Acta Med. Okayama, 2019
Vol. 73, No. 6, pp. 529–532

Acta Medica Okayama
http://escholarship.lib.okayama-u.ac.jp/amo/

Case Report

An Adult Case of Nasal Chondromesenchymal Hamartoma: Imaging Characteristics Including Diffusion-Weighted Images

Takahiro Kitayama[a]*, Shiro Akaki[a], Kento Hisazumi[a], Kotaro Yoshio[a], Daisaku Inoue[a], Nobuhisa Tajiri[a], Tsuyoki Shiode[a], Susumu Kanazawa[b], Shohei Fujimoto[c], Kengo Kanai[c], and Yuji Hirata[c]

Departments of [a]Radiology, [c]Otorhinolaryngology, Kagawa Prefectural Central Hospital, Takamatsu 760-8557, Japan, [b]Department of Radiology, Okayama University Graduate School of Medicine, Dentistry and Pharmaceutical Sciences, Okayama 700-8558, Japan

Nasal chondromesenchymal hamartoma (NCMH), a rare, benign, nasal cavity tumor, typically occurs in children. Differential diagnosis is difficult because NCMH often presents with non-specific findings, including cystic components and invasion of the surrounding area on T2-weighted magnetic resonance images. Here, we present a rare adult case of NCMH, with no clear hyperintensity on diffusion-weighted images (DWI), and bone remodeling on the tumor margins on computed tomography. To the best of our knowledge, this is the first report of DWI on NCMH, and these findings, which suggest benign disease, may be useful in diagnosing NCMH.

Key words: nasal chondromesenchymal hamartoma, computed tomography, magnetic resonance imaging, diffusion-weighted imaging

Nasal chondromesenchymal hamartoma (NCMH) is a rare benign tumor of the nasal cavity. Since the first description of NCMH by McDermott *et al*. in 1998 [1], 48 cases have been reported worldwide. NCMH typically occurs in children, particularly infants aged up to 1 year, with only 8 adult cases reported to date [2]. Histologically, the tumors are composed of island cartilage as well as mesothelial components such as spindle cells and mucinous stroma. In diagnostic imaging, NCMH must be differentiated from a wide variety of tumors, including inverted papilloma, pleomorphic adenoma, squamous cell carcinoma, and adenoid cystic carcinoma. A histopathologic examination is required to reach a definitive diagnosis, and surgery is the primary treatment modality. No recurrences have been reported following complete resection with endoscopic nasal/paranasal surgery. Here, we report an adult case of NCMH that was difficult to differentiate from a malignant nasal/paranasal tumor. We also present the findings from diffusion-weighted images (DWI), which have not been previously reported.

Case Report

The patient was a 23-year-old man who had developed numbness and a feeling of pressure in his left cheek 3 months prior to his visit. He subsequently experienced severe pain behind the eye, occipital pain, lacrimation, and mild protuberance of the left eye, for which he was examined at our hospital's otolaryngology department. Nasopharyngeal fiberscopy showed a smooth-sided, protruding lesion on both sides of the nasal cavity, causing stenosis. Computed tomography (CT) showed a

Received February 15, 2019; accepted July 8, 2019.
*Corresponding author. Phone: +81-86-235-7313; Fax: +81-86-235-7316
E-mail: piwks0e1@icloud.com (T. Kitayama)

Conflict of Interest Disclosures: No potential conflict of interest relevant to this article was reported.

6×5×4 cm irregular tumor with internal calcification, centered in the nasal cavity. Crescent-shaped bone remodeling was observed at the tumor margins, and there was severe displacement of peripheral structures, particularly those in the left orbit (Fig. 1). On magnetic resonance imaging (MRI), the tumor displayed slightly heterogeneous intermediate intensity on T1-weighted images (WI) and heterogeneous hyperintensity on T2WI. Contrast enhancement was observed on the margins in contrast-enhanced T1WI. Intermediate intensity was observed on DWI, with no signs of reduced diffusion capacity. The apparent diffusion coefficient (ADC) was 2.23×10^{-3} mm^2/sec (Fig. 2). Repeated tissue biopsies were performed to reach a final diagnosis of NCMH. It was especially difficult to exclude chondrosarcoma, but the existence of spindle cells was the key to making a final diagnosis of NCMH. Endoscopic nasal/paranasal surgery was performed for maximal possible resection of the tumor, except for the areas near the orbits and the base of the skull. The tumor was removed as multiple fragments (Fig. 3). The residual tumor size increased while the patient was under outpatient observation, so 8 months after the initial surgery, another operation was performed. The patient is currently under observation.

