Chondrosarcoma of the petrous bone: a challenging clinical entity

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Abstract  
Skull base chondrosarcomas are challenging lesions to treat. In this manuscript, we present a brief overview about this clinical entity. Furthermore, we present an illustrated case with chondrosarcoma of the petrous bone and highlight microsurgical treatment options.

Introduction  
Skull base tumours are challenging, and neurosurgeons and ENT specialists probably find these lesions the most difficult to treat. During the last decade, many novel techniques such as endoscopy have been introduced in our clinical armamentarium. This manuscript focuses on the chondrosarcomas of the petrous bone and presents an illustrative case, giving a current overview of the natural history, diagnosis and multimodal treatment strategies.

Epidemiology  
Intracranial chondrosarcomas are most common skull base lesions, whereby extra-skeletal occurrence including falce, parasagittal, parasellar and in the choroid plexus have also described1–5. These tumours arise usually de novo; however, an association with Maffucci syndrome, Olliers and Paget's diseases have also been described in the literature6–8.  
The incidence of primary intracranial chondrosarcomas is estimated to be <0.16% of all intracranial tumours and 6% of all skull base lesions3–5,7.

The origin of skeletal chondrosarcomas is uncertain. Some authors postulate derivation from undifferentiated cells from cartilaginous synchronous chondrosarcomas3–11. Extra-skeletal chondrosarcomas are believed to arise from pluripotent cells of the meninges, parenchyma and choroid plexus3,6.

Histological subtypes  
Histologically, three subtypes are differentiated: classic, myxoid and mesenchymal. Each subtype is associated with different incidence and prognosis. Microscopically, mesenchymal chondrosarcomas appear with a densely cellular stroma of anaplastic cells with associated lacunae of neoplastic chondrocytes and hyaline cartilage. Further characteristic findings of chondrosarcomas are collagen and cytoplasmic glycogen deposits. The mesenchymal subtype was found to be a more malignant type with a higher tendency for recurrence, metastasis and increased vascularity. The myxoid type is characterized by streaks of chondrocytes in a myxoid matrix and has an intermediate histological position. The classic type is well differentiated with minor mesenchymal tissue3,5,6,9. The mesenchymal type occurs more frequently in younger (20–30 years) and the classical subtype in elderly patients (60–70 years)3,4,12. However, chondrosarcomas can occur at any age. A literature review by Korten and co-workers revealed occurrence of these tumours in patients aged 3 months to 76 years4.

Tumour grading is determined by the classification of the World Health Organization (WHO) into three grades. Grade I tumours are well differentiated and grade II tumours are moderately differentiated. Grade III lesions, however, are poorly differentiated and are highly malignant.

Chondrosarcomas tend to grow rapidly with consecutive compression and dislocation of the surrounding brain structures. Furthermore, these tumours are characterized by their infiltrative behaviour, which lead to progressive neurological complaints such as headache and impaired vision6,12.  

Neuroimaging  
Characteristic neuroimaging findings of cartilaginous tumours—more specifically chondrosarcomas—are calcifications in varying degrees. On computed tomography (CT) scans, these tumours appear as isodense to hyperdense lesions, with heterogeneous enhancement3. Even if distinct calcification is missing within the tumour, T1-weighted images show low signal intensity, which can be construed as cartilage lacunae.

The classic type chondrosarcoma is a slow growing tumour. According to this biological behaviour, these tumours appear as extra-axial lesions with good delimitation to the surrounding brain tissue with retained cerebrospinal fluid (CSF) margin, even in infiltrative tumours. This circumstance is best shown on T2-weighted magnetic resonance imaging (MRI) sequences, in that these tumours appear strongly hyper-intense. On the other hand, chondrosarcomas appear hypointense on T1-weighted images. The affection of the bony structures such as "pressure erosion" is best shown on CT scanning1,11. Since chondrosarcomas are usually low or avascular tumours, application of contrast agents may lead to mild enhancement. Some authors describe the "honeycomb" pattern3,14. However, some tumours can present with a high vascularization pattern and may mimic vascular tumours.

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such as hemangiopericitomas. Despite the typical neuroimaging findings of chondrosarcomas, meningiomas and metastases have to be differentiated.

Illustrative case

A 26-years-old male patient presented with left sided abducens nerve palsy, atrophy of the left half of the tongue and progressive headaches since three months. On examination, the patient additionally presented uvula deviation to the left side and dysphagia without hearing impairments. Neuroimaging with cranial MRI and CT showed a tumour of the petrous apex, which extended to the posterior fossa right up to the foramen magnum (Figure 1).

