

Case Report

Anaplastic meningioma: a case report and literature review

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Abstract: The aim of the present case study was to investigate the imaging manifestations, histopathological and immunohistochemical characteristics of anaplastic meningioma (AM). We analyzed and summarized the characteristics of AM in a single case using computed tomography (CT) and magnetic resonance imaging (MRI), histopathological and immunohistochemical examinations. Imaging results of CT and MRI in the present case indicated bone destruction of the left parietal bone with surrounding fusiform soft tissue mass, mild edema of the adjacent brain tissue and compression of the adjacent venous sinus. On postcontrast T1-weighted MRI, the lesion exhibited marked inhomogeneous enhancement and a dural tail sign. Pathological examination identified the lesion as AM originating from the dura mater, and positive for epithelial membrane antigen and vimentin. Most AM lesions appear on brain imaging as cystic-solid masses, with an irregular shape and blurred margin, but when AM invades the adjacent bone and causes bone destruction, it is essential for radiologists to differentiate it from other extracranial tumors. CT and MRI examination are effective diagnostic methods for identifying AM preoperatively given that the uncommon imaging manifestations of AM are well understood.

Keywords: Brain, anaplastic meningioma, recurrence, magnetic resonance imaging, tomography

Introduction

Anaplastic meningioma (AM) is a highly malignant tumor classified as a grade III meningioma according to the 2016 World Health Organization (WHO) classification of tumors of the central nervous system [1]. Although AM has a high recurrence rate and an exceptionally poor prognosis, it occurs rarely, accounting for only 1%-2% of all meningiomas [2]. CT and MR examinations are usually able to diagnose typical AM cases, but when AM invades the surrounding structures and causes corresponding changes, diagnosis becomes more difficult and misdiagnosis may occur. The purpose of this case report is to report atypical or rare imaging features of AM.

Case report

In May 2016, a 27-year-old man presented to The Jingmen No. 1 People's Hospital (Jingmen, China), due to a mass on the left side of the head detected incidentally one month earlier; the patient was asymptomatic. The brain CT

scan revealed a fusiform inhomogeneous soft tissue mass with bone destruction of the left parietal bone (**Figure 1A-D**). The radiologist considered the possibility of a bone tumor, and the patient was referred for further investigation. A brain MRI revealed a space-occupying lesion with mixed intensity and a multiple cystic signal pattern, unclear margins, with mild peritumoral brain edema (**Figure 2A-C**). A magnetic resonance venogram (MRV) revealed that the adjacent superior sagittal sinus was slightly compressed (**Figure 2D**). Gadopentetate dimeglumine- (Gd-DTPA) enhanced MRI demonstrated marked inhomogeneous enhancement of the lesion, with the dural tail sign (**Figure 2E, 2F**). Those manifestations highlighted on CT and MRI led to the diagnosis of eosinophilic granuloma or plasmacytoma of the left parietal bone. Following completion of routine examinations, the patient underwent surgery. Simpson grade IV surgical resection was performed due to the poorly defined boundary between the tumor and adjacent tissue. Pathological examination revealed that the tumor was highly cel-

