Advances in
Clinical and Medical Sciences
Volume 3, Issue 1

Case Report

Primary Intracranial Chondrosarcoma: A Case Report

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Abstract

Background:

Chondrosarcoma (CS) is the second most common primary bone tumor, with only 6% of cases involving skull base neoplasms and 0.15% of all intracranial tumors. CS most often develops in individuals aged 30 to 50, regardless of gender. These tumors are malignant, slow-growing, and do not metastasise until the late stages of the disease. Cartilaginous tumors occasionally arise in extraskeletal tissue, but primary intracranial cartilaginous tumors are rare. The most common subtype of chondrosarcoma is conventional chondrosarcoma, which comprises an estimated 75-80% of tumors. The main symptoms include a long-standing history of headaches and signs associated with increased intracranial pressure.

Case Report:

A 50-year-old woman visited the hospital, reporting dizziness, worsening headaches, progressive right-sided hemiparesis, and sensorimotor aphasia. Radiological findings and postoperative histopathological responses indicate the diagnosis of a gigantic primary intracranial chondrosarcoma (Grade 1). Surgery was performed to resect the tumor.

Conclusion:

Intracranial chondrosarcoma is a rare malignant tumor that can lead to serious complications if left

untreated. The main treatment is surgical resection. This clinical case describes a patient with parasagittal chondrosarcoma (Grade 1), its symptoms, diagnosis, and treatment. CT and MRI scans of the head after gross total resection do not visualize residual tumor tissue.

It is important to draw other healthcare workers' attention to the occurrence of intracranial cartilaginous tumors and the presence of non-specific symptoms. These are essential to recognize for establishing a more accurate diagnosis and planning further therapy.

Keywords: Chondrosarcoma, Brain neoplasm, Dura mater, Neurosurgery

Introduction:

Chondrosarcoma is considered the second most common primary bone tumor, with only 6% of cases representing the skull base neoplasms and 0.15% of all intracranial tumors.[1,2] CS most often develops in people aged 30 to 50, frequency regardless of gender. These tumors are malignant and slow-growing, and, until the late stage of the disease, do not metastasise. [3]

Cartilaginous tumors occasionally arise in extraskeletal tissue, but primary intracranial cartilaginous tumors are rare. [4] Tumor mostly arises from the skull base, but other localisations are reported, for example, the parasagittal falcine origin is sporadic for primary extra-skeletal intracranial chondrosarcomas. [3] The most common subtype of chondrosarcoma is conventional chondrosarcoma, comprising an estimated 75-80% of tumors. [7] The symptoms vary among patients, although the main symptoms are often associated with long-term increased intracranial pressure, such as headaches. [3,4]

The main diagnostic methods are Cranial computed tomography (CT) and magnetic resonance imaging (MRI), but only the postoperative histopathological findings can prove the diagnosis. The standard treatment is radical excision for intracranial chondrosarcoma. [5] Depending on the grade of intracranial chondrosarcoma, postoperative adjuvant radiotherapy may be used after resection to improve prognosis. [8]

This paper reviews a clinical case of a 50-year-old woman that highlights key findings, symptoms, and treatment related to a patient diagnosed with gigantic primary intracranial chondrosarcoma (Grade 1). The patient presented with a progressive headache and dizziness lasting for two years, along with right-sided hemiparesis and sensorimotor aphasia. This article reviews a clinical case of a 50-year-old woman showcasing the main findings, symptoms, and management of a patient diagnosed with a gigantic primary intracranial chondrosarcoma (Grade 1), characterised by a progressive headache and dizziness of two years' duration, right-sided hemiparesis, and sensorimotor aphasia.

Case Report:

A 50-year-old woman visited the hospital, reporting dizziness and worsening headaches that had persisted for the past two years, as well as progressive right-sided hemiparesis over the last two weeks and sensorimotor aphasia. The patient had no history of prior trauma nor any family history of congenital diseases. An initial workup included routine blood tests, a neurological examination, and a magnetic resonance imaging (MRI) scan with intravenous contrast of the patient's head. She was admitted to the Neurosurgery Department. All laboratory tests were within normal limits. A neurological examination revealed right-sided hemiparesis, while her mental status, sensory function, and reflexes remained unchanged. Muscle strength was reduced on the right side of her body.

