Calcified Chondroid Mesenchymal Neoplasm

Ricardo Sánchez Jiménez, MD[1], Alejandro Arbona-Lampaya, BS[2], Alejandro Odeh Couvertier, BS[2], Eduardo J Labat, MD[1]

[1]Department of Radiology, School of Medicine, University of Puerto Rico Medical Sciences Campus, San Juan, Puerto Rico.
[2]School of Medicine, University of Puerto Rico Medical Sciences Campus, San Juan, Puerto Rico.

Corresponding Author:
Ricardo Sánchez Jiménez, MD
Department of Radiology, School of Medicine, University of Puerto Rico Medical Sciences Campus, San Juan, Puerto Rico, 00935
Email:ricardo.sanchez7@upr.edu
Telephone: +1-787-477-7044







What is a Calcified Chondroid Mesenchymal Neoplasm (CMNN)?

A rare, benign soft tissue tumor made of cartilage-type (chondroid) tissue with variable internal calcification, driven by a characteristic gene fusion most often involving FN1 gene.

What is the typical patient population?

Middle-aged adults with mean age ~51–52 years (range 14–87) and equal male-to-female distribution.

Clinical Presentation



Chief Complaint

CCMN typically presents as a slowly enlarging, palpable soft tissue mass/swelling referred for expert consultation out of concern for malignancy.

Site Predilection and Size

- Small at diagnosis: mean size ~2.1 cm, usually well-defined
- Distal extremities (hand, fingers, foot, toes, wrist, forearm) causing painless or mildly painful swelling, sometimes limiting movement
- Temporomandibular joint / temporal / parotid region may present as a preauricular or facial mass, jaw/TMJ swelling, or dysfunction .

Clinical Presentation : Behavior



Benign, indolent course

No reported metastases

Recurrence is rare

Only after incomplete removal

Looks nonspecific

Symptoms overlap with other cartilage-type lesions such as chondrosarcoma

Diagnosis

needs tissue and molecular proof of FN1 fusion, not clinical features alone

Imaging Hallmarks

- Key Finding: Soft tissue mass with variable internal calcification
- Well-circumscribed margins
- Small size: mean ~2.1 cm
- Benign-looking on imaging in every reported case, even though the histology looks worrisome

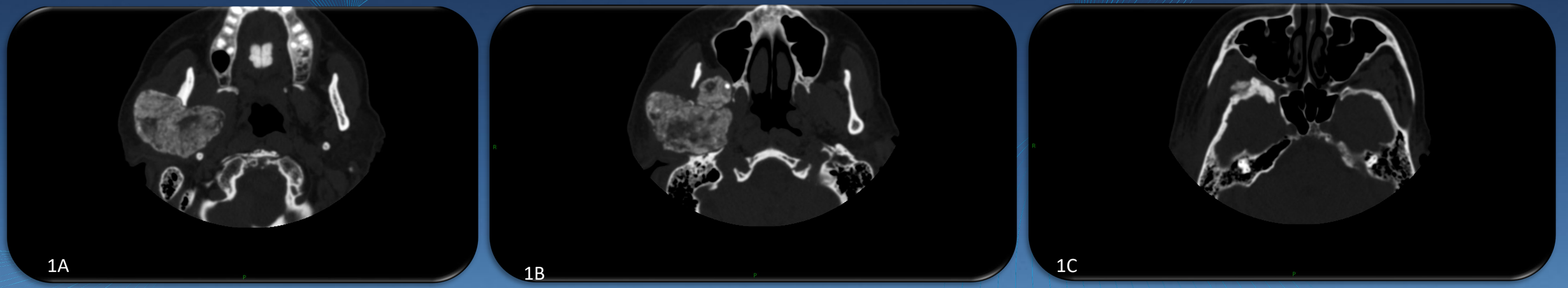
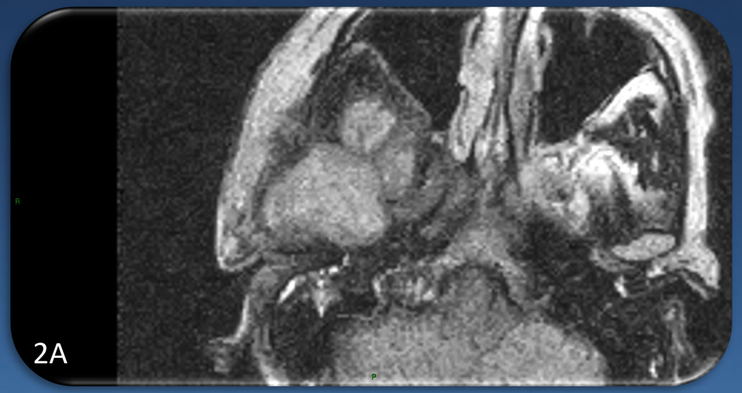
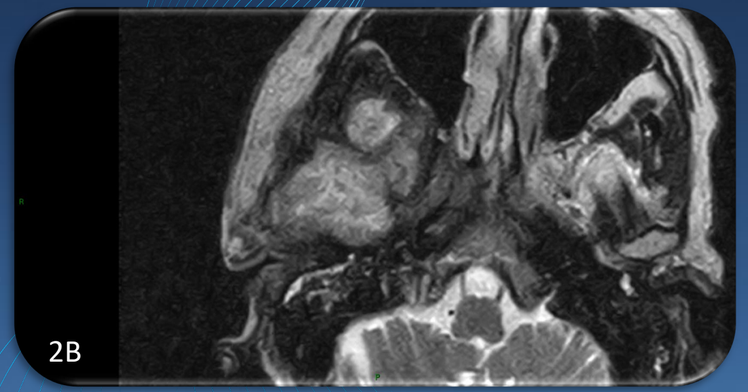