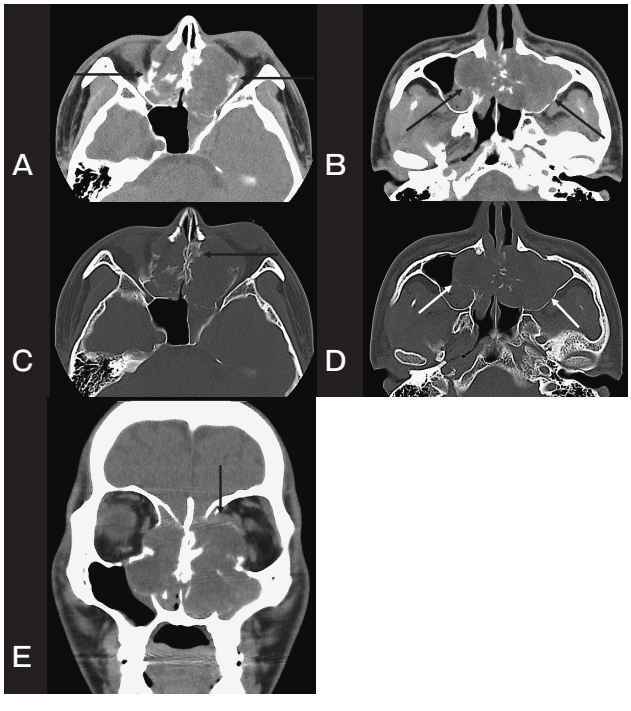

Fig. 1 Axial (**A**–**D**) and coronal (**E**) computed tomography images with soft-tissue and bone windows. A 6×5×4-cm soft-density tumor with distinct and relatively irregular margins centered on the nasal cavity was observed (black arrows in **A**, **B**). Destructive invasiveness of the nasal septum (black arrow in **C**) and other surrounding bone and crescent-shaped bone remodeling at the tumor margins (white arrows in **D**) were observed. Progression into the bilateral maxillary sinuses, orbits, and ethmoid sinuses was observed, along with severe displacement of the left orbital structures (black arrow in **E**). Internally, fine calcification was observed.

