

CASE STUDY

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# Primary intracerebral osteosarcoma: a rare case report and review

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## Abstract

**Background:** Primary intracranial osteosarcoma is an extremely rare disease entity. We describe a case of primary intracerebral osteosarcoma in an adult brain.

**Case description:** A patient who presented with a 1-week history of headaches, and MRI examination was performed. The immunohistochemical diagnosis confirmed primary intracerebral osteosarcoma. The patient was treated with a surgical resection of the tumor.

**Conclusion:** Primary osteosarcomas occurring in the brain are extremely rare. The MRI images did not provide a specific pretreatment diagnosis, and the histopathology was the mainstay in establishing the diagnosis.

**Keywords:** Osteosarcoma, Brain, Primary, MRI

## Background

Osteosarcoma is a highly malignant neoplasm of bone and can occur anywhere in the body, but is most commonly found in the long bones. Extraskelatal osteosarcomas occurring in tissues other than bone, are rare. Primary intracranial osteosarcomas are extremely rare, developing from leptomeningeal sheaths around blood vessels or from vessel walls themselves (Ashkan et al. 1998). Here we report a case of primary intracerebral osteosarcoma.

## Case report

A 54-year-old female was referred to our institution with a 1-week history of headaches. Her past medical history was unremarkable. Magnetic resonance imaging (MRI) revealed a large, partially necrotic and calcified tumor in the parenchyma of the left frontal lobe with marked surrounding edema and mass effect on midline structures, but without any dural attachment. The tumor appeared hypointense on non-contrast T1-weighted images (Fig. 1a) and iso-to hyperintense on T2-weighted images

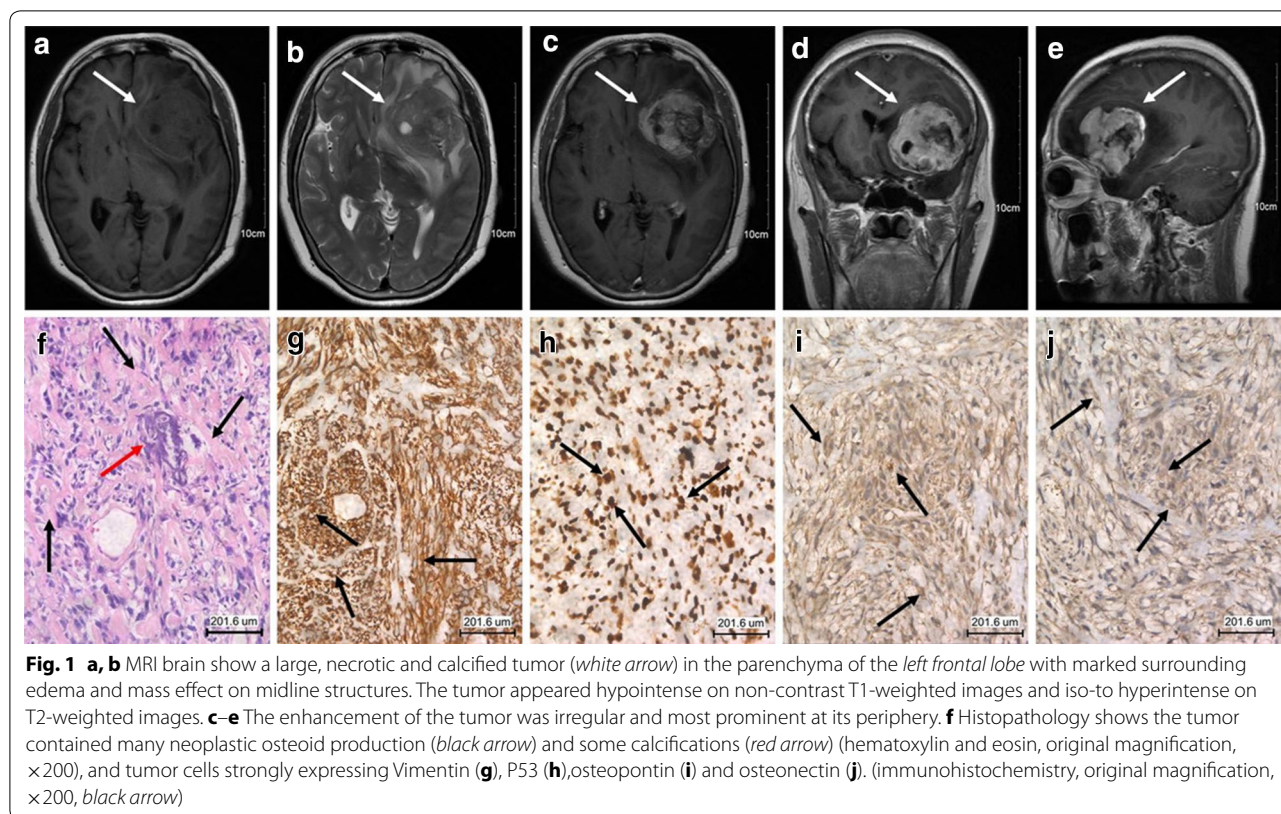
(Fig. 1b). The enhancement of the tumor was irregular and most prominent at its periphery (Fig. 1c–e). The preoperative clinical diagnosis was meningioma or calcified glial tumor. She underwent incomplete resection of the tumor and histopathologic examination revealed a malignant mesenchymal neoplasm displaying a poorly differentiated spindle cells with interspersed eosinophilic osteoid production, calcification intimately associated with the malignant cells, and localized new bone formation (Fig. 1f). Immunohistochemistry was negative for epithelial membrane antigen (EMA), glial fibrillary acidic protein (GFAP), CD34, desmin and neuron specific enolase (NSE) but positive for vimentin (Fig. 1g), P53 (Fig. 1h), osteopontin (Fig. 1i) and osteonectin (Fig. 1j). Antisera against the proliferation marker Ki-67 revealed very variable immunoreactions (80%). Given the microscopic appearance a histopathologic diagnosis of primary intracerebral osteosarcoma was made.

