Internal jugular vein extension of Grade I cerebellopontine angle meningioma

A 67-year-old woman presented with a soft non-tender left cervical swelling, evident for some months. She had a relevant past history of left sided Grade I cerebellopontine angle meningioma, partially resected 12 years earlier and which had remained stable on follow-up imaging, and chronic lymphatic leukaemia (Binet Stage A).

Imaging Findings:

Neck ultrasound revealed a heterogeneous mass distending the left internal jugular vein with associated flow on Doppler examination (Fig. 1a-c). Post-contrast CT confirmed the intra-luminal location of this mass, with only a small rim of contrast between it and the vessel wall (Fig. 2a). Small enhancing vessels were seen within the mass (Fig. 2b). At the skull base, a large enhancing mass was noted at the left petrous apex, the site of the known residual meningioma (Fig 2c). Pre- and post-gadolinium MRI sequences demonstrated that the internal jugular vein mass was low signal on T1 and high signal on STIR sequences, enhancing homogeneously post-contrast (Fig. 3a-d). The left cerebellopontine angle extra-axial mass extended into surrounding structures (including the left jugular foramen and clivus), enhanced uniformly and had a dural tail (Fig. 4a-f). Ultrasound guided fine needle aspiration of the jugular vein mass confirmed intravenous extension of the skull base meningioma.

Discussion:

Meningiomas arise from arachnoid cap cells in the meningeal coverings of the spinal cord and brain [1]. They are the most frequently diagnosed primary central nervous system tumour in adults, accounting for up to a third of these tumours [2, 3]. Meningiomas are classified according to the World Health Organisation (WHO) grading system as Grade I-III, recently updated in 2007 [1]. 80-90% of tumours are Grade I and, although defined as benign, show significant intra-class heterogeneity with recurrence rates of 7-25%.

Extracranial presentation of meningioma is uncommon and may occur with direct extension (most commonly),
metastasis or primary extracranial meningioma. Direct extension into internal jugular vein, although much less common than involvement of the dura, adjacent venous sinuses and jugular foramen, has been described in a small number of case reports with implications for further management [4-6].

Seminal data collected by Simpson in the 1950s showed a clear correlation between completeness of surgical resection and 10-year recurrence rates, with subtotal resection corresponding to a 4-fold greater risk of recurrence compared with complete resection [7]. However, an important recent study has reported minimal differences in recurrence free survival when comparing aggressive with more conservative surgical approaches in the context of Grade I tumours, likely reflecting improvements in surgical technique [8]. Clearly, there is a compromise between complete excision, risk of recurrence/progression and functional outcomes and this is influenced by the precise anatomical location of the tumour. Even within the subcategory of cerebellopontine angle meningioma a representative study reports complete resection rates ranging from 100%-73% with functional preservation of the facial nerve ranging from 93%-73% [9].

Despite the revised WHO meningioma grading, aggressive behaviour is difficult to predict and there is a need for radiological parameters predictive of clinical outcomes. Whilst magnetic resonance (MR) is the most useful imaging modality in meningioma assessment, standard sequences do not readily differentiate benign and non-benign tumours. Furthermore, recent multivariate analysis of general tumour parameters on MR (including tumour size, location, associated calcification and oedema, brain-tumour interface and pial-cortical blood supply) found that only residual tumour volume >3cm3 was predictive of regrowth in residual Grade I tumours [10]. However, functional MR parameters with or without positron emission tomography (PET) are being correlated with clinical parameters and grade in some studies and, although still controversial, are likely to significantly impact on surgical and radiological management in the future [11].

Differential Diagnosis List: Internal jugular vein extension of Grade I cerebellopontine angle meningioma., Schwannoma, Glomus jugulare tumour, Intravascular capillary endothelial hyperplasia, Angiosarcoma

Final Diagnosis: Internal jugular vein extension of Grade I cerebellopontine angle meningioma.

