Case report

Sinonasal teratocarcinosarcoma in a 57-year-old Thai man: a case report of an extremely rare malignant sinonasal tumor

Piriya Sutthiruangwong[1], Thirayost Nimmanon[2], Sitthi Sukauichai[3] and Kulachet Wiwatwarayos[1]

[1]Institute of Pathology, Department of Medical Services, Ministry of Public Health; [2]Department of Pathology, Phramongkutklao College of Medicine; [3]Department of Medicine, Chonburi Cancer Hospital

Abstract:

Sinonasal teratocarcinosarcoma (TCS) is a rare aggressive malignant tumor. This tumor is exclusively found in the sinonasal tract. Clinical manifestations are associated with mass effects, such as nasal obstruction, headache and epistaxis.

Histopathologically, the tumor consists of a mixture of epithelial, mesenchymal, and neuroepithelial elements.

In this report, we are presenting a case of sinonasal TCS a 57-year-old Thai man, who presented with epistaxis. The nasal mass extended to the right anterior cranial fossa.

Keywords: ● *Sinonasal mass* ● *Epithelial element* ● *Mesenchymal element* ● *Neuroepithelial element*

RTA Med J 2023;76(2):75-80.

Received 10 July 2022 Corrected 27 April 2023 Accepted 27 June 2023

Correspondence should be addressed to Kulachet Wiwatwarayos, MD., Institute of Pathology, Department of Medical Services, Ministry of Public Health, Bangkok

รายงานผู้ป่วย

มะเร็งโพรงจมูกชนิดพบได้ยาก Sinonasal Teratocarcinosarcoma ในผู้ป่วยชายไทย อายุ 57 ปี

พิริยา สุทธิเรืองวงศ์[1] ธีรยสถ์ นิมมานนท์[2] สิทธิ สุขอวยชัย[3] และ กุลเชษฐ์ วิวัฒน์วรายศ[1]

[1]สถาบันพยาธิวิทยา กรมการแพทย์ กระทรวงสาธารณสุข [2]ภาควิชาพยาธิวิทยา กองการศึกษา วิทยาลัยแพทยศาสตร์พระมงกุฎเกล้า

[3]กลุ่มงานอายุรศาสตร์ โรงพยาบาลมะเร็งชลบุรี

บทคัดย่อ

Sinonasal teratocarcinosarcoma (TCS) เป็นมะเร็งที่พบได้น้อยมาก มีความร้ายแรงและมีอัตราการตายสูง ที่ผ่านมามีรายงานการเกิดมะเร็งชนิดนี้เฉพาะในโพรงจมูก และบริเวณไซนัส ผู้ป่วยมักมาด้วยอาการที่เกิดจากการอุดตันของโพรงจมูก อาการปวดศีรษะ โดยผู้ป่วยบางรายพบเลือดกำเดาไหลได้ด้วย

มะเร็งชนิดนี้ประกอบด้วยสามส่วนหลัก คือ primitive neuroepithelial elements, มะเร็งของเยื่อบุผิว(malignant epithelium element/carcinoma), และส่วนของมะเร็งเนื้อเยื่อเกี่ยวพัน (mesenchymal elements/sarcoma)

ในรายงานผู้ป่วยนี้ ขอนำเสนอผู้ป่วยชายไทย อายุ 57 ปี มีอาการเลือดกำเดาไหล ตรวจร่างกายพบก้อนที่จมูก และรุกรานไปยัง right anterior cranial fossa

คำสำคัญ: ● *Sinonasal mass* ● *Epithelial element* ● *Mesenchymal element* ● *Neuroepithelial element*

เวชสารแพทย์ทหารบก 2566;76(2):75-80.

