Intracranial osteochondroma: A case report with review of literature

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ABSTRACT

Osteochondromas account for approximately 15% of all bone neoplasms and up to 30-50% of benign bone tumors. Intracranial osteochondromas are very uncommon and represent only 0.1–0.3% of all intracranial neoplasms. We reported a 25-year old previously healthy male presented with malaise, paresthesia, and weakness of the right extremities. The patient was referred to as magnetic resonance imaging (MRI) to confirm the demyelinating disease. Instead of multiple sclerosis, an extra-axial lesion with hemorrhage was suspected, and the patient was hospitalized. Computer tomography (CT) showed a well-circumscribed extra-axial lesion with calcification. The patient had an episode of seizures accompanied by severe morning headaches, dizziness, vomiting, and speech disorders. The patient underwent craniotomy with the excision of the tumor under the control of intraoperative magnetic resonance imaging. Few episodes of Jacksonian Seizures were observed during the surgery and after the procedure. After the surgery patient remained with only slight neurological deficits. Follow-up of the patient with clinical examination and imaging studies showed no evidence of any recurrence.

Keywords: intracranial, osteochondroma, bone tumor, supratentorial

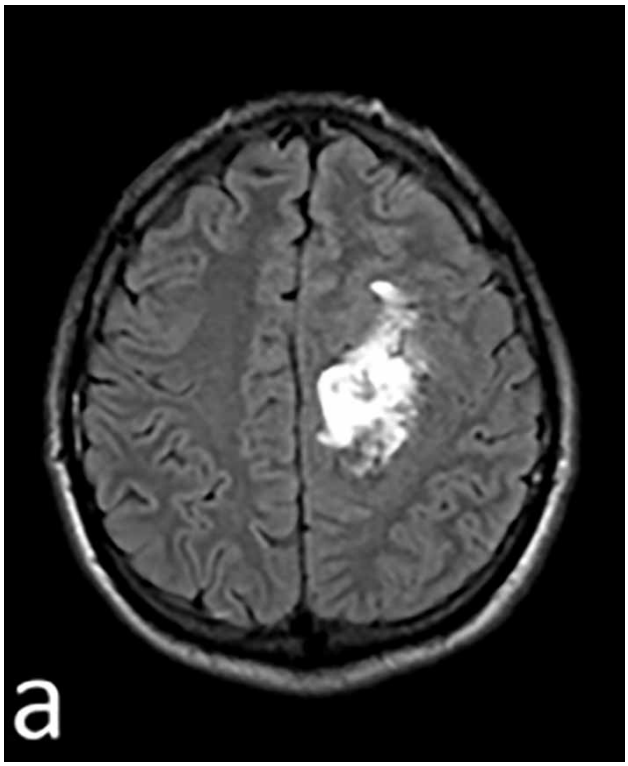
INTRODUCTION

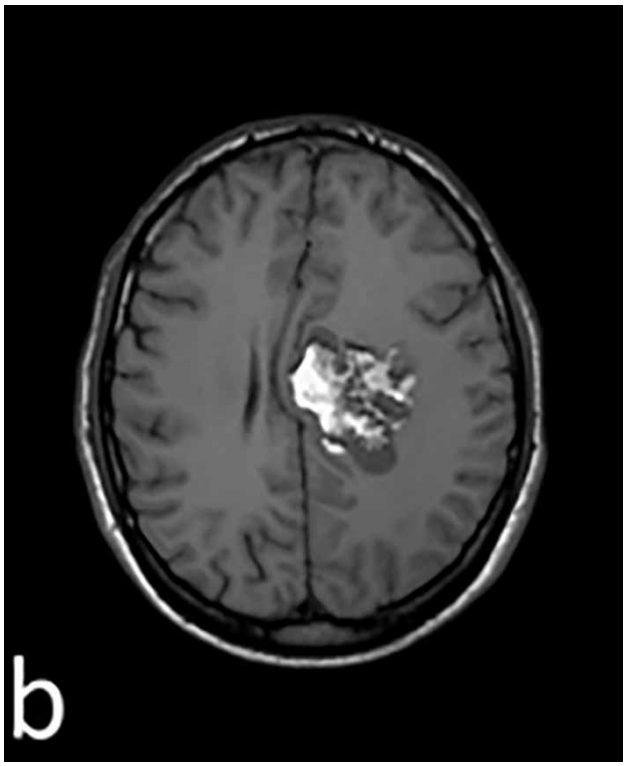
Osteochondroma is a benign neoplasm that arises from mature hyaline cartilage together with the local ossification center. The tumor itself grows slowly, although it can reach well-defined, great sizes [1]. Osteochondromas account for approximately 15% of all bone neoplasms and up to 30-50% of benign bone tumors. They usually occur around the knee or the proximal humerus [2]. Osteochondromas often occur spontaneously, although there are few reports about osteochondromas following radiotherapy [3-5]. However, intracranial osteochondromas are very uncommon and represent only 0.1–0.3% of all intracranial neoplasms [6]. So far only sporadic case reports can be found in the literature. Most of the intracranial osteochondromas arise extradurally from the base of mid-skull, although they may rarely occur from the dura mater of the falx cerebri [7, 8].

