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Primary Anaplastic Intraosseous Meningioma: Illustrative Case

Xiaojing Ma[1], Wenxia Yuan[1], Hengguo Li[2*]

[1]Medical Imaging Center, The First Affiliated Hospital of Jinan University, Guangzhou, China
[2]The First Affiliated Hospital of Jinan University, Guangzhou, China
Email: *lhgjnu@263.net

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Abstract

An 83-year-old woman had been diagnosed with a left frontal apex mass for more than 20 years without any symptoms. Recently, the rate of protrusion of the perceived mass increased rapidly. Malignant lesions were considered by CT/MR, not excluding metastatic lesions; malignant lesions were also considered by PET-CT, and no other tumor lesions were found in the whole body. The histopathological diagnosis was WHO grade 3 anaplastic meningioma. The patient underwent gross total resection (GTR) with no postoperative abnormalities.

Keywords

Primary Intraosseous Meningioma, Anaplastic Meningioma, Primary Epidural Meningioma

1. Introduction

Primary intraosseous meningioma (PIM) is a rare tumor originating in the skull. It is usually a slow-growing benign lesion; however, in rare cases, PIM manifests itself as a malignant tumor with high proliferative capacity, requiring maximal resection, adjuvant radiotherapy, and close follow-up. As such cases are extremely rare, they often lead to misdiagnosis in diagnostic imaging, and preoperative diagnosis is commonly made as osteosarcoma, metastasis and plasmacytoma. Here, we report a case of primary intraosseous meningioma with a histopathologic diagnosis of WHO grade 3 anaplastic meningioma. In the literature, only three cases of anaplastic PIM have been reported, so little is known about its characteristics and treatment. This patient was successfully treated with total tumor resection but was not treated with radiation therapy due to her age. We will continue to follow the patient for signs of recurrence (Table 1).

Table 1. The reported cases of primary intraosseous meningioma, WHO grade 3.

Case	Age	Sex	Symptom	Tumor location	EOR	Pathology	Time to recurrence	Publication year
1	42	Male	Facial nerve paresis	Right temporal	STR	Anaplastic	1.3 year	1993 [9]
2	70	Female	Scalp mass	Left parietal	GTR	Anaplastic	2.5 year	2006 [10]
3	78	Male	Scalp mass	Right frontoparietal	STR	Anaplastic	ID	2023 [11]
Present case	83	Male	Scalp mass	Left frontal	STR	Anaplastic	ID	ID

Abbreviations: EOR, extent of resection; GTR, gross total resection; STR, subtotal resection; ID, indetermination.

For WHO grade 3 (anaplastic) meningiomas, the recurrence interval is short, and the mortality rate is high [1]. Postoperative radiotherapy (RT) is an important consideration, and there is still controversy about the most appropriate radiotherapy after subtotal resection (STR) alone. Some patients do well for many years after undergoing STR alone, while others progress and develop larger, symptomatic tumors more rapidly. There is still no recommendation on whether patients should be observed and treated when the condition progresses, or to treat in advance [2]. Currently, prospective trials have not yet determined the most appropriate tumor target volume, radiation dose, and fractionation scheme for the patient [3].

2. Illustrative Case

2.1. Clinical Information

2.1.1. Medical History

An 83-year-old female patient discovered a mass on the left frontoparietal area about 20 years ago, measuring approximately 1 cm × 1 cm. She did not experience symptoms such as headache, dizziness, or limb weakness, and did not pay attention to it, thus receiving no treatment. Seven months ago, she noticed an accelerated growth of the mass and experienced pain upon pressure. Physical examination revealed no significant positive signs. The local hospital cranial MRI suggests that a mass on the left frontal bone and small patchy abnormal signals in the left occipitoparietal bone, which could not rule out metastatic lesions. The patient's past medical history, occupational history and family history were not specific, and there was no weight loss, and no obvious abnormalities in laboratory tests.

2.1.2. CT

Left frontal sees a class of round soft tissue mass shadow, size of about 5.9 cm × 5.9 cm × 3.7 cm. The lesion partially protruded into the intracranial; its density is not uniform, can see a large piece of low-density necrotic area and a little calcification shadow, adjacent bone can be seen destruction, did not see obvious sclerosis side (see **Figure 1**).