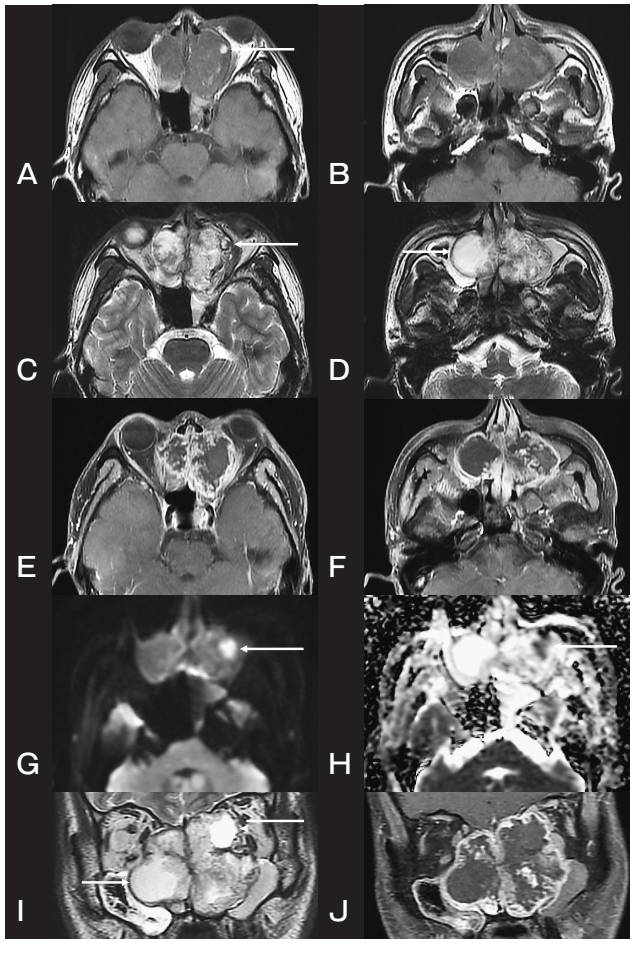

Fig. 2 T1-weighted axial images (**A**, **B**), T2-weighted axial images (**C**, **D**), contrast-enhanced axial images (**E**, **F**), diffusion-weighted image (DWI, b = 800 sec/mm^2; **G**), apparent diffusion coefficient (**ADC**) map (**H**), T2-weighted coronal image (**I**), and contrast-enhanced coronal image (**J**).
The tumor presented with slightly heterogeneous intermediate intensity on T1-weighted images (**A**, **B**) and heterogeneous hyperintensity and cystic formation (white arrows in **D** and **I**) on T2-weighted images (**C**, **D**, **I**). Strong contrast enhancement was observed on the margins in contrast-enhanced T1-weighted images (**E**, **F**, **J**). Intermediate intensity was observed on DWI, with no signs of reduced diffusion capacity (ADC = 2.23×10^{-3}mm^2/sec; **G**, **H**). Some areas were thought to represent subacute hemorrhage (white arrows in **A**, **C**, **G** and **H**).

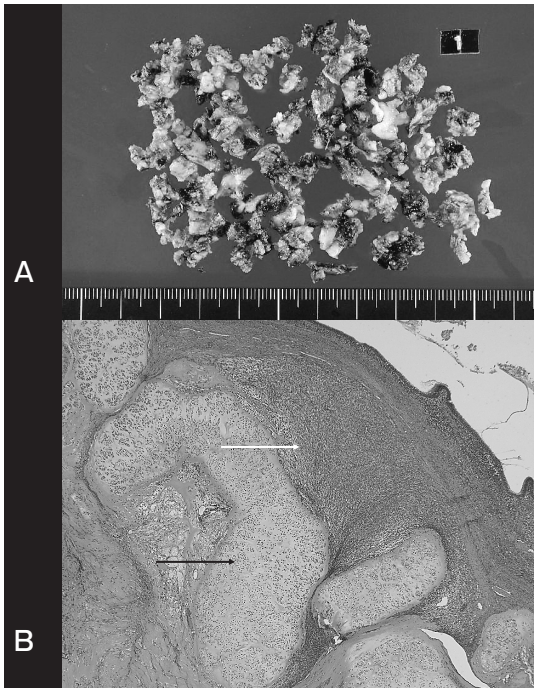

Fig. 3 Tissue fragments removed during surgery (**A**). Hematoxylin- and eosin-stained section of a lesion from the left-inferior nasal concha (**B**). The tumor is composed of spindle cells (white arrow in **B**) and island cartilage (black arrow in **B**).

Discussion

In 2009, Finitsis *et al.* reported CT findings from 23 NCMH cases [3]. The tumors ranged from 1.4 to 5.5 cm in size, exhibited contrast enhancement, and were often accompanied by calcification or cyst formation. The authors also reported bone remodeling and tumor progression into the orbits, paranasal sinuses, and cranium.

In addition, other typical CT findings included tumors with ill-defined margins and without capsules, often accompanied by cystic components. On MRI, T1WI showed heterogeneous intensity, and T2WI depicted cystic components. MRI also has the advantage of superior tissue characterisation and delineation of invasion of the surrounding area [2]. Calcification and bone remodeling are easy to assess on CT. On MRI, hypointense areas on T2WI are reported to correspond to cartilage and fibrosis, while hyperintensity on T2WI corresponds to cysts or mucous [4]. Thus, while a few reports have described the imaging characteristics of NCMH, all of these findings have been non-specific, making it difficult to differentiate NCMH from other nasal/paranasal tumors. Therefore, in addition to the tumor site and imaging characteristics, clinical information such as age, sex, and clinical course need to be considered during differential diagnosis. A histopathologic examination is required to reach a definitive diagnosis.