CASE REPORT

A 25-year old previously healthy male presented with malaise, paresthesia, and weakness of the right extremities. The symptoms first started half a year ago when he experienced paresthesia of the right lower extremity. The condition became extremely severe during the last week because of newly originated paresthesia of the right arm. The patient was consulted by the general practitioner and neurologist, and the demyelinating disease was suspected. He was referred to as magnetic resonance imaging (MRI) at the "Affidea Lietuva" diagnostic center to confirm multiple sclerosis. The MRI of the brain showed brain lesions with a slight midline shift and local compression features in the left frontoparietal region (Figure 1). The differentiation involved arteriovenous malformation and tumor with the subacute hemorrhage. The patient was hospitalized in the Hospital of Lithuanian University of Health Sciences Kaunas Clinics, department of neurosurgery. Physical examination revealed no pathology except for the focal neurological symptoms. Right-sided hemiparesis with muscle weakness, impaired ability to walk and instability in Romberg's pose were detected. Later on computerized tomography (CT) was performed and showed well-circumscribed extra-axial mass

lesions with intense calcification. The lesion was located in the corona radiata of the left frontal region (Figure 2). The midline structures were shifted to the right. Differential diagnosis included extra-axial tumor (calcified meningioma, metastasis, sarcoma), intralesional calcified cavernous hemangioma, maybe fibrous dysplasia. Angiography and electroencephalogram revealed no pathology (Figure 3). The neurosurgical treatment was delayed at that time because of subcutaneous infection found in the thigh. After three months, the patient had his first episode of seizures following the immediate hospitalization. On admission to hospital, the patient also complained of having severe morning holocranial headaches accompanied by dizziness, vomiting, and speech disorders. On physical examination, he was conscious, alert with stable vitals, although neurological deficits appeared to be more severe than three months ago. The patient underwent craniotomy with the excision of the tumor under the control of intraoperative magnetic resonance imaging (Figure 4). Although the patient was previously started on antiepileptic drugs, few episodes of Jacksonian seizures were observed during the surgery and after the procedure. The pathohistological diagnosis of osteochondroma was given. The patient felt well at follow-up after the operation. The improvement of symptoms was seen. However mild right-sided weakness remained. Antiepileptic treatment with phenobarbital, as well as follow-up EEG was recommended. The control MRI taken in the postoperative period of the patient revealed the total removal of the lesion (Figure 5).