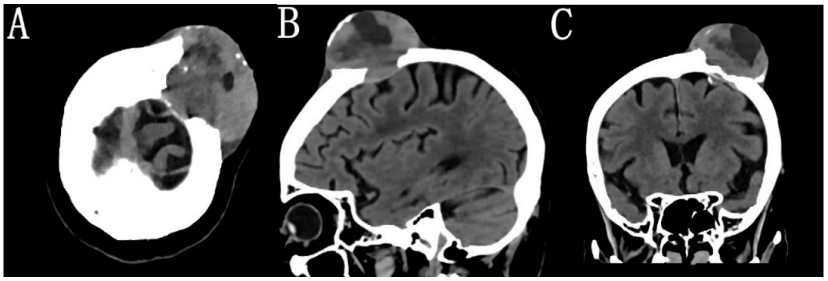

Figure 1. Head CT mass with uneven density, large patchy hypodense necrotic areas and a few calcified shadows; adjacent bone can be seen damaged, but no obvious sclerotic edges are seen.

2.1.3. MRI

The left frontal bone subcutaneous can be seen a class of round mass shadow, the boundary is clear, the signal within it is not uniform, T1WI is low signal, T2WI is uneven slightly high signal, the cystic lesion can be seen within it, local breakthrough of the frontal bone, the neighboring frontal bone can be seen obvious bone damage, the lesion and the dura mater is not clearly demarcated, enhancement of scanning lesions can be seen unevenly obvious enhancement; the diagnosis is: subcutaneous mass of the left frontal bone, considered malignant, not excluding metastasis (**Figure 2**).

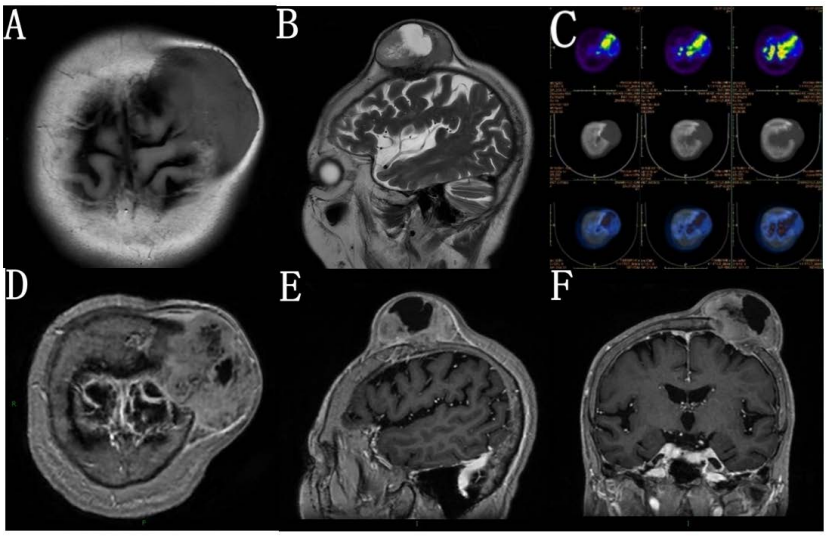

Figure 2. Cranial MR+ PET-CT TIWI (A) showed low signal, T2WI (B) showed uneven slightly high signal, within which cystic lesions were seen, local breakthrough of the frontal bone, obvious bone destruction was seen, the lesion was poorly demarcated from the dura mater, and the lesion was unevenly and obviously strengthened on enhancement scans (D E F); PET-CT (C): osteolytic bone destruction was seen in the left frontal-parietal bone, and the shadow of a round-like soft-tissue mass was seen in the local area, and multiple calcified foci were seen at the edges, FDG uptake, SUVmaxl1.3.

2.1.4. PET-CT

Left frontal parietal bone osteolytic bone destruction, localized round soft tissue mass shadow, the edge of the multiple calcification foci, FDG uptake; the

diagnosis is: left frontal parietal bone osteolytic bone destruction with soft tissue mass, glucose metabolism is increased, consider malignant lesions, and the rest of the whole body did not find other neoplastic lesions (see Figure 2).

2.1.5. Surgery Seen

The tumor was dark red and medium in texture, the tumor was separated along the bone window, the inner and anterior edges of the bone window were hyperplastic and elevated, the entire edge of the bone window was osteoporotic, and supradural vascular hyperplasia was seen, and a thin layer of the tumor was given to see the subdural and the extraarachnoid membrane, and the tumor was seen to be fat-like under the microscope, with numerous points of calcification, and the texture was soft and brittle, and it was given to be clamped off in pieces.