After evaluation of the images, it was clear to us that the tumour had to be treated by combination of two approaches. The first surgical approach was retrosigmoid suboccipital to remove the tumour of the posterior fossa. The first reason for performing this surgery was the clinical symptomatology of the patient, i.e. double vision related to the abducens nerve palsy. The second delayed approach was subtemporal extradural with the necessity of an additional petrous apicectomy to remove the tumour adjacent to the petrous bone to obtain gross tumour resection. Prior to surgery, a lumbar drainage was placed to reduce CSF and enable slight elevation of the temporal lobe. During this surgical intervention, a navigation system with previously obtained CT data was used.

Figure 2 shows the cranial CT images after the retrosigmoidal and Figure 3 shows the post-operative CT after the subtemporal approach. The tumour is with exception of a small part in the petrous bone completely removed. In the post-operative course, the abducens palsy declined completely.

Histologically, the tumour was diagnosed as a chondrosarcoma grade II with extended osteodestructive growth and focally myxoid components. The proliferation index was less than 1% (Figure 4).

Since complete tumour resection was not possible, a heavy ion radiation therapy was performed post-operatively. The patient did not suffer any additional neurological deterioration after treatment without proven tumour growth in the follow-up examinations (follow-up of 16 months).

Surgical approaches and anatomical structures

There are different types of approaches to treat chondrosarcomas in the middle cranial fossa, depending on the extent of the tumours and necessity to protect cranial nerves, especially the cochlear nerve. Therefore, the most suitable surgical strategy needs to be determined on a case-by-case basis, considering the neuroimaging findings carefully. To obtain the best surgical result, neurosurgeons have to be familiar with the anatomical structures of the skull base. Subsequently, the most important anatomical landmarks are recapitulated to make surgical understanding easier. The following descriptions are based on the findings of Waniibuchi, Friedman and Fukushima as well as on the illustrations of Sanna, Sekhar and Yaşargil. The two main approaches to tumours of the petrous bone are subtemporal and retrosigmoidal. By application of each approach, different parts and angles of the area of interest are visible.

Retrosigmoidal approach

The overview of anatomical structures through a right-sided retrosigmoidal approach is displayed in Figure 5. From this point of view, looking over the cerebellum, the relationship between the trochlear nerve and superior cerebellar artery (SCA) as well as the relationship between the SCA and trigeminal nerve is visible. The fifth cranial nerve impresses as a really strong nerve close to the SCA. Next to the trigeminal nerve, the so-called acousticofacial bundle (AFB) can be identified, which contains the facial nerve, acoustic nerve, as well as the...
Critical review

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Figure 2: Tumour removal through a retrosigmoidal approach. (a) The tumour was partially removed by a retrosigmoidal approach. The posterior part of the petrosal bone was drilled to remove the tumour tissue (arrow), and the tumour was removed from the caudal cranial nerves, the abducens and cranial nerves VII/VIII. (b) The carotid artery can be visualized in its petrous part (arrow). (c) The sigmoid sinus entering the jugular foramen is shown with an arrow. ICA is shown in (b) and (c) in close proximity to the tumour. (d) In the bone window of the CCT part of the petrous bone is drilled (white arrow). (e) The bottom part of the tumour is still present as it is seen in contrast-enhanced MRI of the brain. (f) In this MRI image, the limits of tumour removal are shown by white arrows. The green arrow indicates cranial nerves VII and VIII, the blue arrow shows the labyrinth and the red arrow shows the cochlea, which could be left intact during surgery. (g) MRI shows the remaining tumour, which was then removed with a subtemporal approach combined with a petrosal apicectomy (white arrows).

dissection of the dura is done by following the MMA to the foramen spinosum. Rostral to the foramen spinosum, the foramen ovale appears with the third branch of the trigeminal nerve (V3, maxillary nerve). Caudal to the foramen ovale and spinosum, the petrous part of internal carotid artery (ICA) is visible, which is usually covered by a thin bone layer. The greater petrosal nerve (GPN) runs medially to the ICA, which can be followed to the arcuate eminence as another anatomical landmark, which indicates the internal auditory meatus. After dissection of all anatomical landmarks, the petrous apex is visible and petrous apicectomy can be performed.

For drilling, the knowledge of the course of the petrous part of ICA and GPN is important. Mortini and co-workers defined the pentagonal post-trigeminal area, which is marked out by ganglion gasseri, ICA, cochlea, internal auditory canal and superior petrosal sinus. The authors termed the angle between the ganglion gasseri and superior petrosal sinus as the key point at which drilling should start to avoid damage to the surrounding anatomical structures. The anatomical structures are displayed in Figure 6.