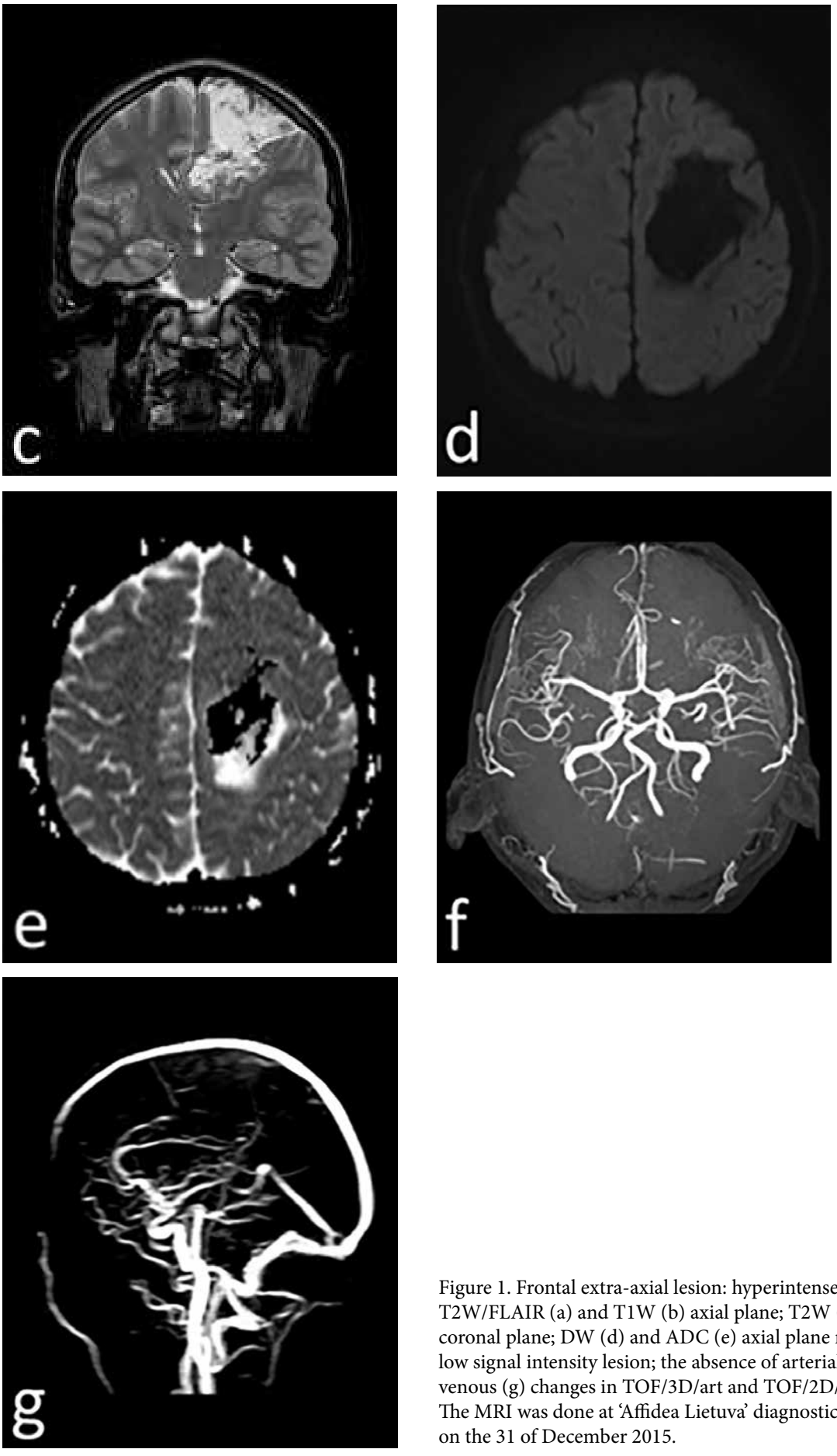


Figure 1. Frontal extra-axial lesion: hyperintense lesion T2W/FLAIR (a) and T1W (b) axial plane; T2W (c) coronal plane; DW (d) and ADC (e) axial plane revealed low signal intensity lesion; the absence of arterial (f) and venous (g) changes in TOF/3D/art and TOF/2D/vein. The MRI was done at 'Affidea Lietuva' diagnostic center on the 31 of December 2015.