2.1.6. Pathologic Findings

Microscopically, tumor cells were seen to be spindle-shaped, with high cell density, lamellar, fascicular or structureless growth, and nuclear schizophrenia was readily seen, locally up to 20/10HPF, and multifocal necrosis was seen.

2.1.7. Immunohistochemistry

CK(−), EMA (foci, +), PR (−), SSTR2 (−), SMA (−), Desmin (−), S-100 (−), SOX-10 (−), CD34 (−), STAT6 (−), GFAP (−), H2K27Me3 (+), Ki67S-100 (+, localized 40%); morphology combined with immunohistochemistry, consistent with anaplastic meningioma, grade WHO 3 (see Figure 3).

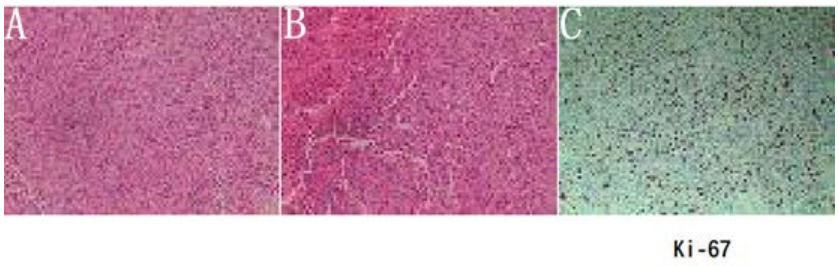

Figure 3. (A) H&E stain showing proliferation of atypical cells (×20); (B) H&E stain showing increased mitotic activity (>20 mitoses/10 high-power fields) (×40); (C) Ki-67 expression is found in ~64% of cells (×40). Original magnifications x20 (A) and x40 (B C).

3. Discussion

3.1. Observations

Meningiomas originate from the arachnoid cap cells of the arachnoid capillaries. Of these, primary epidural meningiomas (PEM) are rare lesions, accounting for less than 2% of all meningiomas, while primary intraosseous meningiomas (PIM) account for only 14% of PEMs and are more common in women [4]. Malignant meningiomas are even rarer. The age distribution of meningiomas shows a bimodal peak, with the first peak at 20 years of age and the second at 50 - 70 years of age [5]. PIM may present as osteoblastic, osteolytic, or mixed lesions, and it is

extremely rare to find PIM with both osteolytic radiologic features and mesenchymal pathology. In this case, the patient presented with a mass that invaded the subcutaneous soft tissues of the scalp and disrupted the cranial bone with significant calcification and mass necrosis associated with a malignant process.

The frontoparietal and orbital regions are the most common sites for intraosseous meningiomas. The clinical presentation of PIM depends largely on the size, location, and occupying effect of the lesion, with scalp masses being its most common clinical manifestation [6]. Cranial nerve dysfunction is usually the main complaint of skull base PIM [7]. In our case, we did not see any neurologic dysfunction, which may be related to its location in the frontal lobe rather than the skull base. Notably, the presence of a scalp mass was a strong predictor of higher grade. Also on MRI, the higher the degree of edema of the lesion, the higher the degree of malignancy [6]. Intracranial meningiomas usually present as asymptomatic, resulting in patients not taking them seriously and thus missing the best chance of treatment. In this case, the patient had a history of more than 20 years without neurological symptoms and might not have developed into a malignant meningioma if treated early.

Primary intraosseous meningiomas are usually well demarcated on CT and tend to involve the inner and outer cranial plates with typical osteophytic manifestations of focal thickening, hyperdensity and enlargement of the skull and disruption of the cortical layer [8]. However, the dural tail sign is not present in cranial PIM [7]. Osteomalacia is most commonly associated with low-grade PIM, and mixed osteomalacia or osteolysis with radiolucent bone destruction with the presence of a scalp mass seems to be more frequently associated with higher-grade PIM [6]. Primary intraosseous meningiomas are low signal at T1WI, high signal at T2WI, high signal at DWI, low apparent diffusion coefficient, and markedly enhanced on enhancement scans, but do not have specific diagnostic features.