ได้รับต้นฉบับ 10 กรกฎาคม 2565 แก้ไขบทความ 27 เมษายน 2566 รับลงตีพิมพ์ 27 มิถุนายน 2566

ต้องการสำเนาต้นฉบับติดต่อ นพ.กุลเชษฐ์ วิวัฒน์วรายศ สถาบันพยาธิวิทยา กรมการแพทย์ กระทรวงสาธารณสุข กรุงเทพฯ

Introduction

Sinonasal teratocarcinosarcoma (TCS) is a rare aggressive malignant tumor that, as the name indicates, exclusively involves the sinonasal tract[1-3]. This tumor is most frequently located in the superior aspect of the nasal cavity with frequent extension into the ethmoid, maxillary, or sphenoid sinuses[1]. Nasopharyngeal involvement is rare[4].

TCS usually arises in the middle-aged men, most commonly between 50-55 years of age, but it can be found in almost every age group, ranging from 10 to 82 years of age[1,4,5]. Clinical manifestations are associated with mass effects, such as nasal obstruction and headache. Some patients have epistaxis[1,2,6-8].

By definition, the tumor comprises three major parts, including the primitive neuroepithelial element (terato-), the malignant epithelial component (carcino-) and the malignant mesenchymal component(sarcoma).

Pathogenetically, this tumor has been associated with biallelic inactivation of *SMARCA4* or activating *CTNNB1* mutation. As a result, the tumor cells are usually shown to have loss of *BRG1*(*SMARCA4*) nuclear expression (either complete or partial) or positive staining for β-catenin by immunohistochemistry, respectively[1,9-11]. In addition, an absence of 12p amplification have also been reported[5,12].

Surgery is considered the main therapeutic modality. Adjuvant therapies including radiotherapy and chemotherapy have been implicated in some reported patients[1,2,4,8,13,14].

Recently, a systematic review with survival analysis revealed a mean 2-year survival rate of 55%[15]. Some studies have reported a mean survival rate of 1.7 years with 60% mortality rate within 3 years[13,16]. However, some patients had a good outcome, having shown to be disease-free at 2 and 9 years in post-operative follow up[1]. Until the year 2021, only approximately 127 cases had been reported in English literatures[15]. Importantly, sinonasal TCS has never been reported in Thai population. We would therefore like to present a case of this unusual tumor.

Case Report

A 57-year-old Thai man presented with epistaxis. A physical examination revealed a nasal mass extending to right anterior cranial fossa. A computed tomography (CT) scan showed a soft tissue mass in the nasal cavity and the ethmoidal sinus with invasion into the skull base and right orbit. The patient was treated by craniotomy with right lateral rhinotomy and tumor removal.

The microscopic examination showed a mixture of three components, consisting of the teratoid component (primitive neuroectodermal cells with rosette formation) (Figure 1), the malignant epithelial component (malignant squamous cells and malignant glands) (Figure 2), and the sarcomatous component (chondroid differentiation) (Figure 3). The immunohistochemical study for BRG1(SMARCA4) was performed, showing a complete loss of expression in this case (Figure 4). The findings were all consistent with the diagnosis of sinonasal teratocarcinosarcoma (TCS).

Due to the large-sized tumor of 7.4 cm with right orbit and anterior cranial fossa extension, the multidisciplinary approach for the proper treatment for this patient was surgical debulking of the tumor with post-operative chemotherapy and radiotherapy. Sixteen months after treatment, the patient came to visit the clinic for the post-treatment follow-up with no evidence of residual or recurrent tumor.

Discussion

TCS is still considered to be one of the extremely rare tumors with aggressive behavior. In the literature, the term teratocarcinosarcoma was firstly described in 1984 by Heffner et al.[17]. Since then, many pathologists have