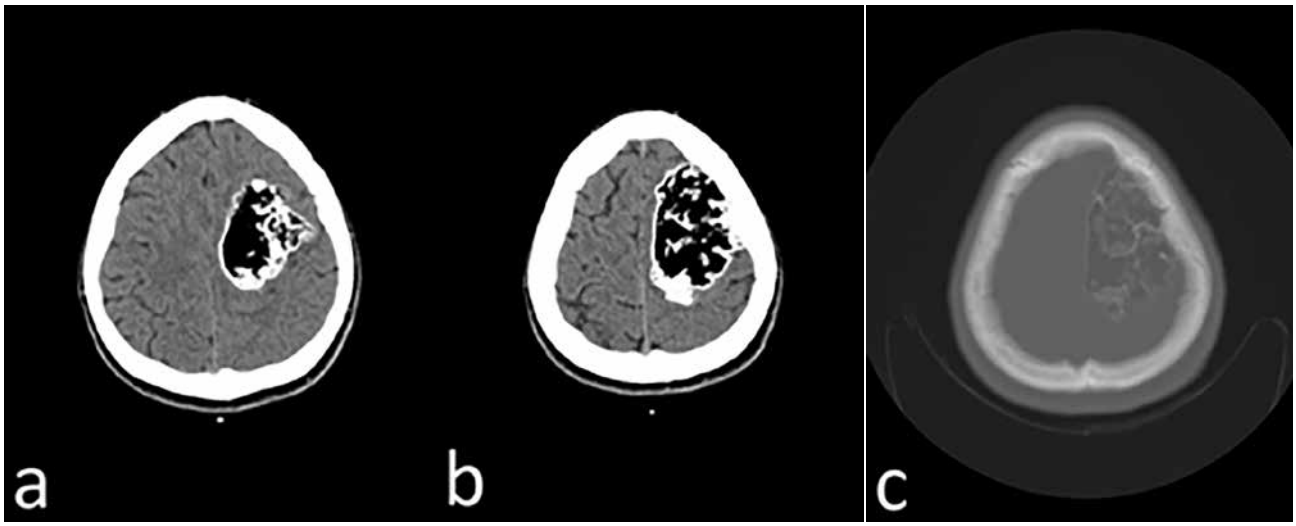


Figure 2. CT 2015 12 31 LSMUL KK: CT axial brain window (a, b), bone window (c): calcified lesion with fat inclusion are seen on the periphery.