Extraskelatal osteosarcoma is rare and is defined as a malignant mesenchymal neoplasm that produces osteoid as well as bone or chondroid material and is located in the soft tissue without any bony attachment (Chung

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**Fig. 1** **a, b** MRI brain show a large, necrotic and calcified tumor (white arrow) in the parenchyma of the left frontal lobe with marked surrounding edema and mass effect on midline structures. The tumor appeared hypointense on non-contrast T1-weighted images and iso-to hyperintense on T2-weighted images. **c-e** The enhancement of the tumor was irregular and most prominent at its periphery. **f** Histopathology shows the tumor contained many neoplastic osteoid production (black arrow) and some calcifications (red arrow) (hematoxylin and eosin, original magnification,  $\times 200$ ), and tumor cells strongly expressing Vimentin (**g**), P53 (**h**), osteopontin (**i**) and osteonectin (**j**). (immunohistochemistry, original magnification,  $\times 200$ , black arrow)

and Enzinger 1987). Primary intracranial osteosarcoma most often represent intracranial invasion from a tumor arising from the skull (Salvati et al. 1993), and meningeal osteosarcomas which arise from the mesenchymal components of the meninges are also reported (Dagcinar et al. 2008). However, primary intracerebral osteosarcoma is rare with very few reported cases in literature.

**Literature search**

We performed a PubMed search for all cases of primary intracerebral osteosarcoma up to September 2016. Cases were analyzed for basic demographic features including

age, sex, chief complaint, location, treatment, and clinical outcome (Table 1).

**Conclusion**

Primary intracerebral osteosarcoma is an extremely rare tumor and its radiological appearance is not pathognomonic. Despite their rarity, primary intracerebral osteosarcomas should be included in the differential diagnosis, especially for other tumors such as meningioma and calcified glial tumor, and histopathology is the mainstay in establishing the diagnosis.

**Table 1 Summary of previously reported cases of primary intracerebral osteosarcoma**

Study	Age/gender	Chief complaint	Location	Treatment	Clinical outcome
Jacques et al. (1976)	53/male	Headaches and left arm weakness	Right temporo-parietal lobe	Surgery, radiotherapy	Dead at 5 months
Reznik and Lenelle (1991)	64/male	Headaches and left side hemiparesis	Right thalamus	Surgery	Dead at 3 days
Ohara et al. (1994)	57/female	Headaches	Right parietal lobe	Surgery, radiotherapy, chemotherapy	Dead at 1 year
Sipos et al. (1997)	16/female	Headaches and impaired vision	Left parietal lobe	Surgery, radiotherapy, chemotherapy	Alive at 4 years
Bauman et al. (1997)	3/female	Seizure	Right temporal lobe	Surgery, radiotherapy, chemotherapy	Alive at 18 months
Hettmer et al. (2002)	78/male	Right side weakness	Left frontal lobe	Surgery	Recurrence at 4 months

**Abbreviations**

MRI: magnetic resonance imaging; EMA: epithelial membrane antigen; GFAP: glial fibrillary acidic protein; NSE: neurone specific enolase.

**Authors' contributions**

XLZ: designed the study and drafted the manuscript. SPC and JLT: acquired, analysed and interpreted the data. All authors read and approved the final manuscript.

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**Competing interests**

The authors declare that they have no competing interests.

**Patient consent**

Signed consent is available from the patient for this report publication.

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