References:

Rigau V (2011) French brain tumor database:5-year histological results on 25756 cases. (PMID: 21554472)
Seo EK (2008) Unusual intracranial parasagittal meningioma extending into the internal jugular vein through the sinuses. (PMID: 2588220)
Taki N (2013) Extracranial intraluminal extension of atypical meningioma within the internal jugular vein. (PMID: 3586463)
Chen CM (2011) Contemporary surgical outcome for skull base meningiomas. (PMID: 21614426)
Description: Transverse ultrasound image showing a large heterogeneous echogenic mass filling and distending the left internal jugular vein. Origin: Department of Radiology, Aberdeen Royal Infirmary, Aberdeen.
Description: Longitudinal ultrasound image showing the mass extending along the length of the left internal jugular vein. The cranial extent of the mass was not possible to determine on ultrasound.

Origin: Department of Radiology, Aberdeen Royal Infirmary, Aberdeen.
Description: Longitudinal Doppler ultrasound image demonstrating signal within the mass and distinguishing it from thrombus. Origin: Department of Radiology, Aberdeen Royal Infirmary, Aberdeen.
Description: Axial image showing a large filling defect in the left internal jugular vein with small enhancing vessels. There is almost complete obstruction of the distended vessel with only a thin rim of contrast around it. Origin: Department of Radiology, Aberdeen Royal Infirmary, Aberdeen.
**Description:** Coronal image showing the craniocaudal extent of the internal jugular vein filling defect. Again, small enhancing vessels are noted within the filling defect. **Origin:** Department of Radiology, Aberdeen Royal Infirmary, Aberdeen.
Description: Axial image of the skull base showing a well-defined partially enhancing mass adjacent to the left petrous apex, in close proximity to the left internal auditory meatus. Occipital skull defect in keeping with previous surgery. Origin: Department of Radiology, Aberdeen Royal Infirmary, Aberdeen.
Description: Coronal STIR image: the craniocaudal extent of the internal jugular vein filling defect can be seen, extending from skull base to brachiocephalic vein. Origin: Department of Radiology, Aberdeen Royal Infirmary, Aberdeen.
Description: Axial T1 image pre contrast: large low signal filling defect noted within the distended left internal jugular vein. Origin: Department of Radiology, Aberdeen Royal Infirmary, Aberdeen.
Description: Axial T1 image post contrast: almost homogenous enhancement of the internal jugular vein filling defect is noted. **Origin:** Department of Radiology, Aberdeen Royal Infirmary, Aberdeen.

Description: Coronal T1 post contrast: the mass shows largely homogenous enhancement throughout its length. **Origin:** Department of Radiology, Aberdeen Royal Infirmary, Aberdeen.
Description: Axial T1 pre contrast: large extra-axial mass at the left cerebellopontine angle extending into left internal auditory canal. The mass is isointense to brain and distorts the pons. Origin: Department of Radiology, Aberdeen Royal Infirmary, Aberdeen.
Description: Axial T1 pre contrast: there is extension of the low signal mass into the clivus on the left side as well as the left jugular foramen. Origin: Department of Radiology, Aberdeen Royal Infirmary, Aberdeen.
Description: Axial T1 post contrast: uniform avid enhancement of the left cerebellopontine angle mass with enhancing tissue also extending into the left internal auditory canal. A dural tail is noted. Origin: Department of Radiology, Aberdeen Royal Infirmary, Aberdeen.
Description: Coronal T1 post contrast: again, uniform enhancement of the left cerebellopontine angle mass is seen with distortion of the adjacent pons. Origin: Department of Radiology, Aberdeen Royal Infirmary, Aberdeen.
**Description:** Sagittal T1 post contrast: large left cerebellopontine mass, uniformly enhancing, with extension towards the left cerebellar hemisphere posteriorly and clivus anteriorly. **Origin:** Department of Radiology, Aberdeen Royal Infirmary, Aberdeen.
Description: Axial T2 post contrast: infiltration of tumour into clivus and left petrous apex with
extension into left internal jugular foramen. Thickening and enhancement of the dura anterior to the
medulla oblongata is noted. Origin: Department of Radiology, Aberdeen Royal Infirmary, Aberdeen.