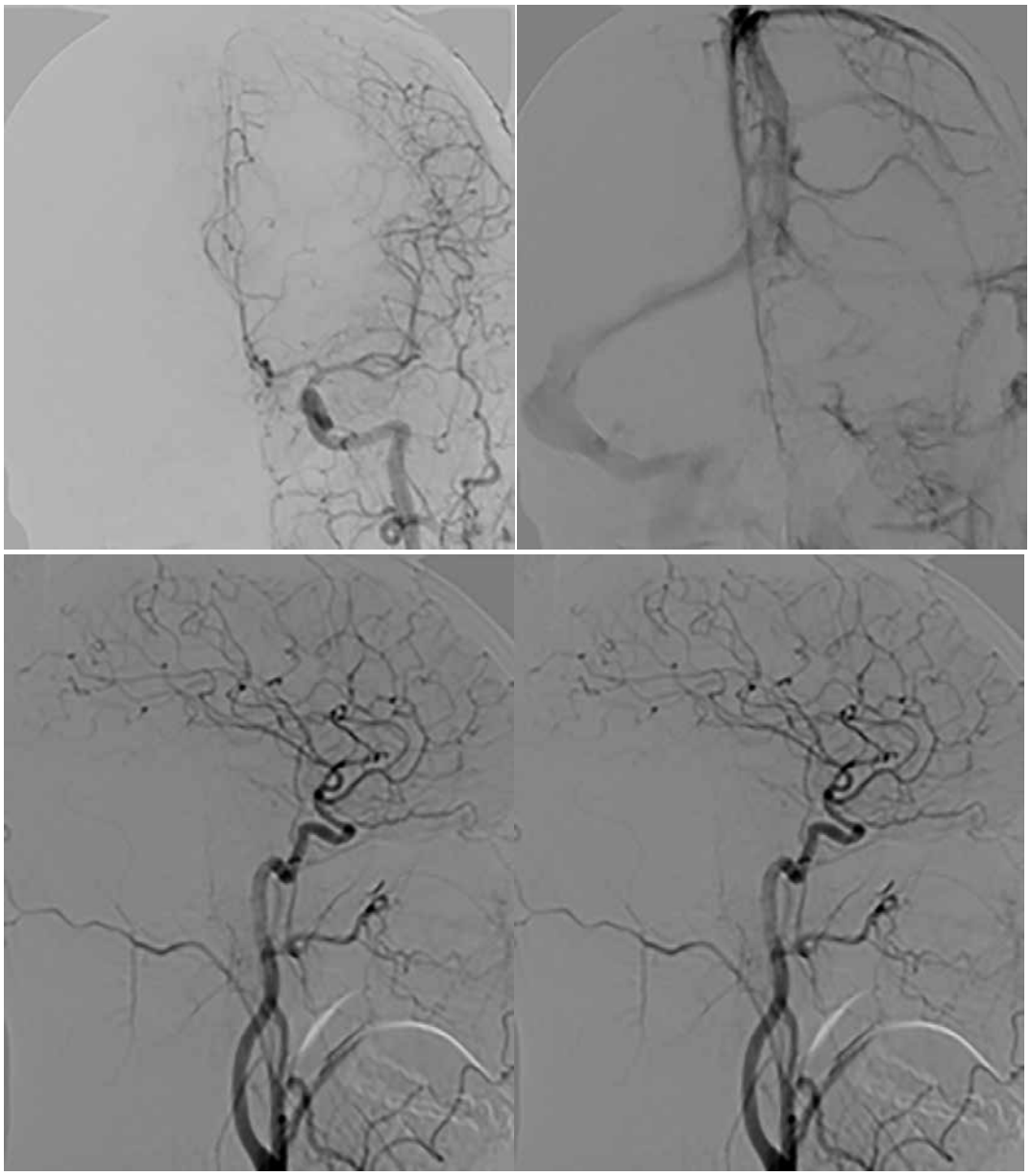


Figure 3. Anterior and lateral view of angiography revealed no pathology in both arterial and venous phases.

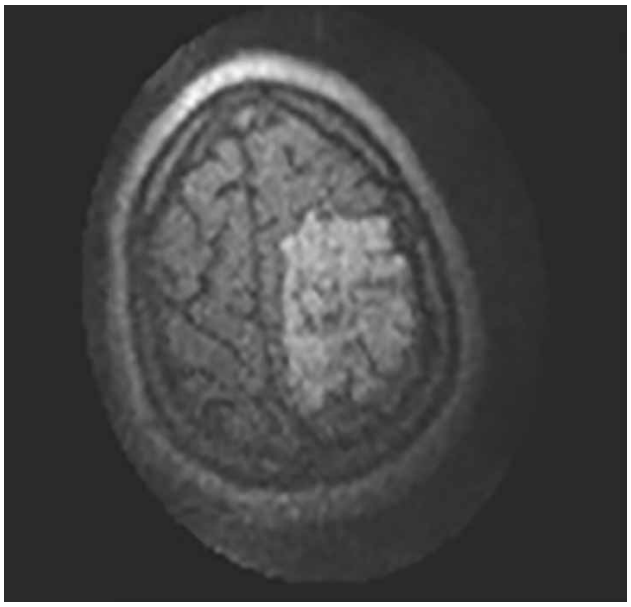


Figure 4. Hyperintense lesion was seen on the intraoperative MRI T1W axial plane.

