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Extraskeletal Mesenchymal Chondrosarcoma of Maxillary Sinus :A Case Report

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1. Abstract

Extraskeletal Mesenchymal Chondrosarcoma (ESMC) originating in the sinonasal tract has rarely been reported. Reports related to its imaging features are even rarer, which makes preoperative diagnosis difficult. Here, we analyse and report the clinical, computed tomography, and magnetic resonance imaging features of a 25-year-old male patient with pathologically confirmed ESMC in the maxillary sinus, hoping to improve the understanding and diagnosis of this disease, reduce misdiagnosis and missed diagnosis. Extraskeletal mesenchymal chondrosarcoma in the maxillary sinus demonstrate typical imaging features, such as calcification , inhomogeneous enhancement, hemorrhage, necrosis and cystic degeneration.

2. Introduction

Extraskeletal Mesenchymal Chondrosarcoma (ESMC) is a rare, highly malignant type of chondrosarcoma originating from the cartilage or chondroblastoid mesenchymal tissue [1,2]. ESMC has a high incidence of metastasis,and the adjacent vital structures are vulnerable to invasion, which leads to a poor prognosis if treated insufficiently.Histopathological examination is the gold standard for diagnosis of ESMC, but the combination of CT and MRI can accurately locate and assist in qualitative diagnosis, and judge the degree of tumor infiltration. However ESMC originate from the sinonasal tract has rarely been reported, especially its imaging features,which makes the preoperative diagnosis difficult.In this report, we focus on describing the Computed Tomography (CT), and Magnetic Resonance Imaging (MRI) features of ESMC in the maxillary sinus to improve the understanding and diagnosis of this disease, reduce misdiagnosis and missed diagnosis and to help provide more reference for the diagnosis before operation.

3. Case Presentation

We reported the case of a 25-year-old male patient who developed right nasal cavity obstruction for more than half a year without obvious incentive, accompanied by nasal itching, sneezing, runny nose, hyposmia, occasional epistaxis, no fever, headache, nasal pain, ghosting, vision loss, etc. Nasal endoscopy revealed that the right nasal cavity and inferior meatus were filled with a pink and hard neoplasm.

Images of CT revealed a soft tissue mass blocked the right maxillary sinus with multiple and diffuse calcification of varying sizes in the lesion.The bone of the upper and medial walls of the right maxillary sinus was destroyed, and there was no enlarged lymph nodes around. The average CT value was about 162HU (Figure 1A).

Images of 3.0T MRI demonstrated a tumor with equal signals on T1-weighted imaging (T1WI; Figure 1B) and mixed equal/high signals on T2-weighted imaging (T2WI; Figure 1C). On the diffusion-weighted imaging sequence (b =1000 s/mm2), limitation of diffusion was observed in the solid part of the mass and revealed slightly high signal(Figure 1D). The corresponding part revealed low signals on the apparent diffusion coefficient maps (Figure 1E). After intravenous injection of Gd-diethy lenetriamine pentaacetic acid (Gd-DTPA),the mass demonstrated obvious and inhomogeneous enhancement (Figure 1F).In the coronal, we can clearly see that the lesion grew into the right nasal cavity.The boundary with the right middle and inferior turbinate was unclear,and the lesion invaded the orbit upward, and the right inferior rectus muscle was pressed inward and upward (Figure 1F).The tumor's boundary was clear with a size of 56 mm ×48 mm × 56 mm.

Then, the patient underwent Caldwell-Luc antrostomy, and Radical antrostomy surgery. The neoplasm was pink. There were bone-derived mass and brown liquefaction necrosis substances inside. Meanwhile, we detected that the mass invaded the middle turbinate, inferior turbinate, pterygopalatine fossa, orbit, and protruded into the orbit.

Pathological examination diagnosed as extraskeletal mesenchymal chondrosarcoma. Histopathological examination revealed that the tumor cells grew in sheets, cords, and nests.The nuclei of the cells were round, oval and polymorphic.The cytoplasm of the cells was eosinophilic and partially clear. Fibrosis and cartilage tissue can be seen in the interstitium(Figure1G). Immunohistochemical analysis results were as following: CK(-),VIM(+),CD31(+),KI-67(30%+),AR(-),Desmin(-),SMA(-),S-100(-), Syn(-), CgA(-), CD117(-), P63(-), P40(-), CD99(+).