MRI revealed an extra-axial, heterogeneous formation along the left cerebral hemisphere (AP 74mm x LL 64mm x CC 54mm) - involving the medial surfaces of the frontal and temporal lobes, as well as the middle parts of the falx cerebri, showing a heterogeneous structure with low-intensity accumulation of contrast material and a pronounced mass effect that displaces midline brain structures (Figures 1-6).

A tumor appears round-shaped in the T2 sequence, exhibiting a heterogeneous structure with hypointense lobulated and hyperintense, possibly chondroid inclusions. It shows a high ADC coefficient and a hypointense signal in the diffusion sequence, with no hemorrhages observed. In the T1 sequence, the formation signal predominantly resembles grey matter, featuring more hypointense inclusions. The accumulation of contrast material is of low intensity, mainly found in the central areas, nodular and stained, accompanied by small contrasted blood vessels. Individual tiny calcifications can be discerned in the dorsal part of the formation. Marginal contrast of the dura mater is not visible (Figure 3-5).

The contrast-enhancing component of the tumor showed no visible signal in the tumor on a diffusion-weighted imaging (DWI) sequence (Figure 6). After diffusion, the ADC coefficient and the contrast-enhanced formation do not correspond to a meningioma or a certain embryonal tumour or lymphoma. Based on the preoperative imaging results, the MRI report concluded that primary intracranial chondrosarcoma was one of the possible diagnoses, with other cartilaginous tumours, such as chondroma, possible as a differential diagnosis. Surgical resection was proposed to the patient for the evacuation of the tumour (gross total resection) via osteoplastic trepanation. The patient agreed to the surgery.

Surgery and Pathology

The patient received general anesthesia and was placed in a supine position. For the tumor excision, an osteoplastic trepanation was performed on the patient's left frontotemporal bone. The dura mater

was incised to expose the brain. A cerebrotomy along the falx cerebri was performed. On the left side of the falx cerebri, abnormal tissue was found, which was relatively hard and difficult to extirpate. A monopolar loop was used for more convenient tissue extirpation. Total tumor evacuation was performed within the limits of visibility. Hemostasis was achieved using SurgiCel Snow. The dura mater was then surgically closed, and sealed with TachoSil, the bone fragment was reinserted and fixed with tree absorbable rivets. The subcutaneous and cutaneous layers were sutured with separate sutures. After the surgery, the patient was admitted to the Intensive Care Unit (ICU) for further observation. The removed tumor tissue (2.4×1.7×1.6 cm) was sent for pathohistological examination.

Histopathological evaluation reveals a tumor with nodular growth architecture embedded in a cartilaginous matrix, consistent with Grade I chondrosarcoma (stained with Hematoxylin and Eosin, x100). Figure 7A highlights chondrocytes exhibiting bland morphology, embedded within a hyaline matrix, and separated by a minimal fibrous stroma. The cellular arrangement is sparse, exhibiting minimal cellular atypia. The absence of necrosis or mitotic figures further supports the diagnosis of a low-grade neoplasm (Figure 7A).

Figure 7B shows that tissue consisted of neoplastic chondrocytes with round, small monomorphic nuclei, mild pleomorphism, slightly increased cellularity and lobular growth (stained with H&E, ×400).

Based on morphological and immunohistochemical examination data the diagnosis of primary intracranial chondrosarcoma (Grade 1) was determined, as well as radiological data that indicated an extra-axial, heterogeneous formation along the left cerebral hemisphere with chondroid inclusions.