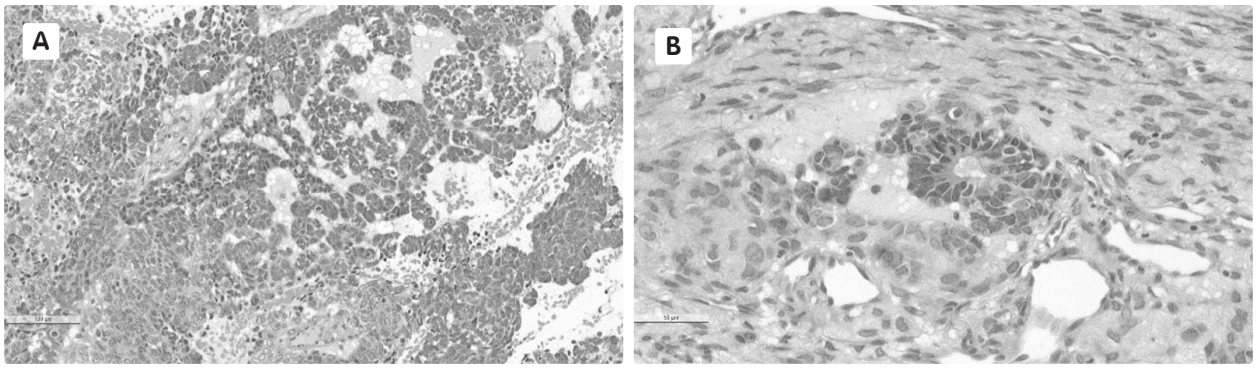

Figure 1 The primitive neuroectodermal component at 200x magnification (A) and 400x magnification with rosette formation (B) (H&E stain)