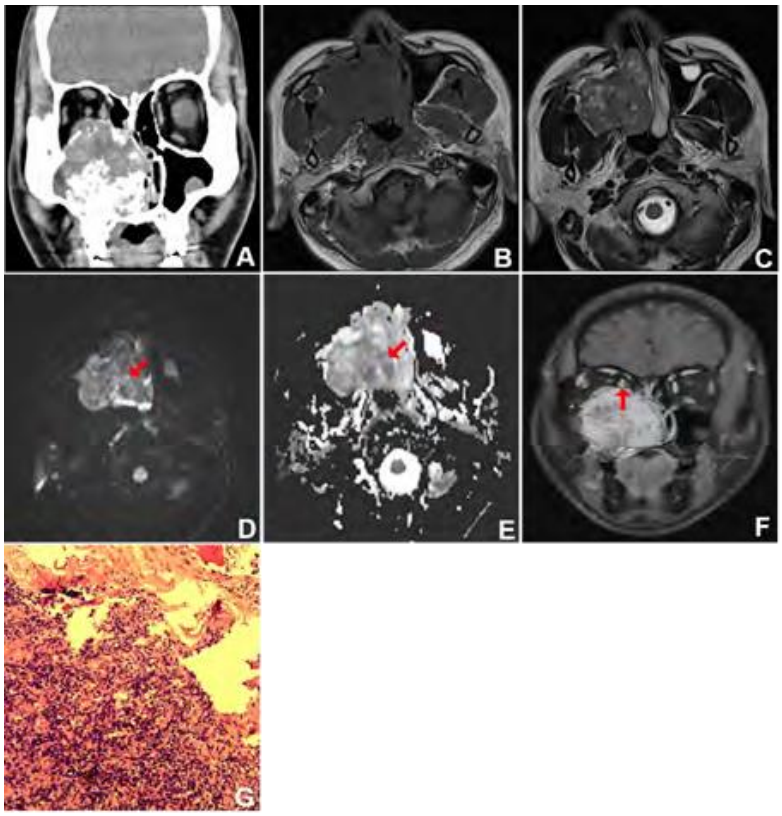

Figure 1: Radiological and pathological images from a 25-year-old male patient with extraskeletal mesenchymal chondrosarcoma(ESMC). (1A): Unenhanced computed tomography (CT) showed a soft tissue mass in the maxillary sinus. Note the multiple and diffuse calcification inside the tumor (attenuation value = 482 Hu). The bone of the upper and medial walls of the right maxillary sinus was destroyed. (1B and 1C): 3.0T magnetic resonance images demonstrated that the tumor is isointensity on T1-weighted imaging (T1WI) and mixed hyperintensity on T2-weighted imaging (T2WI). (1D and 1E): Limitation of diffusion was observed in the solid part of the tumor and revealed slightly high signal On the diffusion-weighted imaging (DWI) (b =1000 s/mm2) and low signals on the apparent diffusion coefficient (ADC). (1F): The tumor was enhanced obviously and inhomogeneously. Note the lesion grew into the right nasal cavity.The boundary with the right middle and inferior turbinate was unclear,and the lesion invaded the orbit upward, and the right inferior rectus muscle was pressed inward and upward(arrow).(1G): Histopathological examination revealed that the tumor cells grew in sheets, cords, and nests.The nuclei of the cells were round, oval and polymorphic.The cytoplasm of the cells was eosinophilic and partially clear. Fibrosis and cartilage tissue can be seen in the interstitium (hematoxylin and eosin [H&E], ×10).

4. Discussion

Chondrosarcoma is the third most common malignant bone tumor , however, the one that occurs in the extraskeletal system is even rarer, only about 1% reported in the literature [2]. The main pathological types of extraskeletal chondrosarcoma are: mesenchymal, mucinous and common. Extraskeletal mesenchymal chondrosarcoma was first reported by Dowling in 1964 [1].Extraskeletal chondrosarcoma mostly occurs in people aged 40-50 years, and the mesenchymal type is more common in young adults aged 15-40 years old, with an average age of 30 years (the age of this case is 25 years old), and it is more likely to occur in women. The majority of ESMC occurs in the soft tissue of the lower extremities, the orbit, and meninges [2].Only a few cases of ESMC originate from the nasal cavity [3] or sinonasal tract [4]. The clinical symptoms of ESMC are mostly caused by mass compression, such as progressive nasal congestion, runny nose, repeated epistaxis, pain, facial swelling or bulging eyes and so on. The tumor is often accompanied by destruction or erosion of adjacent bones, and easily invades surrounding structures [5].The growth of ESMC is slow, and the metastases appear late. The lungs are a common site of me-

tastases [6]. Therefore, for confirmed patients, chest CT examination should be performed routinely to check whether there is lung metastasis. In this case, it took up to 6 months from the onset of nasal congestion to the time of consultation and no distant metastasis was found.However, we need to closely observe and monitor the in situ recurrence and distant metastasis of the tumor.