Outcome and Follow-up

After the surgery, the patient had right-sided hemiparesis. During the neurological evaluation, sensations were intact. After surgery, muscle strength in the lower and upper limbs was reduced, also sensory and motor aphasia persisted. The patient received rehabilitation in the postoperative period, which improved both hemiparesis and aphasia. The patient gradually regained strength and mobility on the right side and is now able to move independently without any assistive devices. Postoperative CT scan showed no residual tumor mass, a slight shift of midline structures to the right, and residual hemorrhage in the right lateral ventricle. [Figure 8]

After a follow-up visit 3 months after the surgery, CT and MRI scans showed no residual tumor within the brain. Postoperative scar tissue and hemosiderin inclusions could be seen around the surgical site. [Figures 9,10,11]

Discussion:

The World Health Organization 2020 (WHO 2020) classification divides chondrogenic bone tumors as benign, intermediate or malignant. Intracranial chondrosarcomas are divided into three subtypes based on histological classification: Grade 1 - Classical type (well-differentiated), Grade 2 - Myxoid type (intermediate) and Grade 3 - Mesenchymal (undifferentiated) type. [7] Naik et al study included a review of chondrosarcomas where 192 cases of intracranial chondrosarcoma were included, 62% of all cases were classical subtypes, while the mesenchymal type was 30%, but myxoid types only 8%. This study reports the mesenchymal type as malignant, while the classical type is the most benign. Results of the study indicate that 37% of the tumors were located in the petrous bone, 23% in the occipital bone and clivus, 20% in the sphenoid bone, and 14% occured in the frontal, ethmoid, and parietal bones. Only 6% were located in dural tissue, which typically does not contain cartilage. [5] Tumor mostly arises from the skull base, but also other localisations are reported, for example, the parasagittal and falcine origine. [3] Falcine and parasagittal chondrosarcomas have a better outcome after the radical excision and a lower risk of recurrence, compared to tumors at the skull base. [9] Hasegawa et al the study about Grade 1 and 2 skull base chondrosarcoma long-term outcomes reviewed 32 cases of intracranial chondrosarcoma, 50% of all cases were classical and 50% were myxoid subtypes. This study reports that the 10-year survival of the postoperative radiation therapy group was not significantly different from the surgery-only group and radiotherapy was more effective in cases where there was recurrence or tumor progression. [12]

On CT scans, intracranial chondrosarcoma will appear as an isodense/hyperdense lesion with varying degrees of calcification and heterogeneous structure. [8,10] MRI reveals a hypointense mass on T1-weighted images and hyperintense lesions on T2-weighted images. [11] In our case, MRI was used, which helped identify the tumour's presence and localisation. Since CS and other tumors such as chordoma have similar CT and MRI findings, it requires a biopsy to determine the final diagnosis.

On immunohistochemistry examination, chondrosarcomas mostly are S100 and vimentin-positive, they are cytokeratin (CK AE1 / AE3) and brachyury negative, while scattered proliferating cellsshow positive proliferation marker Ki-67, allowing to differentiate CS from other benign or malignant bone and other

tumors. [2] For example, cytokeratin expression helps distinguish chondrosarcoma from chordoma, but negative brachyury expression will indicate a chordoid meningioma. [13]

The main treatment method for intracranial chondrosarcoma is tumor radical excision. Studies have reported that postoperative adjuvant radiation therapy improves the outcome of patients with intracranial chondrosarcoma, especially for mesenchymal-type lesions or invasive tumors and reduces

the risk of recurrence. [1, 8, 9] According to a Ravindran et al study, recurrence within 5 years after surgical treatment varies from 5 to 39 %, but after postoperative radiotherapy from 1.5% and 42.9%. (1) In our case report, the tumor was localized to the surface of the left frontotemporal region, not invading adjacent structures and well-circumscribed from the surrounding tissue. In this case, radical excision was performed without adjuvant radiation therapy afterwards, being decided by a multidisciplinary neuro-oncology patient council.

Conclusion:

We presented a rare case of gigantic primary parasagittal chondrosarcoma. Primary intracranial chondrosarcoma is a rare malignant cartilaginous tumor. This type of tumor mostly arises from the skull base. [1,2] Differential diagnosis is often challenging due to similar imaging findings with chondroma and other tumors. [14] Pathohistological diagnosis is the gold standard and radical resection is the first choice treatment. [15] According to this rare case of low-grade, classical intracranial parasagittal chondrosarcoma, this study has provided a better understanding of the incidence and identification of this tumor for physicians managing these patients.