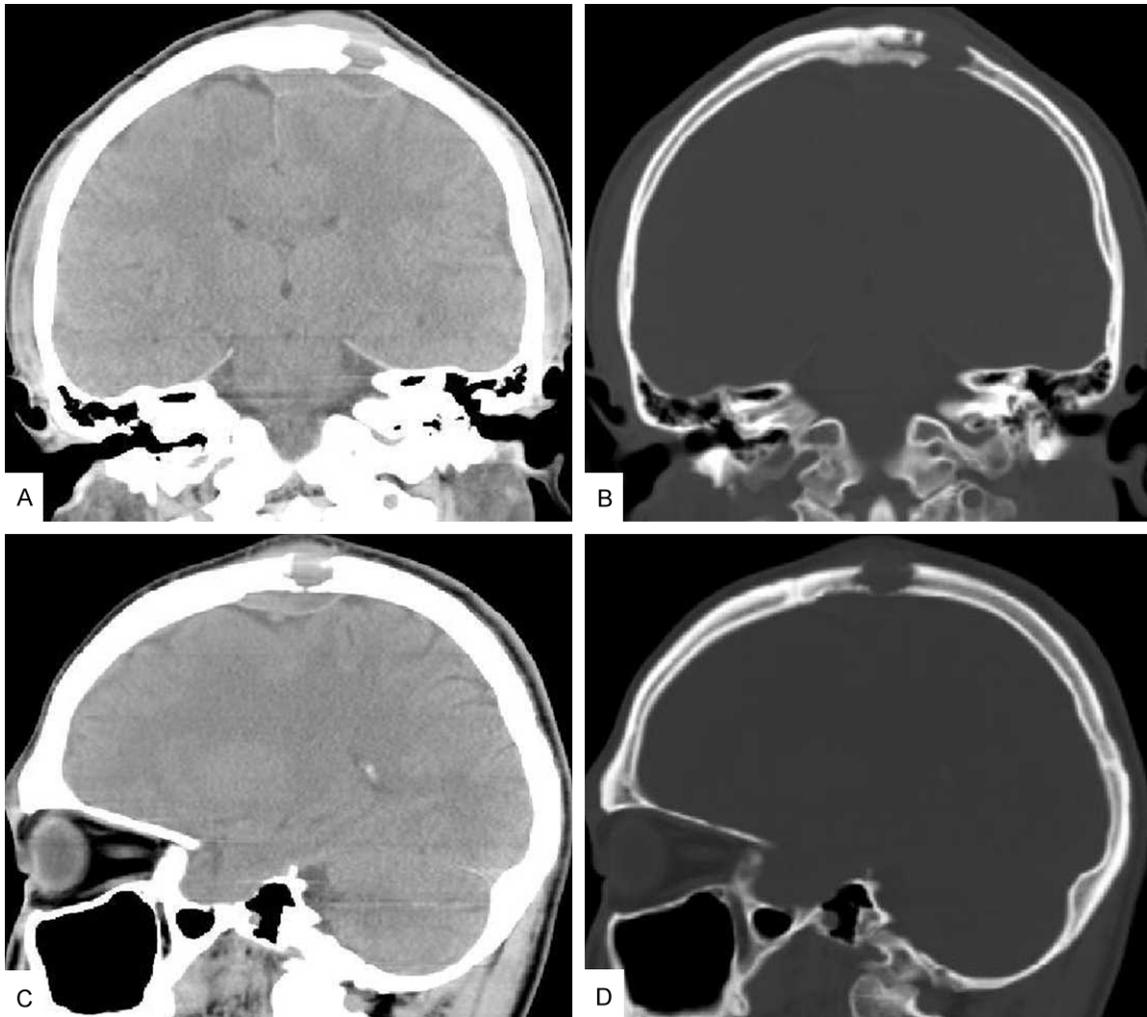


Figure 1. Brain CT showed the lesion to be located on the left top of the head, seen as a fusiform inhomogeneous soft tissue mass with osteolytic destruction of the left parietal bone.

ular, with cells exhibiting prominent and overlapping nucleoli, or multinucleated vesicular cells (**Figure 3**). Immunohistochemistry results revealed that the tumor cells were weakly positive for epithelial membrane antigen (EMA) (**Figure 4A**), diffusely positive for vimentin (**Figure 4B**), with patchy staining for progesterone receptor (PR) (**Figure 4C**), and a Ki-67 index of 20% (**Figure 4D**). Taken together, these results led to the diagnosis of AM of the left parietal region.