The present case also exhibited cystic components, progression into the surrounding area, and bone remodeling. In addition to these imaging findings, marginal enhancement in contrast-enhanced T1WI had a "cerebriform" appearance, so we considered inverted papilloma as a possible diagnosis [5]. Pleomorphic adenoma was also considered because the tumor with cystic components appeared to arise in the nasal septum [6]. However, it was difficult to exclude malignant tumors such as squamous cell carcinoma or adenoid cystic carcinoma because of the heterogenous intensity on T2WI and the invasion of the surrounding area [5,7]. It was difficult to make a diagnosis based solely on these images, and a biopsy had to be performed.

Differential diagnosis of NCMH is not only challenging because of its rarity, but also because the image findings are non-specific, and characteristic findings have not yet been established. Further, while NCMH is histologically benign, it is composed of immature cells that proliferate quickly, and differentiation tendencies vary by case, which can make it difficult to distinguish from malignant tumors on images.

With the exception of areas thought to represent subacute hemorrhage, areas of clear hyperintensity were not observed on DWI in the present case, and the diffusion capacity was not clearly reduced, even on an ADC map. The areas with reduced diffusion capacity were thought to represent subacute hemorrhage because hyperintensity was observed on both T1WI and T2WI (white arrows in A, C, G, and H; Fig. 2). Histologically, NCMHs are composed of island cartilage, as well as mesothelial components such as spindle cells and mucinous stroma. Despite the rapid proliferation of the tumor cells in the present case, these histological characteristics may have affected the lack of restricted diffusion.

In the present case, marginal enhancement on contrast-enhanced T1WI was thought to represent normal mucosa displaced by the tumor and tumor granulation tissue. Hyperintensity on T2WI was thought to represent cysts or mucosas, and the other areas of heterogeneous signals were thought to represent cartilage and mesothelial components such as spindle cells and mucinous stroma. However, they could not be confirmed

histologically because the tumor was removed in the form of multiple fragments.

In conclusion, regardless of the invasive proliferation tendency, the lack of restricted diffusion and the presence of bone remodeling may help differentiate NCMH from malignant nasal tumors. However, more reports on DWI and other imaging findings of NCMH cases are needed, as well as comparisons with histological findings.

References

1. McDermott MB, Ponder TB and Dehner LP: Nasal chondromesenchymal hamartoma: an upper respiratory tract analogue of the chest wall mesenchymal hamartoma. Am J Surg Pathol (1998) 22: 425–433.
2. Mason KA, Navaratnam A, Theodorakopoulou E and Chokkalingam PG: Nasal Chondromesenchymal Hamartoma (NCMH) : a systematic review of the literature with a new case report. J Otolaryngol Head Neck Surg (2015) 44: 28.
3. Finitsis S, Giavroglou C, Potsi S, Constantinidis I, Mpaltatzidis A, Rachovitsas D and Tzioufa V: Nasal chondromesenchymal hamartoma in a child. Cardiovasc Intervent Radiol (2009) 32: 593–597.
4. Yao-Lee A, Ryan M and Rajaram V: Nasal chondromesenchymal hamartoma: correlation of typical MR, CT and pathological findings. Pediatr Radiol (2011) 41: 675–677.
5. Koeller KK: Radiologic Features of Sinonasal Tumors. Head Neck Pathol (2016) 10: 1–12.
6. Tatekawa H, Shimono T, Ohsawa M, Doishita S, Sakamoto S and Miki Y: Imaging features of benign mass lesions in the nasal cavity and paranasal sinuses according to the 2017 WHO classification. Jpn J Radiol (2018) 36: 361–381.
7. Kawaguchi M, Kato H, Tomita H, Mizuta K, Aoki M, Hara A and Matsuo M: Imaging Characteristics of Malignant Sinonasal Tumors. J Clin Med (2017) 6: 116.