Acknowledgements:

The authors gratefully acknowledge the financial support for the publication of this article from the Department of Radiology at Riga Stradins University.

Figure Legends

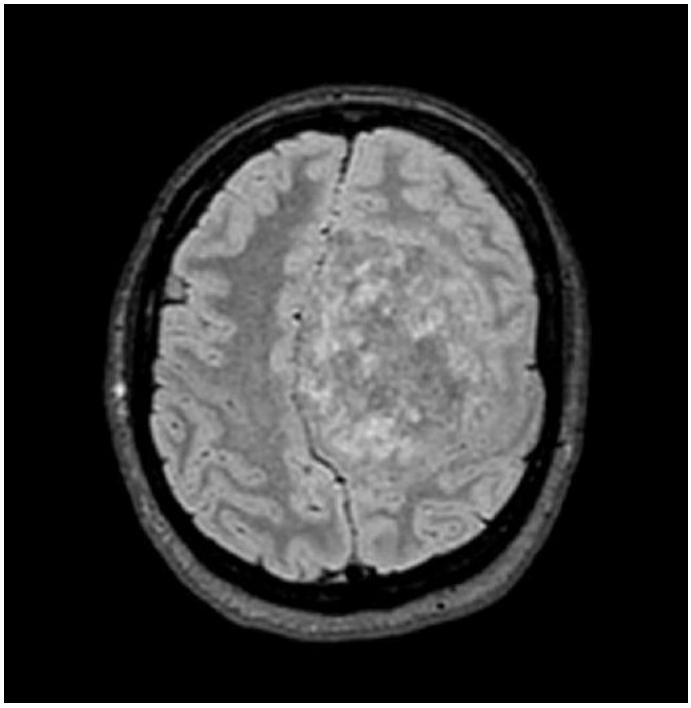

Figure 1. Axial FLAIR image. Extra axial, smoothly contoured, inhomogeneous signal intensity mass, located parasagittal to the left of the falx cerebri. Midline dislocation to the right. Left frontal, and parietal lobes are affected.

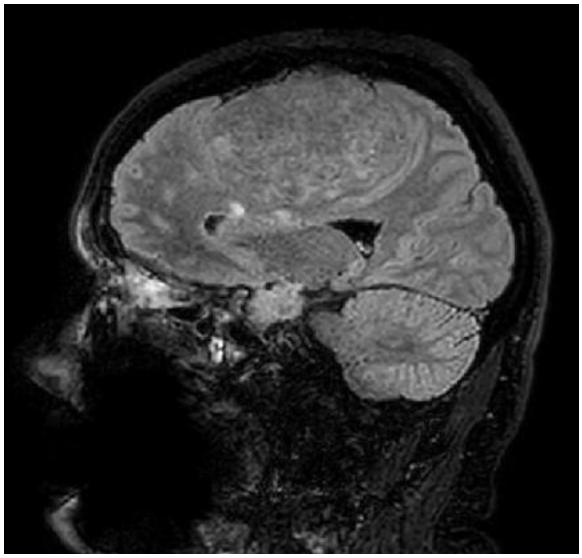

Figure 2. Sagittal FLAIR image. Extra axial, smoothly contoured, inhomogeneous signal intensity mass, located parasagittal to the left of the falx cerebri. Midline dislocation to the right. Left frontal, and parietal lobes are affected.