Following surgery, the patient received radiotherapy, with GTV 64Gy/30F and CTV 56Gy/30F five times a week. After 28 courses of radiotherapy, the first re-examination with Gd-DTPA-enhanced MRI demonstrated thickening and marked enhancement of the meninges in left

temporal area, which indicated AM recurrence (**Figure 5A**). The patient then underwent a second surgery of the left temporal meninges, and AM recurrence was confirmed (**Figure 5B**). Over the next two months, the patient continued to undergo radiotherapy and received a course of chemotherapy (temozolomide, 250 mg, once a day for five days), with unsatisfactory results. The last MRI scan in April 2017 indicated metastasis of the left parapharyngeal space involving the adjacent bone and muscle tissue (**Figure 6**). The patient then underwent seven courses of local palliative radiotherapy (GTV, 30Gy/10F), with unsatisfactory results and many complications. The patient died in September 2017. Survival time after the initial diagnosis was 15 months.

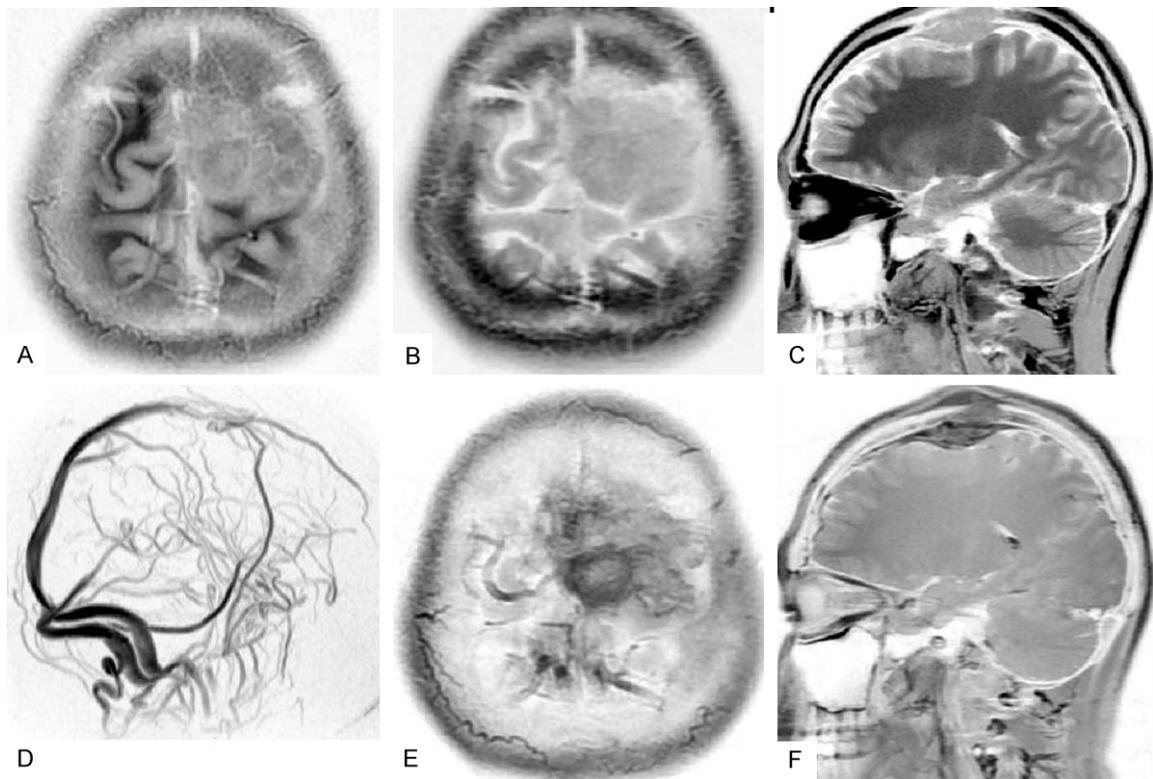


Figure 2. Brain MRI revealed that the lesion was heterogeneous with a long signal on T1-weighted images (B and C) and T2-weighted imaging (A) with small cystic longer T1-weighted imaging and T2-weighted imaging areas. MRV (D) revealed compression of the adjacent superior sagittal sinus by the lesion. Gd-DTPA-enhanced MRI (E and F) revealed marked inhomogeneous enhancement of the lesion, with the dural tail sign.

