A rare case of atypical skull base meningioma with perineural spread

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ABSTRACT

Atypical meningioma is a rare cause of perineural tumour spread. In this report, we present the case of a 46-year-old female with an atypical meningioma of the skull base demonstrating perineural tumour spread. We describe the imaging features of this condition and its distinguishing features from other tumours exhibiting perineural spread.

CASE REPORT

Presentation

A 46-year-old female presented with three months of left sided facial numbness, jaw pain and headache on a background of progressive ipsilateral hearing loss over two years. There was no past medical, social or family history of note. On examination, there was sensory loss in the distribution of the maxillary and mandibular branches of the left trigeminal nerve, complete left sided deafness and a decreased left corneal reflex.

Imaging

Magnetic resonance imaging (MRI) revealed a large left sided skull base mass. The mass extended above and below the skull base by perineural spread through the foramen ovale and foramen rotundum. The mass measured 4.8 x 4.8 x 6.4cm in maximal dimensions. The mass was isointense to muscle on T1 weighted (T1) imaging (Fig. 1a-c) and hyperintense on T2 weighted (T2) imaging (Fig. 2). Post-contrast, the tumour displayed intense, homogeneous enhancement (Fig.1d-f). On diffusion-weighted imaging (DWI), there was restricted diffusion and low apparent diffusion coefficient (ADC) value throughout the tumour (Fig. 3). Inferiorly, the tumour involved the nasopharynx resulting in obstruction of the left Eustachian tube (Fig. 4). Intracranially, the tumour was extra-axial, extending along the surface of the greater wing of the left sphenoid via an enhancing dural tail (Fig. 1d,e). Posteriorly, the mass bulged through Meckel's cave into the preptontine cistern where it was inseparable from the trigeminal nerve (Fig. 5). Anteriorly, the mass extended into the pterygopalatine fossa and through the wall of the sphenoid sinus and sphenopalatine foramen into the nasal cavity (Fig. 6,7). There was denervation atrophy of the left pterygoid, temporalis and masseter muscles (Fig. 8).

Computed tomography (CT) of the head and neck revealed internal calcification within the intracranial component of the mass (Fig 9). There was widening and erosion of the foramen rotundum and foramen ovale (Fig. 11). There was hyperostosis of the surrounding sphenoid bone and bony erosion of nasal septum, the posterior maxillary alveolar margin and the left lateral wall of the sphenoid sinus (Fig. 8,10,12).

Histology

Tissue samples were initially obtained by a percutaneous CT guided core biopsy. The samples were fragmented and difficult to assess histologically. Two pathologists reviewed the samples independently arriving at differing histological diagnoses. One pathologist diagnosed a plasmacytoid neoplasm and the other diagnosed a benign meningioma. Given this discrepancy, a second percutaneous CT-guided core biopsy was performed. This sample exhibited differentiated...
meningothelial cells with an increased mitotic rate and prominent nucleoli (Fig.13a). Throughout the sample there were areas of patternless sheet-like growth and small foci of necrosis (Fig 13b). Based upon these features, a definitive diagnosis of atypical meningioma was made.

**Management**

The tumour was deemed too extensive for complete resection. The surgical team performed a two-step partial tumour resection - a maxillotomy to debulk the extracranial portion of tumour and a posterior fossa craniotomy to excise of the cerebellopontine angle tumour. Two months after surgery, the patient commenced a six-week course of adjuvant intensity-modulated radiotherapy with 54 Gray delivered in 30 daily fractions.

**Follow-up**

At the last clinical review, the patient was four months post radiotherapy and had benefitted from alleviation of her headache and facial pain. All other clinical features were stable. Follow-up MRI scans have shown a stable appearance of the residual tumour (Fig.14).

**DISCUSSION**

Meningiomas are meningothelial cell neoplasms, classically attached to the inner surface of the dura mater. They are subclassified as benign, atypical and malignant corresponding to World Health Organisation (WHO) grades I, II, and III respectively(1). Meningiomas possess mixed features of epithelial and mesenchymal cells. Atypical meningiomas are distinguished from benign meningiomas by increased mitotic activity, prominent nucleoli, uninterrupted patternless growth and foci of spontaneous necrosis. Malignant meningiomas have a greatly increased mitotic rate and features of frank anaplasia(1). Perineural spread is a mechanism whereby tumour spreads along the loose connective tissues of the perineurium.