ESMC usually manifests as a soft tissue mass on CT with calcification of different forms, which can cause surrounding bone destruction [5,7]. There are different reports on the morphology of calcification: for example,Song Chengru [8]reported that there was only a small amount of patchy calcification in the mass, and there were not annular calcification and sclerotic edge .Ghafoor S [6] found that both skeletal and extraskeletal MCS commonly presented with chondroid-type calcifications (80%). Chen M [9] reported that there was isolated punctate calcification in the tumor. In this maxillary sinus ESMC mass , there was multiple diffuse calcification of different sizes ,which was consistent with the above literature reports. Calcification is common in ESMC and it's an important imaging feature of the disease.But the calcification forms of ESMC are diverse, and there is on very specific calcification form, which suggests that it may be related to the tumor location, pathological type and degree of differentiation.

Combining literature reports [2,6,9] with this case, it can be concluded that the tumor demonstrated isointensity or slightly hypointensity on T1WI and mixed hyperintensity on fat-suppressed T2WI, depending on the amount and distribution of low T1/T2 signal calcification.On the diffusion-weighted imaging sequence (b =1000 s/mm2), the solid part of the tumor demonstrated high signal and the corresponding part revealed low signals on the apparent diffusion coefficient maps.Images after gadolinium contrast revealed obvious and inhomogeneous enhancement.Combined with the pathology, we speculate the enhanced part to be the solid component of the tumor, corresponding to the white structures seen in the gross specimen. The remaining part was speculated to be liquefaction necrosis substances and calcification.

Maxillary sinus ESMC needs to be differentiated from maxillary sinus cancer, maxillary sinus osteosarcoma, maxillary sinus ossifying fibroma, fibrous dysplasia, maxillary sinus hemangioma with calcification, and maxillary sinus fungal infection.Maxillary sinus cancer is more common in middle-aged and elderly people, and typically presents as an irregular soft tissue mass with osteolytic bone destruction.Calcification in maxillary sinus cancer is rare.When the tumor invades the bone of the sinus wall, the signal of the fat band outside the sinus wall is abnormal or interrupted, and this feature is of great significance for the differential diagnosis.Maxillary sinus osteosarcoma primarily exhibits osteolysis and/or osteoblastic destruction, as well as having an irregular tumour margin on CT imaging [10].The primary features are local or patchy high-density shadows in the medullary cavity with varying degrees of bone destruction and periosteal reaction. Ossifying Fibroma (OF) of the maxillary sinus is benign bone lesions, most frequent in young children [11].CT shows that the tumor lesions grow expansively around the medullary cavity as the center, the sinus cavity is deformed but the boundary is clear [12].The intratumoral density is uneven, and it is composed of mature dense bone tissue, immature ground-glass bone tissue, soft tissue and cystic degeneration. Therefore, it shows mixed signal on MRI. The adjacent structures are compressed but not destroyed.Fibrous Dysplasia (FD) is a skeletal developmental anomaly in which normal medullary bone is replaced by fibrous tissue [11].The lesion grows diffusely and merges with the surrounding bone, with ill-defined borders, and the involved bone expand with a ground-glass appearance. Maxillary sinus hemangioma is a vascular tumor , and a small number of cases may appear with small pieces of calcification or phleboliths. On fat-suppressed T2WI the tumou demonstrates markedly high signal (bulb sign) and on Enhanced T1-weighted image it shows heterogeneous intense enhancement. Maxillary sinus fungal balls (MSFBs) mostly occur in older individuals. According to the literature, they show a higher frequency of calcification and partial opacification with an irregular surface on CT scans compared with maxillary sinusitis and other maxillary sinus lesions [13]. There is no enhancement of the MSFBs in the enhanced scan, but the marginal mucosa can be enhanced.