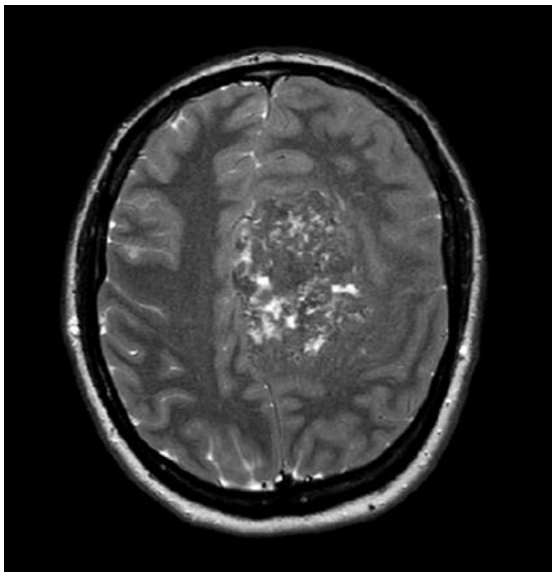

Figure 3. Axial T2 weighted image. Extra axial, smoothly contoured, inhomogeneous signal intensity mass, located parasagittal to the left of the falx cerebri). Midline dislocation to the right. Left frontal, and parietal lobes are affected.

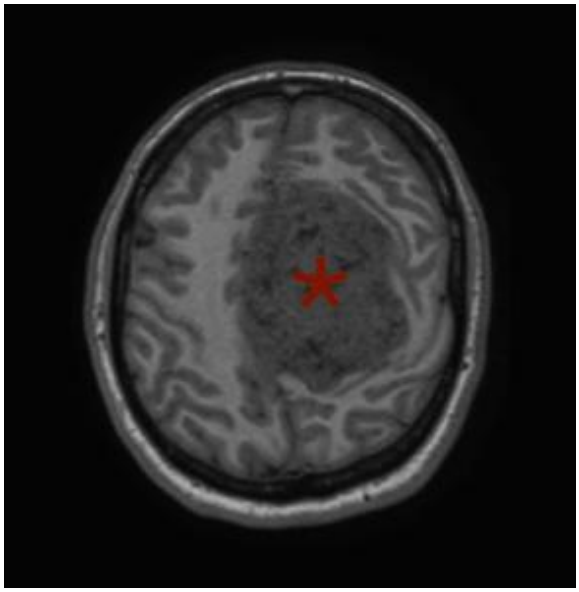

Figure 4. Axial T1 weighted image pre-Gd administration. Extra axial, smoothly contoured, inhomogeneous isointense signal intensity mass, located parasagittal to the left of the falx cerebri (marked with a red star).

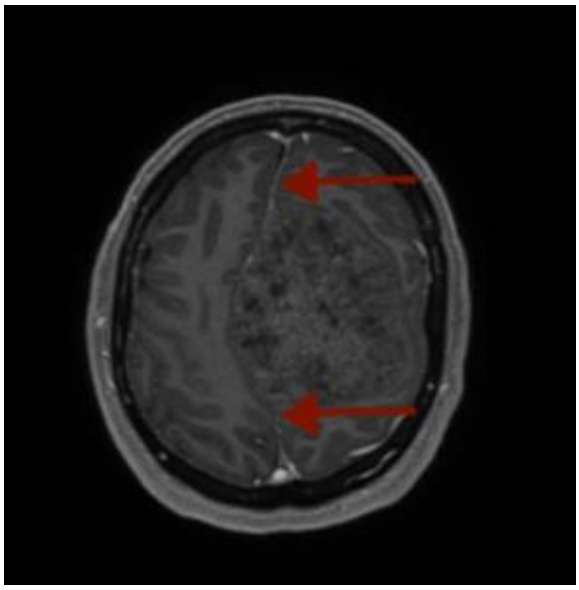

Figure 5. Axial T1 weighted image post-Gd administration. Midline dislocation to the right (shown by red arrows). Slight contrast enhancement is seen on post contrast image.