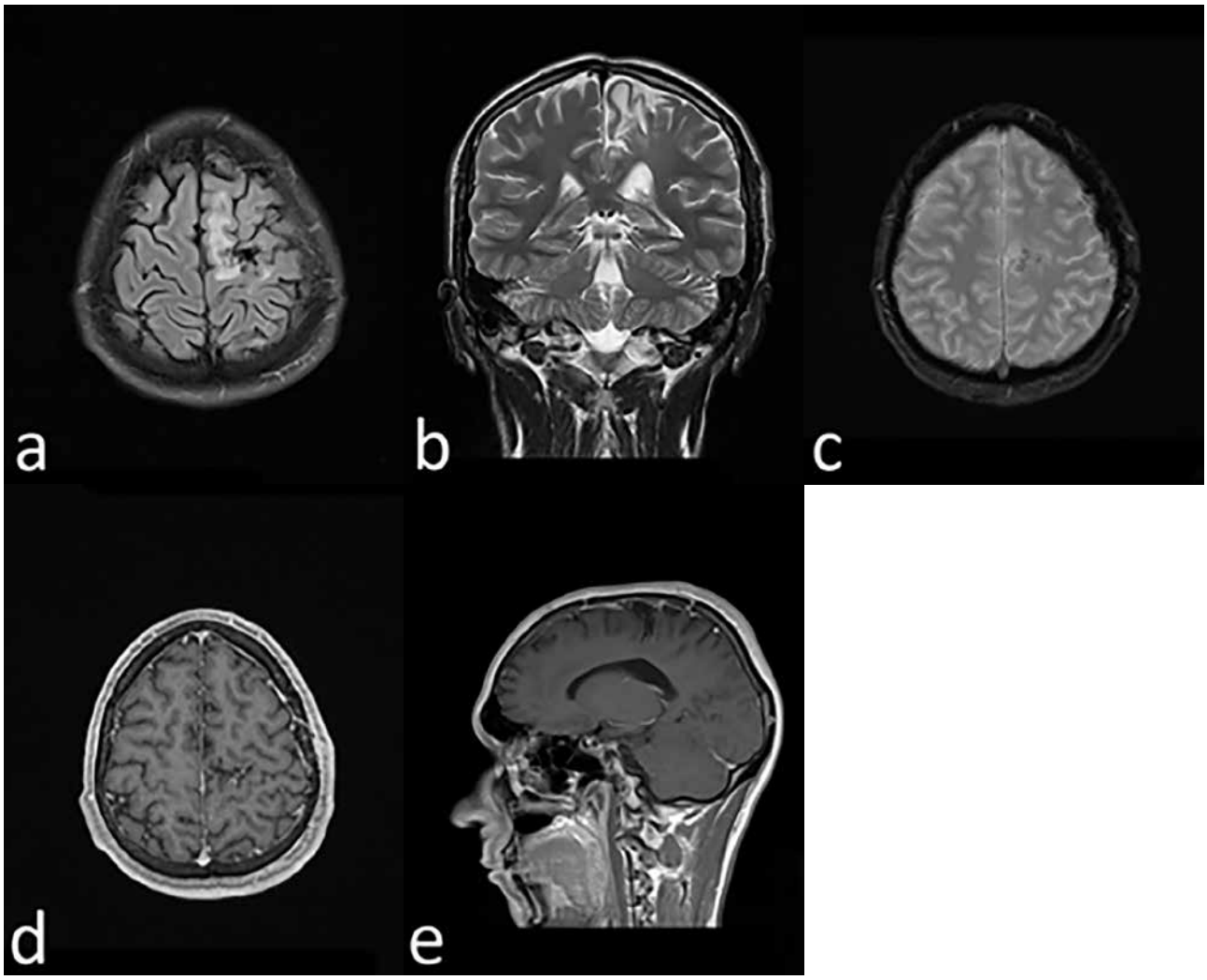


Figure 5. The postoperative control MRI (2016/2018). The postoperative cystic gliotic area at the left frontal lobe: T2W/FLAIR axial (a), T2W coronal (b), T2W/fl2d/hemo axial (c), T1W post c/m, axial, sagittal (d, e).