**Epidemiology:**

Meningiomas are the most common non-glial tumours of the central nervous system. With an annual incidence of 6 per 100,000, they account for 16-20% of all intracranial tumours(2–4). Atypical meningiomas represent 4.7-7.2% of all meningiomas(5). Benign meningiomas are more common in those aged over 60 years and in females (M:F 1:2.3) whereas atypical meningiomas occur 10 years earlier on average and are more common in men (M:F 1:0.9)(6). Though perineural tumour spread is a common phenomenon in several head and neck malignancies, it has rarely been reported in meningiomas, with only 3 published cases to date(7–9).

**Etiology:**

Benign meningiomas arise following a genetic mutation on chromosome 22q12(4,10). This genetic locus encodes for the tumour suppressor protein schwannomin (also known as merlin), the same protein that is involved in neurofibromatosis type 2 (NF2). Loss of Schwannomin has a variety of biological effects that lead to a higher risk of developing benign meningiomas. Accumulation of further chromosomal mutations is thought to cause the transformation to atypical and malignant meningiomas. This transformation can occur de novo in existing benign meningioma or during tumour recurrence(4,10). Exposure to ionising radiation is the only widely recognised environmental risk factor for meningiomas(4,10,11).

**Clinical Findings:**

Common clinical findings in skull base meningiomas include impaired vision, hearing and smell as well as headache, exophthalmos and seizures depending on the position of the tumour(11–14). Perineural spread commonly effects the maxillary and mandibular branches of the trigeminal nerve(15,16). It is important to recognise that even when there is extensive perineural spread, a large proportion of patients (30-45%) remain asymptomatic with a normal cranial nerve examination(16–18). When clinical features are present, they include pain and sensory loss in a maxillary and mandibular distribution and subtle unilateral weakness of the muscles of mastication. Due to vague and non-specific features, affected patients are commonly misdiagnosed with Bell’s palsy or trigeminal neuralgia at initial presentation(19,20).

**Imaging findings:**

*Meningioma*

Meningiomas typically appear as homogeneous, hemispheric, lobular, broad-based, extra-axial masses with well-circumscribed margins and a homogeneous, avid enhancement pattern on cross-sectional imaging(11,21). The most reliable imaging feature is a “dural tail,” the presence of which infers a diagnostic sensitivity of 58% and a specificity of 94%(11). On CT, meningiomas are homogeneously hyperattenuating (72%) and cause peritumoral vasogenic oedema in the adjacent brain parenchyma (52%). They frequently display areas of internal calcification (27%) and cause peritumoral hyperostosis of the adjacent bone (18%)(21). On MRI, meningiomas are iso- to hypointense on T1 sequences and iso- to hyperintense on T2. A cerebrospinal fluid (CSF) cleft is a commonly encountered (80%) and is a reliable indicator of an extra-axial position(3).

Magnetic resonance spectroscopy (MRS) is used in cases where image findings are atypical. Meningiomas demonstrate increased choline and decreased creatine(3).

Diffusion weighted imaging has been used to distinguish between benign and atypical meningiomas. A recent prospective study of 24 patients showed a significantly lower preoperative ADC value in atypical meningiomas compared to benign meningiomas indicating restricted diffusion within the atypical group(22). If this finding is confirmed in larger studies, DWI may provide a reliable non-invasive method of differentiation between benign and atypical meningiomas.

On FDG-PET, meningiomas are universally avid. FDG-PET has also shown promise as a method of differentiating grades of meningioma, with high grade meningiomas showing a significantly higher uptake than low grade meningiomas(23).
Perineural spread

Imaging of perineural spread is more sensitive with MRI than CT(24). The first feature of perineural spread visible on imaging is caused by the breakdown of the blood-brain barrier, which results in increased permeability of the endoneurial capillaries. This process is demonstrated most conspicuously on post-contrast T1 fat suppressed sequences, seen as increased enhancement of the affected nerve. When the tumour cells proliferate further, the nerve’s diameter increases. This process results in effacement of perineural fat by hypointense tumour, seen best on T1 non fat suppressed sequences. Finally, the nerve becomes compressed and infiltrated by tumour resulting in denervation muscle atrophy of the associated muscle groups. In the acute and sub acute phases, this process results in T2 hyperintensity and mild T1 post-contrast enhancement in the muscles. In the chronic phase, this process causes progressive fatty muscle atrophy on T1 and fast spin echo T2 sequences(16,25). CT can demonstrate the widening and destruction of the neural foramina seen in perineural tumour spread. CT can also demonstrate obliteration of the perineural fat at the foraminal openings or pterygopalatine fossa, though less conspicuously than non fat suppressed T1 MRI(16,26).