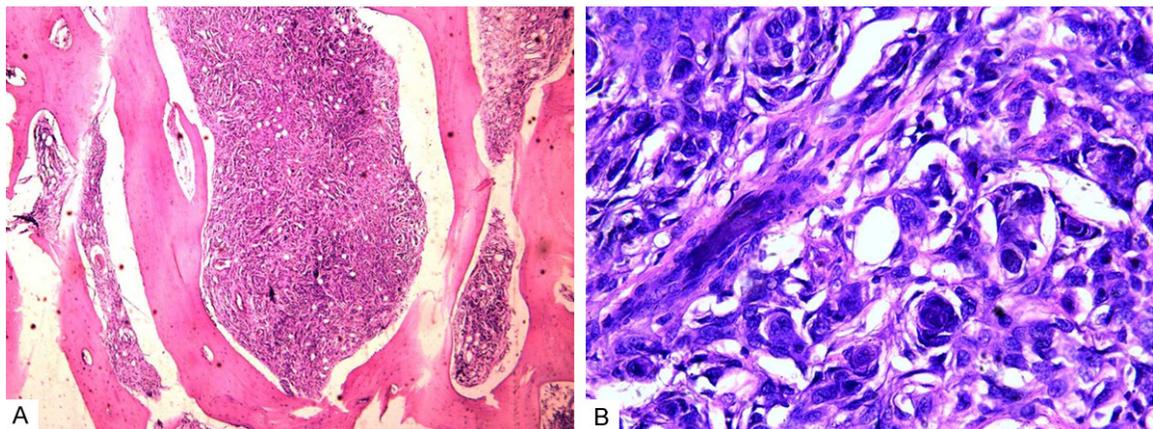


Figure 3. Hematoxylin and eosin staining revealed that the tumor was highly cellular, with large cells exhibiting prominent and overlapping nucleoli, or multinucleated vesicular cells. Magnification, $\times 40$ (A) $\times 400$ (B).

Discussion

Meningioma is the most common intracranial brain tumor, accounting for over one-third of primary brain neoplasms [3]. Meningioma is divided into 15 subtypes and 3 grades [1];

grade III has three variants, namely anaplastic, rhabdoid and papillary. AM is the most common grade III type, which has a high degree of malignancy and a high recurrence rate. The age at onset of AM ranges from 18 to 70 years old [4]. Female predominance is noted only in