Wide surgical excision is the main treatment for the ESMC of maxillary sinus,but some studies have shown that radiotherapy is an effective remedy for reducing the recurrence rate in patients whose tumor cannot be completely excised [14].Van found that the bcl-2 family and TGF β are highly expressed in ESMC, indicating that patients may benefit from bcl-2 family inhibitors and TGFβ-targeting monoclonal antibody therapy [15].

5. Conclusion

In summary, the ESMC of the maxillary sinus is a rare tumor that occurs in young people with progressive nasal obstruction and epistaxis as the main clinical symptoms. In practice, ESCM should be included in the differential diagnosis of mesenchymal-derived tumors occurring in the sinuses. Extraskeletal mesenchymal chondrosarcoma in the maxillary sinus revealed typical imaging features, such as calcification, inhomogeneous enhancement, hemorrhage, necrosis and cystic degeneration. Histopathological examination is still required for the diagnosis of maxillary sinus ESCM. But CT can better show calcification and bone destruction,while MRI can indicate the tumor's composition and invasion to surrounding soft tissue. The combination of CT and MRI can accurately locate and assist in qualitative diagnosis.

References

1. Dowling EA. Mesenchymal chondrosarcoma. J Bone Joint Surg Am. 1964; 46: 747-754.
2. Shapeero LG,Vanel D, Couanet D, Contesso G, Ackerman LV. Extraskeletal mesenchymal chondrosarcoma. Radiology. 1993; 186(3): 819-826.
3. Liu J, Guo HX, Yuan L, He ZY. Extraskeletal mesenchymal chondrosarcoma of nasal cavity: report of a case .Chinese Journal of Pathology. 2009; 38(3): 204-205.
4. Knott PD, Gannon FH, Thompson LD. Mesenchymal chondrosarcoma of the sinonasal tract: a clinicopathological study of 13 cases with a review of the literature. Laryngoscope. 2003;113(5):783-790.
5. Hanakita S, Kawai K, Shibahara J, Kawahara N, Saito N. Mesenchymal chondrosarcoma of the orbit--case report. Neurol Med Chir(Tokyo). 2012; 52(10): 747-750.
6. Ghafoor S, Hameed MR, Tap WD, Hwang S. Mesenchymal chondrosarcoma: imaging features and clinical findings. Skeletal Radiol. 2021; 50(2): 333-341.
7. Khuroo MS, Bilal S, Khuroo NS, Quyoom N. Nasal osteogenic chondrosarcoma. J Cytol. 2012;29(3):190-191.
8. Song C, Zhang Y, Cheng J. Extraskeletal Mesenchymal Chondrosarcoma in Nasal Cavity: A Rare Case Report. Ear Nose Throat J. 2021; 1455613211029797.
9. Chen M, Lai Q. Primary intra-and extradural extramedullary mesenchymal chondrosarcoma with isolated punctate calcification: case report and literature review. BMC Neurol. 2022; 22(1): 112.
10. Luo Z, Chen W, Shen X, Qin G, Yuan J, Hu B, et al. Head and neck osteosarcoma: CT and MR imaging features. Dentomaxillofac Radiol. 2020; 49(2): 20190202.
11. Rabelo N, da Silva VTG, do Espírito Santo MP, et al. Orbit ossifying fibroma - Case report and literature review. Surgical neurology international. 2020; 11: 35.
12. An XS, Li N, Cao WD, Solla D, Oberman DZ, da Costa BS, et al. Retrospective analysis on the clinical characteristics and surgical methods of ossifying fibroma of the sinuses. Chinese Archives of Otolaryngology-Head and Neck Surgery.2021; 28(12): 750-753.
13. Lee DH, Yoon TM, Lee JK, Lim SC. Computed tomography-based differential diagnosis of fungus balls in the maxillary sinus. Oral Surg Oral Med Oral Pathol Oral Radiol. 2020; 129(3): 277-281.
14. Kawaguchi S, Weiss I, Lin PP, Huh WW, Lewis VO. Radiation therapy is associated with fewer recurrences in mesenchymal chondrosarcoma. Clin Orthop Relat Res. 2014; 472(3): 856-864.
15. van Oosterwijk JG, Meijer D, van Ruler MA, van den Akker BEWM, Oosting J, Krenács T, et al. Screening for potential targets for therapy in mesenchymal, clear cell, and dedifferentiated chondrosarcoma reveals Bcl-2 family members and TGFβ as potential targets. Am J Pathol. 2013; 182(4): 1347-1356.