: 202-4.
- [4] Dehner LP, Jarzembowski JA, Hill DA. Embryonal rhabdomyosarcoma of the uterine cervix: a report of 14 cases and a discussion of its unusual clinicopathological associations. *Mod Pathol* 2012; 25: 602-614.
- [5] Haider N, Nadim MS, Piracha MN. Primary embryonal rhabdomyosarcoma of the liver in a

Embryonal rhabdomyosarcoma of the paranasal sinuses

- young male. *J Coll Physicians Surg Pak* 2013; 23: 750-751.
- [6] Arul AS, Verma S, Arul AS, Verma R. Oral rhabdomyosarcoma-embryonal subtype in an adult: A rarity. *J Nat Sci Biol Med* 2014; 5: 222-225.
- [7] Li D, Li Y, Wang K, Li H, Tang Y, Wei X, Wang Y. Embryonal rhabdomyosarcoma of the tunica dartos in the scrotum. *J Ultrasound Med* 2011; 30: 105-109.
- [8] Cohen M, Ghosh L, Schafer ME. Congenital embryonal rhabdomyosarcoma of the hand and Apert's syndrome. *J Hand Surg Am* 1987; 12: 614-617.
- [9] Papacharalampous GX, Vlastarakos PV, Chrysovergis A, Saravakos PK, Kotsis GP, Davilis DI. Olfactory neuroblastoma (esthesioneuroblastoma): towards minimally invasive surgery and multi-modality treatment strategies - an updated critical review of the current literature. *J BUON* 2013; 18: 557-563.
- [10] Mao Y, Sang X, Liang N, Yang H, Lu X, Yang Z, Du S, Xu Y, Zhao H, Zhong S, Huang J, Millis JM. Peripheral primitive neuroectodermal tumors arising in the pancreas: the first case report in Asia and a review of the 14 total reported cases in the world. *Hepatobiliary Surg Nutr* 2013; 2: 51-60.
- [11] Asai N, Ohkuni Y, Kawamura Y, Kaneko N. A case of obstructive sleep apnea syndrome caused by malignant melanoma in the nasal cavity and paranasal sinus. *J Cancer Res Ther* 2013; 9: 276-277.
- [12] Terada T. Small cell carcinoma of the oral cavity (cheek mucosa): a case report with an immunohistochemical and molecular genetic analysis. *Int J Clin Exp Pathol* 2013; 6: 780-787.
- [13] Raney RB, Meza J, Anderson JR, Fryer CJ, Donaldson SS, Breneman JC, Fitzgerald TJ, Gehan EA, Michalski JM, Ortega JA, Qualman SJ, Sandler E, Wharam MD, Wiener ES, Maurer HM, Crist WM. Treatment of children and adolescents with localized parameningeal sarcoma: experience of the Intergroup Rhabdomyosarcoma Study Group protocols IRS-II through-IV, 1978-1997. *Med Pediatr Oncol* 2002; 38: 22-32.
- [14] Luu QC, Lasky JL, Moore TB, Nelson S, Wang MB. Treatment of embryonal rhabdomyosarcoma of the sinus and orbit with chemotherapy, radiation, and endoscopic surgery. *J Pediatr Surg* 2006; 41: e15-17.