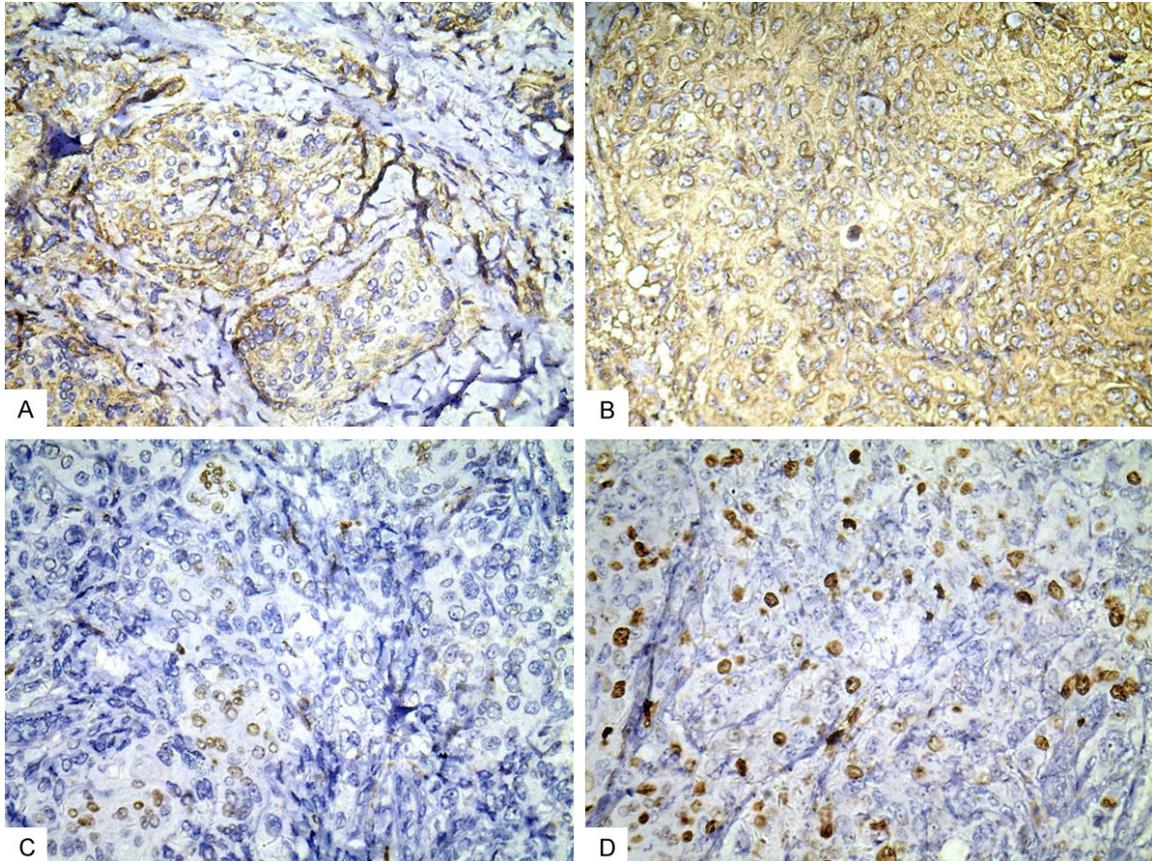


Figure 4. On immunohistochemical staining the tumor was (A) focally positive for epithelial membrane antigen and (B) diffusely positive for vimentin. (C) the staining for progesterone receptor was patchy and (D) the Ki-67 index was ~20%. Magnification, $\times 400$.