Treatment

Surgery is the primary means of treatment, allowing for definitive diagnosis, reduction in mass effect and the chance of a complete cure. The aim of surgery is to perform a complete resection of tumour, dura and abnormal surrounding bone. In skull base lesions, involvement of major cranial nerves and vessels often precludes complete resection. For these cases, a balance is struck between the completeness of resection and the morbidity associated with resection of the structures involved.

Preoperative embolisation is used to decrease intraoperative blood loss and reduce tumour volume. Major risks include embolisation outside the tumour field, carotid dissection, and an increased risk of acute haemorrhage due to tumour necrosis. As a result, neurosurgical consideration of preoperative embolisation and the choice of material used varies widely(4).

Adjuvant radiotherapy use depends primarily on the completeness of surgical resection. Following subtotal resection, adjuvant chemotherapy is widely deemed beneficial(27). Following gross total excision, the literature is conflicting, with multiple retrospective studies arriving at opposing conclusions when balancing the risks and benefits of adjuvant radiotherapy(28–31).

Clinical trials of antineoplastic medical therapies for the adjuvant treatment of atypical meningioma have shown promising results though no agents are clinically available at present(32).

Prognosis

Atypical meningiomas are more aggressive than benign meningiomas, with higher rates of recurrence (29%) and higher risk of transformation to malignant meningioma(33). Perineural tumour spread often precludes complete resection and can cause irreversible neurological impairment. An early imaging diagnosis of these conditions would seem likely to result in a significant reduction in morbidity and mortality.

Differential Diagnosis

The appearance of a skull base tumour with perineural tumour spread can be caused by ascending extracranial tumours, descending intracranial tumours or tumours arising from the cranial nerves themselves.

Ascending perineural spread

Nasopharyngeal squamous cell carcinoma (SCC) is the most common cause of ascending perineural spread, occurring in 5-14% of cases(34). On T1 MRI, SCC is hypo- to isointense and commonly causes adjacent bone destruction. Post contrast, SCC exhibits mild, homogeneous enhancement. On T2 sequences, SCC is classically iso- to hyperintense and frequently causes markedly hyperintense obstructed middle ear secretions. CT commonly shows large necrotic lymph nodes and adjacent bone destruction. Fluorodeoxyglucose positron emission tomography (FDG-PET) examination shows marked avidity of the tumour and involved lymph nodes(35).

Adenoid cystic carcinoma of the salivary glands (ACC) is far less common than SCC but has a great propensity for perineural spread, which occurs in up to 60% of cases(36). On MRI, ACC is usually isointense on T1, iso- to hyperintense on T2 and exhibits moderate post contrast enhancement. CT demonstrates an infiltrative mass commonly causing erosion of the bones of the skull base, hard palate, maxilla and mandible. ACC is usually hypermetabolic on FDG-PET. Associated lymphadenopathy is an uncommon feature(37).

Non-Hodgkin lymphoma (NHL) is seen as a large mass on cross-sectional imaging. NHL is usually isointense on T1 MRI and exhibits moderate, homogeneous post contrast enhancement. The appearance of NHL on T2 sequences depends on the tumour cellularity, though it is classically isointense. CT shows non-necrotic bulky nodal disease in approximately 50% of cases, and frequently demonstrates deep invasion of the surrounding bones of the skull base. NHL is avid on FDG-PET(38).

Nerve based tumours

Trigeminal schwannomas classically arise from the mandibular branch of the trigeminal nerve. They are well-defined tumours, which extend intra- and extracranially along the path of least resistance. On MRI, Schwannomas are iso- to hypointense on T1 sequences and iso- to hyperintense on T2. Post-contrast, Trigeminal schwannomas can enhance homogeneously or heterogeneously, with interspersed non-enhancing intramural cysts considered characteristic. On CT, schwannomas classically cause smooth enlargement of the foramen ovale without bone erosion. Trigeminal schwannomas are avid on FDG-PET(39).

