A 70-year-old woman attended her general practitioner complaining of binocular diplopia for the last 15 days. Relevant medical history includes only general cardiovascular risk factors.

Neurologic examination demonstrates binocular diplopia to levoversion with a left VI cranial-nerve (CN) palsy. Other CNs preserved.

Imaging Findings:

Non-enhanced computed tomography (NECT) demonstrates a clival lesion infiltrating the skull base, extending into the nasopharynx and endocranium (Fig 1a). It indents the anterior aspect of the pons, and invades both cavernous sinuses, and the left Meckel's cave (Fig 1b). The lesion contains multiple internal calcifications that are reminiscent of dots and commas (chondroid-pattern). Contrast-enhanced CT (CECT) shows a moderate honeycomb-like pattern of enhancement (Fig 1c).

Magnetic resonance imaging (MRI) confirms this mass, with homogeneously low signal intensity on T1SE (Fig 2a), which surrounds and encases both internal carotid arteries with erosion of both carotid canals. T2TSE signal is heterogeneously hyperintense, with multiple cystic areas (Fig 2b). Post-contrast images show moderate honeycomb-like, predominantly peripheral enhancement (Fig 2c). Diffusion-weighted imaging shows moderate restriction, with relatively low ADC values (615*10^-6 +/- 65*10^-6 mm^2/s) (Figs 2c, d).

Discussion:

Chordomas are rare malignant tumours derived from the primitive notochord (extending from Rathke's pouch to the coccyx). They can be identified anywhere along the vertebral column and are relatively evenly distributed between cranium, sacrum and vertebral bodies [1, 2].

This tumour can occur at any age, with a slight male predilection, though spheno-occipital chordomas tend to be observed in younger age groups than sacral chordomas [1]. Spheno-occipital chordomas are slow-growing, very locally aggressive tumours. Distant metastases are rare [2].

These patients most commonly present with VI CN diplopia or headache [1]. VI CN is affected relatively early due to clival-centered expansion of the tumour, which promptly involves the abducens nerve's interdural segment in Dorello's canal just over the petroclival fissure [3, 4, 5].

CT of spheno-occipital region chordomas demonstrates an infiltrative mass with extensive bone destruction, and
areas of calcification, which may correspond to sequestrae. Frank tumoral calcification may be observed in the chondroid chordoma variant. The lesion is most commonly located around the clivus, and may infiltrate both intra and extracranial skull base structures. Endocranial pre-pontine invasion often demonstrates the classical “thumbprinting” sign, in which the endocranial part of the tumour indents the anterior aspect of the pons.

MRI demonstrates the same extensive bone-infiltration as NECT, along with intracranial and extracranial spread. T1WI signal is intermediate/low, with possible small foci of T1-hyperintensity due to intrallesional haemorrhage. T2WI shows a heterogeneously hyperintense lesion due to abundant fluid component combined with vacuolated-cystic areas, along with less intense areas of proteinaceous and haemorrhagic content. Enhancement is typically moderate to intense, with a heterogeneous “honeycomb” appearance due to areas of necrosis and cystic degeneration [1, 5].

DWI has been described as being useful in differentiating chordomas from clival-chondrosarcomas. One study showed chondrosarcoma ADC values to be significantly higher than those of chordomas, which in turn were higher than those of atypical chondrosarcomas [6].

The rarity of distant metastasis calls for radical surgical excision. This is limited and complicated by invasion of skull-base structures. Residual tumour is treated with different forms of radio or brachytherapy [2, 7].

Local recurrence is common. Periodic follow-up MRI should be performed [1].

Take-Home-Messages
- Locally-aggressive tumours infiltrating the skull-base, with intra and extracranial invasion.
- Low T1, High T2 with cystic areas. Heterogeneous “honeycomb” -enhancement.
- Lower ADC than chondrosarcoma.
- Prepptine thumbprinting by the intracranial component.
- Binocular diplopia: most common initial symptom (VI-CN palsy)
- Metastases are very rare. Aggressive local treatment is indicated.

Differential Diagnosis List: Clival chondroid chordoma, Invasive pituitary macroadenoma, Skull base chondrosarcoma, Skull base plasmacytoma, Skull base metastasis, Skull base meningioma

Final Diagnosis: Clival chondroid chordoma

References:

Description: Infiltrative clival lesion with intralesional calcifications, some chondroid-like (dots-&-commas). Invades posterior nasopharynx, carotid canals (a), and endocranium, bulging and "thumbprinting" the anterior aspect of the pons (b). Also invades both cavernous venous sinuses (b).

Origin: Hospital Unviersitari de Bellvitge
**Description:** Infiltrative clival lesion with intrallesional calcifications, some chondroid-like (dots-&-commas). Invades posterior nasopharynx, carotid canals (a), and endocranium, bulging and "thumbprinting" the anterior aspect of the pons (b). Also invades both cavernous venous sinuses (b).

**Origin:** Hospital Unviersitari de Bellvitge
Description: Fig 1c. CECT shows heterogeneous, honeycomb-like enhancement of the clival lesion (c).

Origin: Hospital Universitari de Bellvitge
Description: Axial T1SE. Homogeneously intermediate-hypointensity. Right carotid artery encasement.
Origin: Radiology Department. Hospital Universitari de Bellvitge.
**Description:** Coronal T2TSE. Heterogeneously hyperintense with cystic foci. **Origin:** Radiology Department. Hospital Universitari de Bellvitge.
Description: Sagittal T1+contrast. Heterogeneous “honeycombing” enhancement, with a predominantly peripheral distribution. Endocranial invasion is evident, with pontine “thumbprinting”.

Origin: Radiology Department. Hospital Universitari de Bellvitge.
Description: DWI. Discrete hyperintensity of the lesion. Origin: Radiology Department. Hospital Universitari de Bellvitge.
Description: ADC map. Region of interest (red circle) relatively low ADC values (615E-6 +/- 65E-6 mm^2/s). Origin: Radiology Department. Hospital Universitari de Bellvitge.