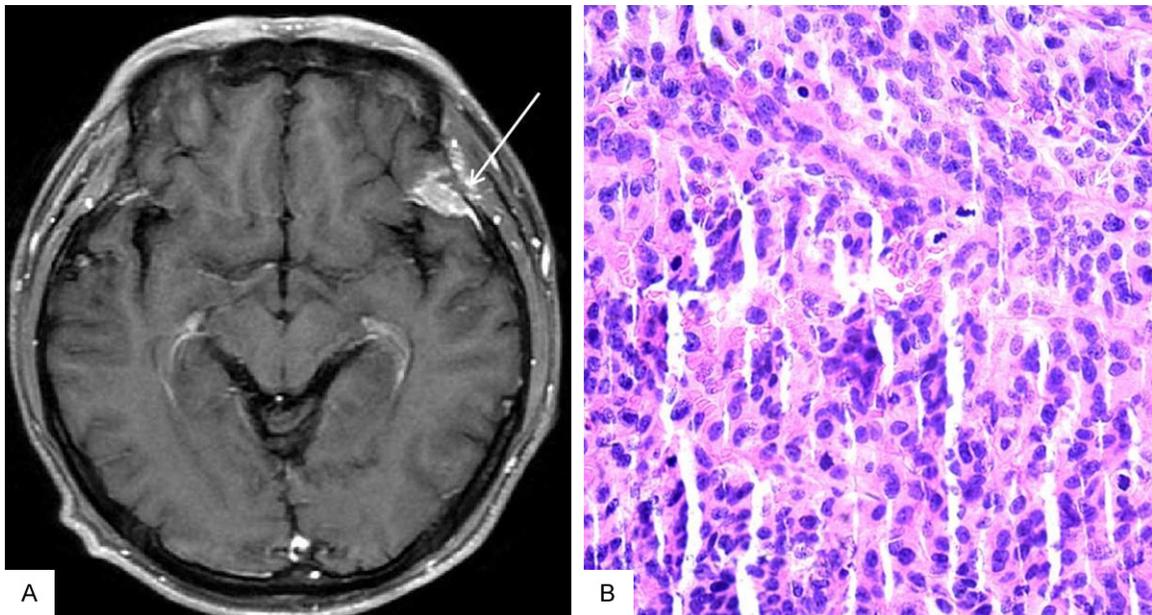


Figure 5. Postoperatively, the patient underwent the first reexamination in June 2016. (A) Gd-DTPA-enhanced MRI revealed thickening of the meninges in left temporal area with obvious enhancement (arrow). (B) After the second surgery, hematoxylin and eosin staining of the left temporal meninges revealed that the tumor was metastasis of AM. Magnification, $\times 400$.

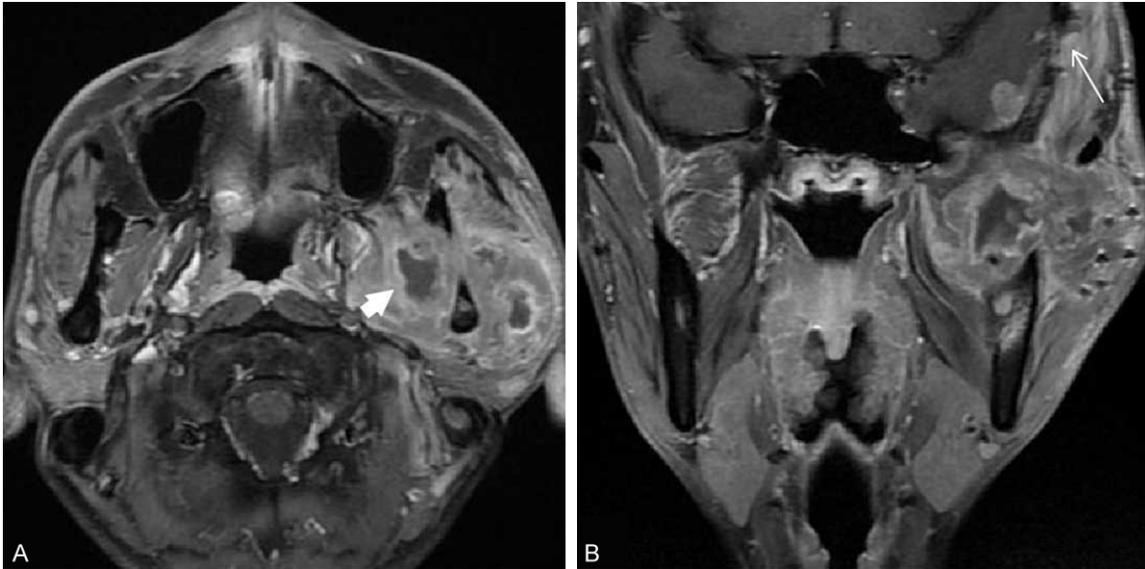


Figure 6. The last enhanced MRI scan in April 2017 indicated metastasis of the left parapharyngeal space involving the adjacent bone and muscle tissue (A, short arrow). Partial absence of left temporal bone after the second surgery (B, long arrow).