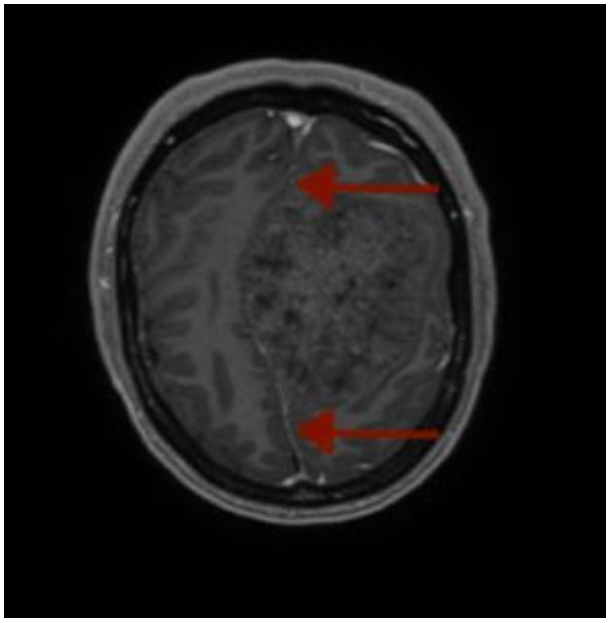

Figure 6. Axial DWI. Extra axial, smoothly contoured, mass located parasagittal to the left of the falx cerebri. No signal is visible in tumor.

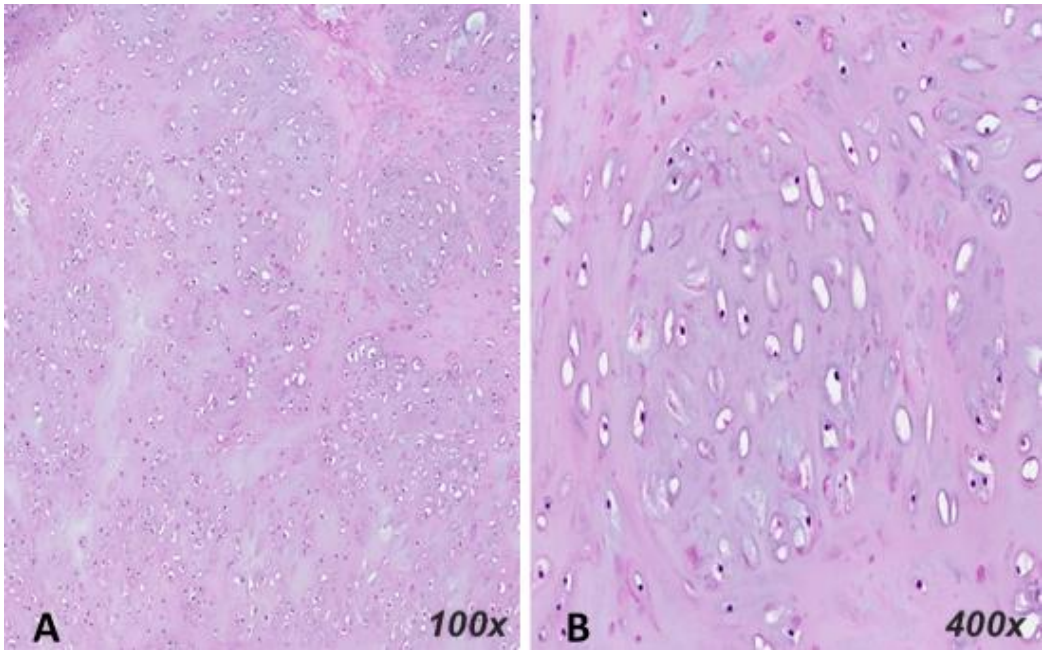

Figure 7 (A) **Figure 7 (B)**

Figure 7 (A): Microscopic evaluation at 100x magnification reveals a tumour with nodular growth architecture embedded in a cartilaginous matrix, consistent with grade I chondrosarcoma. The image highlights chondrocytes exhibiting bland morphology, embedded within a hyaline matrix, and separated by a minimal fibrous stroma. The cellular arrangement is sparse, exhibiting minimal cellular atypia. The absence of necrosis or mitotic figures further supports the diagnosis of a low-grade neoplasm. The specimen is stained with Hematoxylin and Eosin (H&E)

Figure 7 (B): At 400x magnification, the image shows mildly atypical features of chondrocytes within the cartilaginous matrix. Chondrocytes display a consistent uniformity in cell shape and size, with occasional binucleation observed. Notably, there are no significant pleomorphism, necrosis, or mitotic figures present. H&E staining.