DISCUSSION

Osteochondromas typically occur in the metaphyseal end of skeletal bones, such as the distal femur, proximal tibia, and proximal humerus. Intracranial osteochondromas are seen uncommonly. The majority occur from the base of the skull. Only on rare occasions, they arise from the dura attached to the falx cerebri in the frontoparietal area. Therefore, the localization of the tumor makes our case unique. Intracranial osteochondromas may arise at any age with the peak incidence in the third decade, as seen in our patient [9]. Clinical manifestation of the disease mostly depends on the tumor mass effect and location [10]. The supratentorial tumor may cause a mass effect with contralateral midline shift. This may lead to rapid deterioration of the clinical condition. Meanwhile, tumors arising from the base of the skull manifest earlier then supratentorial. Because of the rare incidence of intracranial osteochondromas, the differential diagnosis with other more commonly found tumors, such as meningioma becomes essential. CT is the first choice diagnostic method to investigate neurologic signs or symptoms. This is the reason why the majority of meningiomas are detected using CT. Native CT scans usually show from slightly hyperdense to normal brain tissue, and up to 30% of cases have some calcification. Post-contrast CT reveals the homogenous enhancement of contrast material. Osteochondromas that arise from the convexity dura usually imitate meningiomas on MRI [11]. Osteochondromas appear with the central hypointensity on T1W and T2W sequences, which can be similar to calcification, with a slight chance of intratumoral hemorrhage. Calcifying or ossifying centers can be observed in meningiomas with metaplastic change. On rare occasions, meningiomas might also present with necrotic and hemorrhagic areas. The features of meningiomas include more homogenous contrast enhancement and the dural tale, unlike in osteochondromas, which show heterogeneous enhancement on contrast. More specific findings of osteochondromas are the hyperostosis and absence of surrounding edema [9]. Due to the heterogeneous appearance of the lesion, the diagnosis of the dermoid tumor should also be excluded. Most commonly dermoid tumors present with signal intensity features similar to fat. Unlike intracranial lipomas that follow fat density on all sequences, intracranial dermoid cysts have various signal characteristics on MRI. They appear as hyperintense on T1W-weighted scans and hypointense on T2W scans. Dermoid tumors and osteochondromas are similar to the fact that they both may be inhomogeneous. The heterogeneity of dermoid tumors depends on the presence of hair follicles, calcifications, and cellular debris. Rupture of the dermoid tumor can lead to fat drops in the subarachnoid spaces or ventricles, with T1W high signal intensity. Fat-fluid levels can appear in the lateral ventricles. Similarly to our case, CT scans of FD show calcified lesion. After contrast administration, enhancement is not common, and if present should be limited to the peripheral edge. Another tumor that should be ruled out before the diagnosis of osteochondroma can be confirmed is fibrous dysplasia (FD). Radiographically, FD usually has a non-symmetrical ground-glass appearance on CT scan. 20% of FD cases present with cystic lesions. FD merges with skull bone and leads to the thinning of the cortical bone [12]. Developing FD might demonstrate a cauliflower-like appearance, which is more commonly found in osteochondromas. Moreover, if the tumor invades the meninges, vasogenic edema can be found in the surrounding brain tissue. Although MRI is not the first choice in diagnosing FD, it can be sometimes used in unclear cases. T1W and T2W reveal heterogeneous signal, usually from low to moderate intensity. Contrasting gives the heterogeneous appearance to the lesion. It is also important to exclude intracranial metastasis when considering the origin of the tumor. Although, brain metastases are often multiple, up to 50% of cases demonstrate only a single lesion as seen in our case. The majority of metastatic tumors appear at the grey-white matter junction or in the arterial watershed areas. On native CT scans the mass may be isodense, hypodense or hyperdense comparing to normal brain tissue. There is a different amount of vasogenic edema found in the surrounding brain tissue. After contrast material is administrated, punctate, nodular and intensive enhancement can be observed. If

the tumor overgrows surrounding vasculature, a typical sign of ring-enhancing is seen. Meanwhile on MRI scans, metastatic tumors appear as well-circumscribed, clearly enhancing, extra-axial, dura based, heterogeneous lesions. T1W typically reveals iso- to hypointense signal intensity while T2W and/or FLAIR commonly show hyperintense lesions. Similarly to CT after contrast, an MRI enhancement pattern can be uniform, punctate, or ring-enhancing, but it is usually intense. On DWI edema is inconsistent with the size of the tumor and appears dark. While discussing treatment options, the main treatment of intracranial osteochondromas is complete surgical removal of the tumor. Although osteochondromas tend to be benign, there is a slight possibility of 1% for malignant transformation [13]. The thickness of the cartilage cap seen in MRI helps to assess the possibility of malignant transformation. Cartilage cap thickness of more than 2 cm indicates the possible malignant change [14]. Our case represents the sporadic incidence of osteochondroma. Rarely this tumor occurs as part of generalized hereditary multiple exostoses syndrome, which is an autosomal dominant condition. In our case, the tumor was completely removed, and the patient remained with only slight neurological deficits. Follow-up of the patient with clinical examination and imaging studies showed no evidence of any recurrence.

CONCLUSION

The surgical removal of osteochondromas is the gold standard of treatment. Although intracranial osteochondromas are rare, they should be involved in the differential diagnosis of intracranial extraxial neoplasms with a rare form of calcification. Despite the benign growth pattern of the tumor, the risk of recurrence should be closely monitored after the excision.

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