Solitary neurofibromas of the head and neck are rare and not usually associated with neurofibromatosis type 1(40). Neurofibromas are well-defined tumours that most commonly arise from the ophthalmic and maxillary branches of the trigeminal nerve. On MRI, neurofibromas are classically
hypointense on T1 sequences, hyperintense on T2 sequences and exhibit homogeneous post contrast enhancement. In some cases, they may possess a hypointense centre that enhances post contrast relative to the periphery (the “target sign”) or contain multiple small hypointense foci that represent fascicular bundles (the “fascicular sign”). Neurofibromas are classically hypointenuating on CT (5–25 hounsfield units) and cause smooth enlargement of the surrounding neural foramen. Solitary neurofibromas are FDG-PET avid(41).

TEACHING POINT
Perineural spread of a skull base tumor has a wide differential diagnosis. Skull base tumors are often difficult to access for biopsy, rendering radiological diagnosis of paramount importance. Though atypical meningioma is a rare cause of perineural spread, the high overall incidence of meningioma warrants its consideration in the differential diagnosis. When considered, the distinct imaging characteristics of meningioma such as a dural tail, internal calcification and hyperostosis of the adjacent bone differentiate it from other possible diagnoses.

REFERENCES


Neuroradiology: A rare case of atypical skull base meningioma with perineural spread

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FIGURES

Figure 1: 46-year-old female with an atypical skull base meningioma. FINDINGS: Pre contrast (a-c) and post contrast fat-saturated (d-f) T1 sagittal, coronal and axial sections demonstrating maximal tumour dimensions, isointense tumour signal pre contrast and homogeneous tumour enhancement post contrast. TECHNIQUE: GE Medical Systems Discovery MR450 1.5 Tesla, (a-c) Pre contrast T1 Fast Spin Echo (FSE): TR 545ms, TE 16.64ms. (d-f) Post contrast fat saturated FSE: TR 627ms, TW16.64. Gadolinium intravenous contrast.
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Figure 2: 46-year-old female with an atypical skull base meningioma. FINDINGS: T2 axial section demonstrating iso- to hyperintense tumour signal. TECHNIQUE: GE Medical Systems Discovery MR450 1.5 Tesla, T2 Fast Spin echo, TR 6790ms, TE 98.98ms.

Figure 3: 46-year-old female with an atypical skull base meningioma. FINDINGS: Diffusion Weighted Imaging (b1000) and Apparent Diffusion Coefficient axial sections demonstrating restricted diffusion on DWI images (a) and a low ADC value (b). TECHNIQUE: GE Medical Systems Discovery MR450 1.5 Tesla, Diffusion Weighted Imaging and Apparent Diffusion Coefficient, TR 8000ms, TE 82.6ms.

Figure 4: 46-year-old female with an atypical skull base meningioma. FINDINGS: T2 axial sections demonstrating extension of tumour into the nasopharynx (white arrow) resulting in obstruction of the left Eustachian tube and fluid opacification of the left mastoid air cells (red arrows). TECHNIQUE: GE Medical Systems Discovery MR450 1.5 Tesla. T2 FSE: TR 6790ms, TE 98.98ms.
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Figure 5: 46-year-old female with an atypical skull base meningioma. FINDINGS: Post contrast T1 fat-saturated (a) and T2 (b) axial sections showing (a) tumour within the cavernous sinus and Meckel’s cave invading into the sphenoid sinus anteriorly and (b) tumour in the preoptic cistern, inseparable on imaging from the proximal trigeminal nerve. TECHNIQUE: GE Medical Systems Discovery MR450 1.5 Tesla. (a) Post contrast T1 fat saturated FSE, TR 627ms, TE16.64. Gadolinium intravenous contrast. (b) T2 FSE, TR6790, TE 98.98.

Figure 6: 46-year-old female with an atypical skull base meningioma. FINDINGS: Post-contrast T1 fat saturated coronal and axial sections demonstrating tumour eroding through (a) the wall of the sphenoid sinus and (b) through the sphenopalatine foramen into the nasal cavity. TECHNIQUE: GE Medical Systems Discovery MR450 1.5 Tesla. Post contrast fat saturated FSE: TR 627 ms, TW16.64. Gadolinium intravenous contrast.
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Figure 7 (left): 46-year-old female with an atypical skull base meningioma. FINDINGS: Axial CT section demonstrating widening of the sphenopalatine foramen and tumor extension into the nasopharynx. TECHNIQUE: Non contrast axial CT, 130 mAs, 120 kV, 1.5mm slice thickness.