The imaging characteristics of malignant PIM include: expansile or osteolytic changes in bone; proliferation and sclerosis of bone; formation of soft tissue and peritumoral edema; and possible bony shell or irregular calcification. The differential diagnosis of malignant PIM mainly includes metastatic tumor, cranial osteosarcoma and plasmacytoma. Metastatic tumors have an advanced age of onset, a history of primary tumor, a short course of disease, and no intra-tumoral calcification, and the typical features of MRI are focal osteolytic, osteogenic, and mixed destruction, with obvious uneven enhancement; cranial osteosarcoma, which is mostly a soft-tissue mass with irregular morphology accompanied by bone destruction, with visible tumor bone, and the tumor-induced periosteal reaction is laminar or radiating; and plasmacytoma, which has bone destruction of the cranial bone and soft-tissue filling and can for plasmacytoma, the skull bone is destroyed, the soft tissue fills in and can break through the inner and outer plates to form a biconvex soft tissue mass, there is no sclerosis of the surrounding bone, and the contour of the original bone plate inside the tumor can still be distinguished after the destruction of the skull bone, and the enhancement by MRI is

obvious and homogeneous. Metastatic tumors or sarcomas may progress more rapidly than meningiomas, but are difficult to identify preoperatively [9].

For WHO grade 3 (anaplastic) meningiomas, the recurrence interval is short, and the mortality rate is high [1]. Gross total resection (GTR) is the preferred treatment method. In addition, there is radiotherapy and chemotherapy. Regardless of whether it is after subtotal resection (STR) or gross total resection (GTR), the possibility of tumor recurrence is very high. Therefore, when GTR is not completed, postoperative radiotherapy (RT) is an important consideration. There is still controversy about the most appropriate radiotherapy after STR, some patients have done well for many years after undergoing single stereotactic radiosurgery (SRS) alone, while others have progressed and developed larger, symptomatic tumors more rapidly [2]. There are still no recommendations on whether patients should be observed and treated when the condition progresses, or to treat in advance. It still needs our research and exploration.

Conventional fractionated external beam radiotherapy (EBRT) is suitable for a broader range of patients. Hug studied a mixed group of WHO grade 2 and 3 meningiomas, determining that patients receiving ≥ 60 Gy, CGE (cobalt gray equivalent) had a 5-year local control of 100%, while a lower dose was 0%; 8-year local control was 33% and 0%, respectively. This would mean that a high dose of CGE has a better prognosis for malignant meningiomas. Prospective trials have not yet determined the most appropriate patient, tumor target volume, radiation dose, and fractionation scheme [2]. Chemotherapy is only used for unresectable, growing WHO grade 1 and all WHO grade 2 and 3 PTM [3].

In the surgical process, the unclear boundary between the tumor and the dural membrane increases the difficulty of the operation. Microsurgical techniques can be used to carefully separate the tumor from the dural membrane to reduce damage to normal brain tissue. During surgery, osteoporosis at the edge of the bone window and vascular proliferation on the dura mater were observed. Tumor separation should be performed with even greater care to avoid unnecessary bleeding and damage. The tumor contains numerous calcification points, making its texture hard and brittle, increasing the difficulty of piecemeal tumor removal. A gradual and cautious piecemeal resection strategy should be adopted to ensure complete tumor removal. Considering the anaplastic characteristics and high proliferative capacity of the tumor, a total resection should be performed to reduce the possibility of recurrence.

For follow-up, postoperative MRI imaging should be performed within 48 hours to determine the extent of resection. For benign PEM, monitoring should be conducted every 2 years. Patients with atypical histological presentations or tumors that are incompletely or completely resected with malignancy should have closer follow-up, with imaging examinations conducted every 3 months [7].

3.2. Lessons

For malignant meningiomas in the skull, total surgical resection is the preferred

treatment method. Whether the postoperative normative adjuvant chemoradiation is still unclear, and further exploration is needed. In addition, for patients with a longer history, we may need to intervene in advance to prevent malignant changes.

Conflicts of Interest

The authors declare no conflicts of interest regarding the publication of this paper.

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Abbreviations

CT	Computed tomography
MRI	Magnetic resonance imaging
PEM	Primary extradural meningioma
PIM	Primary intraosseous meningioma
WHO	World Health Organization
PET-CT	Positron emission computed tomography
H&E	Hematoxylin and eosin staining of tumor
STR	Subtotal resection
GTR	Gross total resection