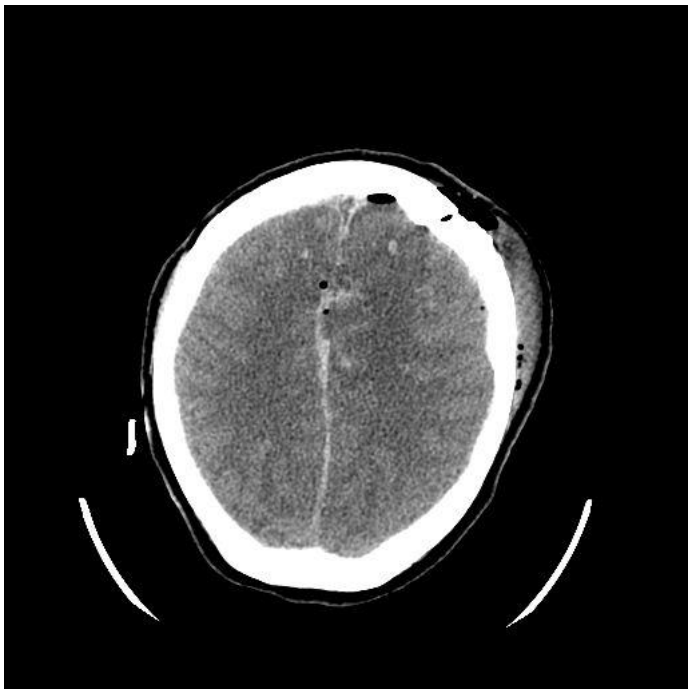

Figure 8: Postoperative CT scan showed no residual tumor mass, and a slight shift of midline structures to the right.

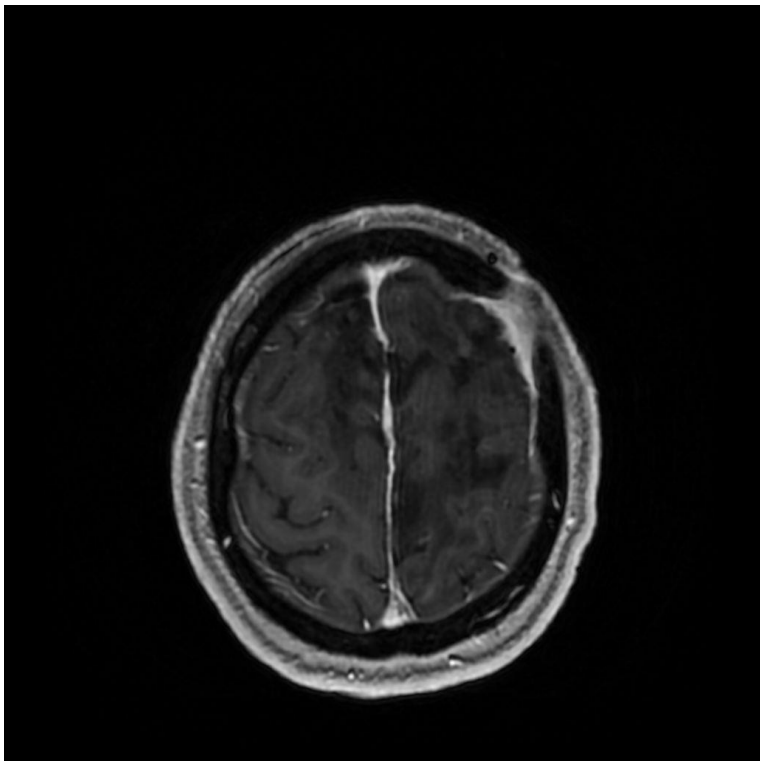

Figure 9: Postoperative MRI scan. Axial T1 weighted image post-Gd administration. Data on tumor residual tissues are not obtained. Post-therapeutic gliosis can be visualized, as well as small remnants of haemorrhages in the post-operative areas.