Figure 8 (bottom): 46-year-old female with an atypical skull base meningioma. FINDINGS: Pre contrast T1 axial sections demonstrating denervation atrophy of the (a) pterygoid and masseter muscles and (b) temporalis muscle on the left side due to long-standing perineural spread. TECHNIQUE: GE Medical Systems Discovery MR450 1.5 Tesla. Pre contrast T1 FSE, TR 545ms, TE 16.64ms.
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Figure 9: 46-year-old female with an atypical skull base meningioma. FINDINGS: Axial, coronal and sagittal CT sections demonstrating internal calcification of the intracranial portion of the mass and hyperostosis and erosion of the greater wing of the sphenoid. TECHNIQUE: Non-contrast axial CT with coronal and sagittal reconstructions, 218mAs, 120 kV, 1.5mm slice thickness.

Figure 10 (left): 46-year-old female with an atypical skull base meningioma. FINDINGS: Axial CT of the skull base demonstrating tumour extending into the nasopharynx (white arrow) with associated erosion of the left pterygoid plate and posterior maxillary alveolus (red arrow). TECHNIQUE: Non-contrast axial CT, 218mAs, 120 kV, 1.5mm slice thickness.
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Figure 11: 46-year-old female with an atypical skull base meningioma. FINDINGS: Sagittal (a,c) and Coronal (b,d) CT sections demonstrating foramen ovale expansion (a,b) and foramen rotundum expansion (c,d) with hyperostosis of the surrounding sphenoid wing. TECHNIQUE: Non contrast axial CT, 218mAs, 120kV, 1.5mm slice thickness.

Figure 12 (left): 46-year-old female with an atypical skull base meningioma. FINDINGS: Axial CT section demonstrating mixed hyperostosis and erosion of the greater wing of the sphenoid. TECHNIQUE: Non-contrast axial CT, 218mAs, 120kV, 1.5mm slice thickness.
Figure 13: 46-year-old female with an atypical skull base meningioma. FINDINGS: Pathological sections showing (a) increased mitotic activity and prominent nucleoli at 40x magnification and (b) uninterrupted patternless, sheet-like growth at 20x magnification. These are key diagnostic features of atypical meningioma. TECHNIQUE: Hematoxylin-eosin stained core biopsy sections at (a) 40x and (b) 20x magnification.

Figure 14: 46-year-old female with an atypical skull base meningioma. FINDINGS: Post contrast T1 axial sections showing the post-operative appearances of the tumour following (a) maxillotomy to debulk the extracranial portion of tumour and (b) a posterior fossa craniotomy to excise of the cerebellopontine angle tumour. TECHNIQUE: GE Medical Systems Discovery MR450 1.5 Tesla, Post contrast T1 FSE: TR 552ms, TE 10.41. Gadolinium intravenous contrast.
## Etiology
- Genetic mutation at chromosome 22q12 causes the loss of tumour suppressor protein
- Loss of tumour suppressor protein “Schwannomin” causes an increased risk of developing benign meningiomas.
- Further accumulation of chromosomal mutations leads to transformation to atypical and malignant meningioma.

## Incidence
- 4.7-7.2% of all meningiomas.
- Approximately 3 in 1,000,000.

## Gender ratio (M:F)
- 1:0.9 (compared to 1:2.3 for benign meningiomas)

## Ethnicity
- Benign meningiomas are common in African-Americans.
- No ethnic predominance has been seen in small studies of atypical meningioma.

## Age predilection
- Middle age, approximately 10 years earlier than benign meningioma.

## Risk factors
- Neurofibromatosis Type 2
- Previous ionising radiation exposure.

## Treatment
- Surgery +/- adjuvant radiotherapy +/- preoperative embolisation

## Prognosis
- 5-year survival: 86%, 10 year survival: 61%
- 5-year recurrence free survival: 48%
- Overall recurrence: 26%
- Median time to recurrence: 3 years

## MR imaging
- CSF cleft (80%); Dural tail (58%)
- T1: iso- to hypointense
- T2: iso- to hyperintense
- +C: Homogeneously enhancing
- MRS: High choline, Low creatine
- DWI: Decreased ADC values

## CT imaging
- Homogeneous hyerattenuation (72%)
- Frequently tumoral calcification (18%)
- Peritumoral vasogenic oedema (52%)
- Hyperostosis of the adjacent bone (18%)

## FDG-PET imaging
- FDG- avid

**Table 1**: Summary table for atypical skull base meningioma.
### Table 2: Differential diagnosis table for atypical skull base meningioma with perineural spread.