Subtemporal extradural approach

The subtemporal extradural approach, which is used in our illustrated case, is a widely used approach for extradural lesions. Preoperative lumbar CSF drainage provides better retraction of the temporal lobe and better exposure of the anatomical structures. After temporal craniotomy, the dura is dissected from bone and middle meningeal artery (MMA), as the first anatomical landmark is visible. Further top, the vertebral artery gets in contact with the accessory nerve.

Subtemporal intradural approach

The subtemporal intradural approach provides a closer look to the structures, which are not visible by the retrosigmoidal approach. Combination of both approaches enables an overview of the adjacent structures of the petrous bone in intradural lesions. After retraction of the temporal lobe, anatomical structures are visible very clearly (Figure 7). The ICA and optic nerve, which passes behind the artery, can be easily identified. Branches of the ICA, in terms of the posterior communicating artery, as well as the posterior cerebral artery, run horizontally and nearly parallel to the temporal lobe. The pituitary stalk is underlying the posterior communicating artery.
Close to the pituitary stalk, both optic nerves cross each other. The basilar artery can be identified above the ipsilateral oculomotor nerve. The arched branch is the superior cerebellar artery, which is running straight ahead to the trochlear nerve at the lateral margin of this image.

### Endoscopic approaches

Surgical resection should be the aim and can be achieved by microsurgical as well as endoscopic techniques. In the clivus, for example, endoscopic techniques have become more and more familiar in the resection of tumorous lesions in greater skull base centres, because they provide minimally invasive approaches to deep-seated lesions. Combination of classical microscopic and endoscopic techniques can increase of the quality and expansion of the field of application of surgical procedures21–27.

### Adjuvant therapy

Since gross or total tumour resection of skull base chondrosarcomas is not possible in every case, adjuvant therapy models, in terms of postoperative irradiation, can enable tumour control. However, the efficacy of the presented treatment modalities is controversial.

Iyer and co-workers performed stereotactic radiosurgery (SRS) in 22 patients with skull base chondrosarcomas. The median dose was 15.0 Gy. Seven patients died at the 75 months follow-up as a result of tumour progression. The authors postulated that patients >40 years without prior radiotherapy may benefit from early SRS after microsurgical tumour resection28.

Hauptman et al. reviewed the data of 13 patients with skull-based chordomas and chondrosarcomas, treated with linear accelerator stereotactic radiotherapy (SRT) or SRS. Under consideration of possible radiation-induced complications such as endocrinopathy, cranial neuropathy and visual impairments, the authors concluded that SRT or SRS are safe.
as an adjuvant therapy after surgical tumour resection.

A systematic review of the literature concerning proton therapy after maximal surgical resection of skull base chondrosarcomas by Amichetti and co-workers revealed a very high probability of medium- and long-term tumour control without the risk of significant complications.

Tabarkiewitz and co-workers treated one patient with dendritic cell based immunotherapy. Despite their encouraging in vitro results on chondrosarcoma cells, this therapeutic approach remains experimental and needs further investigation.

Förander and co-workers investigated the therapeutic efficacy of combination of microsurgery and Gamma Knife surgery for intracranial chondrosarcomas in nine patients. The authors concluded that local tumour control in combination of these two treatment modalities could be achieved in low-grade chondrosarcomas. On the other hand, tumour control was not satisfactory in the mesenchymal tumour type.

Nonetheless, a review of 60 cases of intracranial chondrosarcomas by Gay and co-workers summarized that chondrosarcomas do not show a dose-response after radiotherapy. Adjuvant radiotherapy seems to increase the disease-free survival, but has no influence on the total survival time. In contrast, proton beam therapy seems to increase the survival, whereby the success of this treatment is highly dependent on the resection status of the tumours.

Prognosis
Since chondrosarcomas are malignant tumours with an aggressive infiltrative behaviour, the grade of surgical resection is the key for recurrence-free survival. Brackmann and Teufert analysed the long-term outcome of eight patients with skull-based chondrosarcomas and concluded that gross total tumour resection should be achieved with adjuvant proton beam therapy to obtain the longest possible disease-free survival.

Bloch and co-workers determined the prognosis of patients with chondrosarcomas depending on different factors. The authors pointed out that the extent of tumour resection, the histopathological pattern and the application of adjuvant radiotherapy are the most significant predictive factors. The overall 5-year mortality was estimated to be 11%, with an average survival...
time of 53.7%\(^8\). Gay and co-workers determined the overall recurrence-free survival to be 80% at 3 years and 76% at 5 years. Patients with recurrence had a lower survival rate of 51% at 2 years and 26% at 3 years. In agreement with other studies, the authors confirm that the histopathological mesenchymal type is the most aggressive and is associated with lowest survival rates\(^12\).

### References

23. Frank G, Sciarretta V, Calbucci F, Farnetti G, Mazzaantena D, Pasquini E. The endoscopic transnasal transphenoidal...