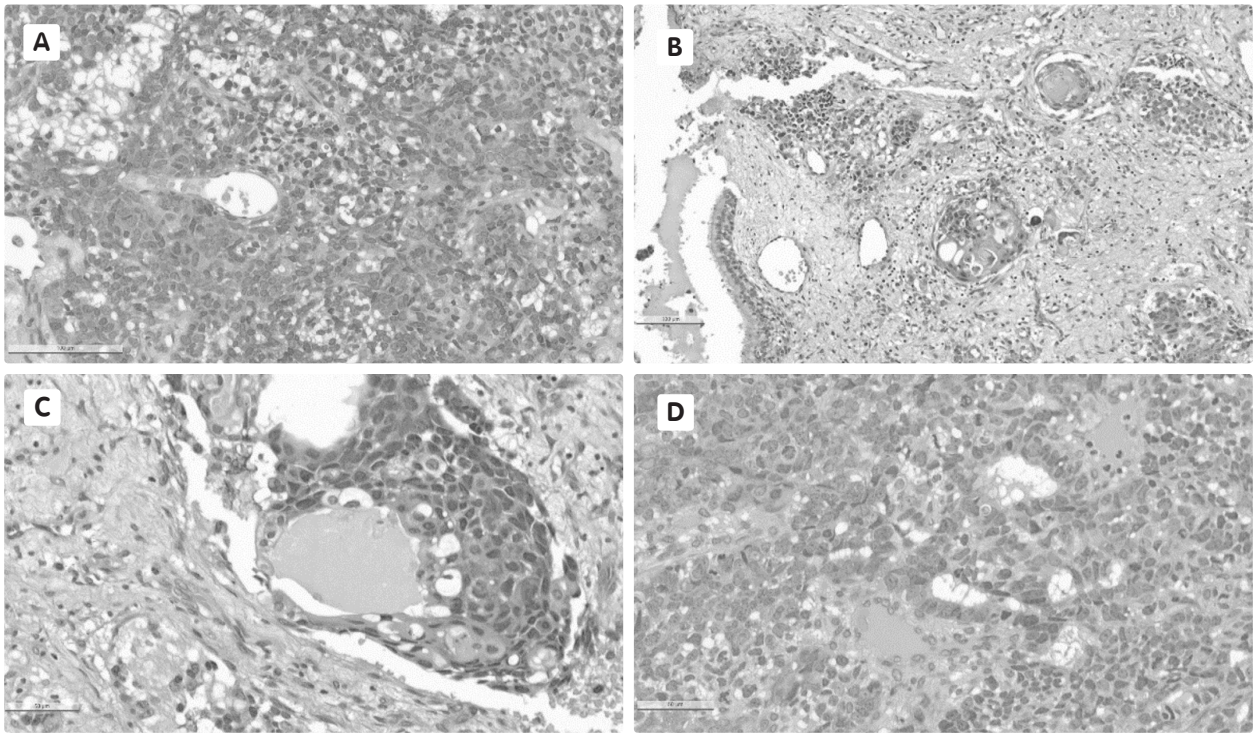

Figure 2 The carcinomatous component, consisting of malignant squamous cells (A-C) and glandular cells (D). The squamous and glandular elements have a clear-cell appearance, reminiscent of fetal tissues. (200x magnification (A-B) and 400x magnification (C-D) (H&E stain)

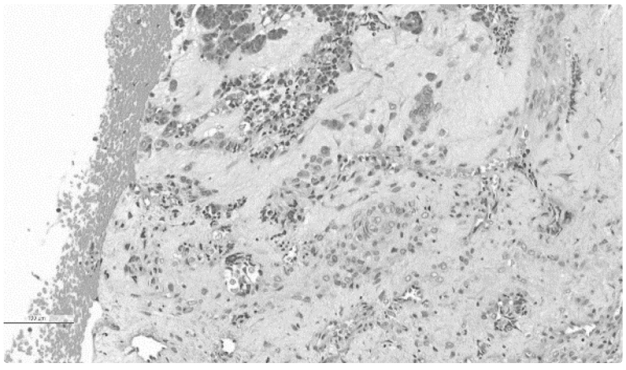

Figure 3 The sarcomatous component with chondroid differentiation at 200x magnification (H&E stain)

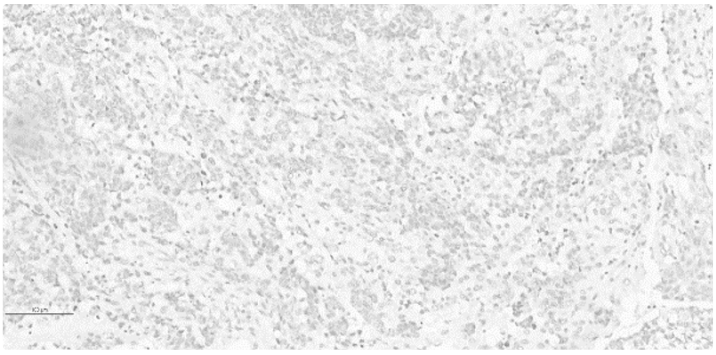

Figure 4 Loss of BRG1(SMARCA4) nuclear expression in the tumor area (intact nuclear stain in endothelial cells) (immunohistochemistry, x200 magnification)

become interested in it with an increasing number of published case reports with molecular data. According to the 5th edition of WHO classification of head and neck tumors, TCS was still considered as a distinct entity.

Histologically, the tumor comprises three different components, consisting of epithelial, mesenchymal, and neuroepithelial components. The epithelial part can be either squamous cell carcinoma, adenocarcinoma, poorly differentiated carcinoma, or mixed. Immature neoplastic squamous cells and the neuroectodermal component with rosette formation are included in the fetal appearance and teratoid features. The sarcomatous component may include poorly differentiated sarcoma-like areas/undifferentiated sarcoma, chondroid differentiation with foci of endochondral calcification, and muscular differentiation[3,18-21]. Consistent with these previous reports, we detected similar features within the tumor. In order to confirm the diagnosis, the immunohistochemistry study for BRG1(SMARCA4) were also performed in our case, showing a complete loss of its nuclear expression as expected.

Conclusion:

Sinonasal TCS is an aggressive malignant neoplasm with poor prognosis. Because this type of neoplasm is extremely rare, it can be easily misdiagnosed as another type of malignant sinonasal tumors, especially when a small biopsy is obtained. Making the correct diagnosis and providing early treatments with combined surgery and adjuvant therapy can reasonably improve the prognosis.

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