<table>
<thead>
<tr>
<th>MRI</th>
<th>CT</th>
<th>PET/CT</th>
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<tbody>
<tr>
<td><strong>Atypical meningioma</strong></td>
<td>T1: Iso- to hypointense</td>
<td>Tumoral calcification (20%)</td>
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<td></td>
<td>T2: Iso- to hyperintensity.</td>
<td>Expansion +/- erosion of neural foramina</td>
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<td>T2: CSF cleft</td>
<td>FDG avid</td>
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<tr>
<td></td>
<td>T1+C: Homogeneous enhancement</td>
<td></td>
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<tr>
<td></td>
<td>T1+C: Enhancing dural tail</td>
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<td></td>
<td>MRS: High choline, low creatinine</td>
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<td></td>
<td>DWI: Decreased ADC values</td>
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<tr>
<td><strong>Squamous Cell Carcinoma</strong></td>
<td>T1: Hypo- or isointense</td>
<td>Large necrotic lymph nodes</td>
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<td></td>
<td>T2: Iso- to hyperintense</td>
<td>Bony destruction</td>
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<td></td>
<td>T2: Hyperintense obstructed middle ear secretions</td>
<td>FDG avid tumour, lymph nodes and metastases</td>
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<td></td>
<td>T1+C: Mild, homogeneous enhancement</td>
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<tr>
<td><strong>Adenoid cystic carcinoma</strong></td>
<td>T1: Isointense</td>
<td>Erosion of the mandible or maxilla.</td>
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<td>T2: Iso- to hyperintense</td>
<td>Enlarged lymph nodes not usually a feature</td>
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<td>T1+C: Moderate, homogeneous enhancement</td>
<td>FDG avid</td>
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<td><strong>Non-Hodgkin Lymphoma</strong></td>
<td>T1: Isointense</td>
<td>Hyperdense</td>
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<td>T2: Variable depending on tumour cellularity usually isointense</td>
<td>Non-necrotic bulky lymph nodes (50%)</td>
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<td>T1+C: Moderate, homogeneous enhancement</td>
<td>Deep erosion of the surrounding bone</td>
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<tr>
<td><strong>Schwannoma of CNV3</strong></td>
<td>T1: Iso- to hypointense</td>
<td>Smooth enlargement of foramen ovale without erosion.</td>
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<td></td>
<td>T2: Iso- to hyperintense</td>
<td>FDG avid</td>
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<tr>
<td></td>
<td>T1+C: Variable enhancement pattern</td>
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<td></td>
<td>T1+C: Intratumoral cysts are characteristic</td>
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<tr>
<td><strong>Neurofibroma of CNV3</strong></td>
<td>T1: Hypointense</td>
<td>Hypodense (5-25 HU)</td>
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<td>T1: “Target sign” - hypointense centre with hyperintense periphery</td>
<td>Smooth enlargement of neural foramina</td>
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<td>T1: “Fascicular sign” - multiple small hypointense foci</td>
<td>FDG avid</td>
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<td>T2: Hyperintense</td>
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<td>T1+C: Homogeneous enhancement</td>
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<td>T1+C: “Target sign” - centre enhances relative to periphery</td>
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### ABBREVIATIONS

ACC – Adenoid cystic carcinoma  
ADC – Apparent diffusion coefficient  
CSF – Cerebrospinal fluid  
CT – Computed tomography  
DWI – Diffusion weighted imaging  
FDG-PET – Fluorodeoxyglucose positron emission tomography  
MRI – Magnetic resonance imaging  
MRS – Magnetic resonance spectroscopy  
NF2 – Neurofibromatosis type 2  
NHL – Non- Hodgkin lymphoma  
SCC – Squamous cell carcinoma  
WHO – World health organisation  

### Keywords

Atypical meningioma; perineural spread; skull base tumour; skull base mass

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