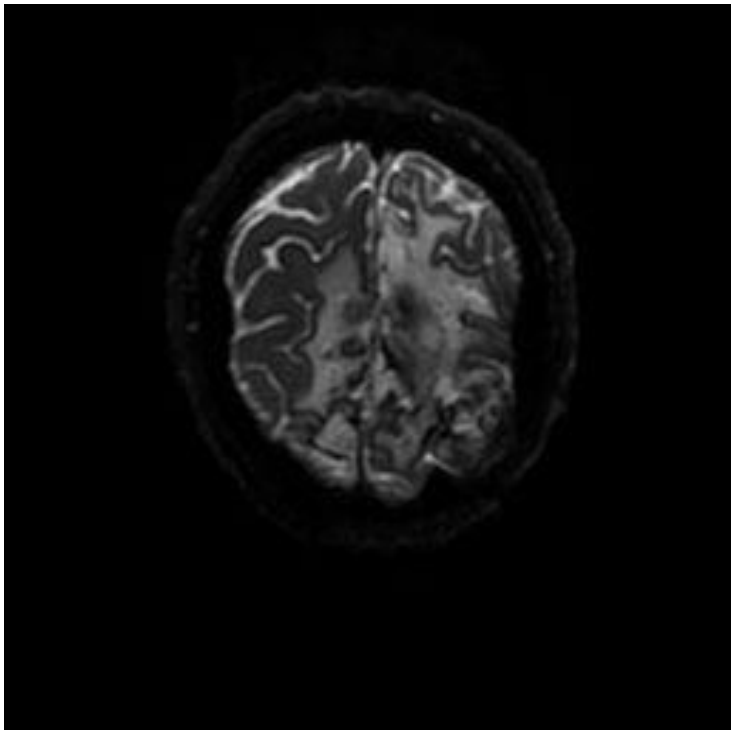

Figure 10: Postoperative MRI scan. Axial DWI does not visualize residual tumor tissue.

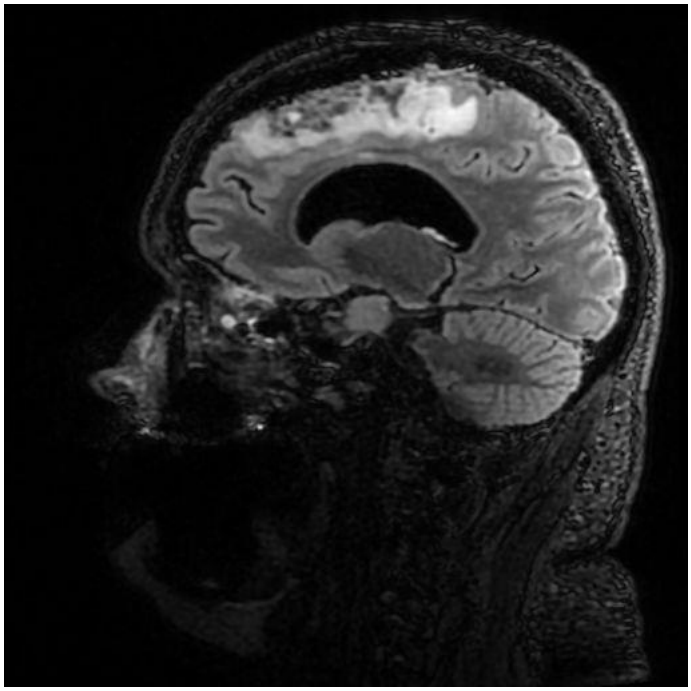

Figure 11. Sagittal FLAIR image. Post-therapeutic gliosis and small remnants of haemorrhages in the post-operative areas can be visualised. No residual tumor tissue is visualized.

Conflicts of Interest/Disclosures

The authors declare that they have no financial or other conflicts of interest in relation to this research and its publication.

Informed consent

The authors declare that written informed consent for publication was obtained from the patient.

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Citation: Elina Ozolina, Aija Tumova, Māris Buks, Ardis Platkājis, Arvīds Jakovļevs, Kārlis Bicāns, Kaspars Auslands, Adv Clin Med Sci, "Primary intracranial chondrosarcoma: A Case Report". 2025; 3(1): XXXX

Received Date: December 30, 2024; Published Date: January 06, 2025