benign meningioma and male predominance is found in atypical and malignant variants [5]. The clinical characteristics of AM are not typical, most patients present with signs and symptoms attributable to mass effect at the tumor site, including headache, seizure, and hemiparesis, while some patients are asymptomatic [6]. The locations of AM include convexity dura mater, skull base, tentorium, falx, or intraventricular space [7, 8]. Variable reports of median overall survival are found in the literature, with some series reporting a survival of 1.5-3.5 years, a 5-year survival rate of 35%-61% [9-11], which may be due to variations in the times of distant metastasis. A retrospective study demonstrated that the factors associated with the progression-free survival of AM were preoperative Karnofsky performance status, extent of tumor resection, radiotherapy, tumor location and a history of meningioma [12]. Approximately 80% of patients with recurrence develop tumor regrowth and metastasis, but extracranial metastases in these cases is rare, accounting for only 0.1%. Although surgery is considered the primary treatment of choice for such cases, the high recurrence and metastasis rates require other adjuvant therapeutic modalities, such as external beam radiotherapy [6, 13]. Various targeted chemotherapies such as somatostatin analogues are actively under investigation [14].

AM may exhibit various appearances on CT and MRI but may also share some common characteristics. Most AM cases have a wide tumor base and are usually lobulated or irregular fusiform in shape as in the present case. On conventional CT scan, AM appears as a low-density shadow with areas of cystic degeneration and calcification. On conventional MRI, the tumor is iso- to hypointense on T1-weighted imaging and iso- to hyperintense on T2-weighted imaging [12]. Both CT and MRI scans demonstrated that the tumor in our case had an unclear boundary. Mild to severe peritumoral brain edema was seen, which was attributed to compression of the adjacent venous sinuses and tumor invasion of the surrounding brain tissue. On contrast-enhanced CT or MRI, the AM appeared to have significant inhomogeneity enhancement with the dural tail sign. Of note, although the dural tail sign is specific for meningioma, it is not unique to meningioma. It may also indicate invading tumor cells, hyperplastic fibrous connective tissue, and abundant and expansive blood vessels [15].

CT and MRI have specific advantages in the diagnosis of AM: CT scan is used mainly to evaluate bone invasion, and MRI to evaluate the relationship between the tumor and adjacent structures. However, when the AM causes adjacent bone destruction, as in the present case,

differentiating it from bone tumors may be difficult. It may be helpful to analyze the relationship between mass and bone destruction; the geometric center of the bone destruction and the mass are asymmetric, and this characteristic can be useful in the differential diagnosis of AM from eosinophilic granuloma and plasmacytoma. Due to the exceptionally high recurrence and metastatic rates of AM, enhanced MRI is recommended for routine postoperative check-ups.

The diagnosis of AM relies on the presence of morphological characteristics and immunohistochemical markers expressed in the tumor cells. The gross specimen of AM is greyish red. Under the microscope, the tumor either exhibits malignant cytology (carcinoma-, sarcoma-, or melanoma-like histology), or a markedly elevated mitotic index (≥ 20 mitoses per 10 high-power fields), without papillary architecture or rhabdoid cytology, and >1 per high-power field [16, 17]. On immunohistochemistry analysis, like meningiomas of all grades, patchy positive immunoreactivity for EMA will be noted in AM, and some immunopositivity for vimentin [18]. Together these characteristic results suggest that the tumor originates from the arachnoid membrane. PR tends to be negative or with a patchy positive pattern in AM compared with lower grade meningiomas [19]. S-100 is a marker for epidermal differentiation, which is mainly used for the identification of meningioma and schwannoma but is less expressed in AM [20]. Recently, somatostatin receptor 2a has emerged as a highly sensitive and specific diagnostic marker for meningiomas of all grades, and appears to be superior to EMA and PR, particularly in cases of AM [21].

In conclusion, since AM is prone to metastasis and recurrence, it represents a major challenge in terms of treatment making accurate diagnosis essential. AM exhibits the general imaging characteristics of malignant tumors, but when it causes bone destruction, observing whether symmetry exists between bone destruction and mass may provide clues to the diagnosis. AM can be accurately diagnosed through a combination of imaging and pathological findings. From a radiological point of view, imaging can help with the timely identification of metastasis following tumor resection and radiotherapy.

The patient provided signed information consent for this case report to be produced. The investigation for this case report was approved by the Medical Ethics Committee of The Jingmen No. 1 People's Hospital.

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Disclosure of conflict of interest

None.

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