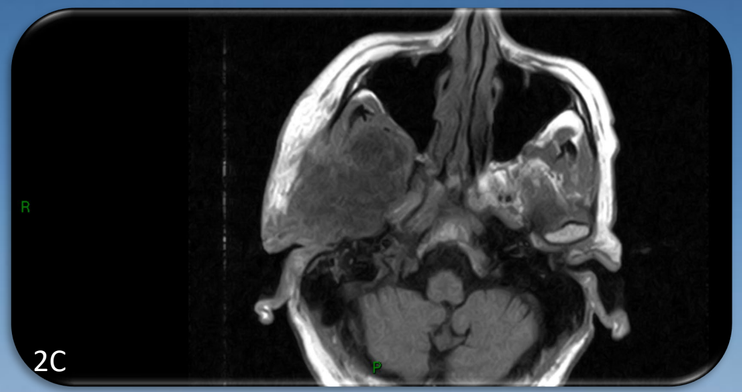
Figure 1 . Lobulated chondroid mass involving the right mandibular condyle. Axial CT image (A) shows a lobulated predominantly chondroid mass with scattered calcific foci and internal areas of hypoattenuation, replacing the right mandibular head/neck, scalloping anteriorly at the mandibular body (1a) and superiorly at the greater wing of the sphenoid bone (1b), while also showing hyperostosis (1c).

Imaging Hallmarks

- Calcification described as "grungy" and/or "lace-like" on imaging–pathology correlation
- May scallop nearby bone but no aggressive cortical destruction
- Our case is associated bone hyperostosis, but causal vs. incidental relationship undetermined







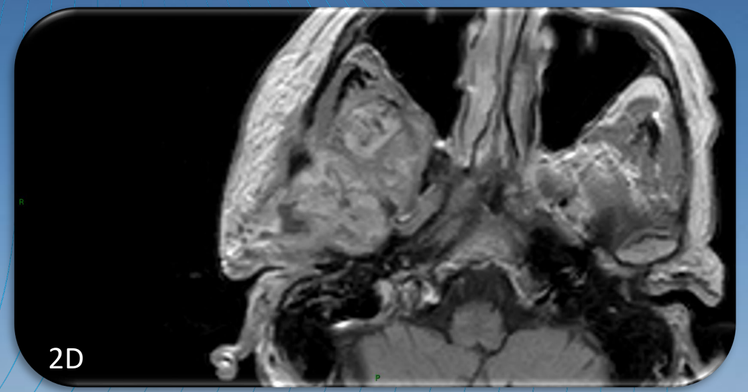


Figure 2. Lobulated chondroid mass involving the right mandibular condyle. Axial MRI shows a lobulated heterogeneous predominantly T2/FLAIR hyperintense mass (2a/2b) with T1hypointensity(2c) and heterogeneous avid enhancement (2d) with areas of internal non-enhancement.

Management

First-line treatment is surgery

Complete surgical excision with negative margins to prevent recurrence

Molecular testing

Look for FN1 rearrangements, most commonly FN1::FGFR2, but also FGFR1, TEK, MERTK, NTRK1

Less often PDGFRA::USP8

Diagnosis

Must be differentiated from chondrosarcoma to avoid overtreatment

Test Methods

RNA sequencing / FISH are most helpful.

Immunohistochemistry is of limited diagnostic help.

Chemotherapy or Radiation

Management is surgical given the benign, indolent biology.

Mimics

Soft tissue chondroma, tenosynovial giant cell tumor, tophaceous pseudogout, phosphaturic mesenchymal tumor, chondrosarcoma

Outcomes

Benign behavior

No malignanttransformation or metastasis in the literature

Excellent disease -free survival

The largest series with follow-up (15/33 patients), all disease free except one wrist recurrence after incomplete excision.

Recurrence is uncommon

Only 2 reported, both after incomplete excision.



Key message

Recognition prevents misdiagnosis as chondrosarcoma and avoids unnecessary aggressive therapy

Take Home Points

- Think CCMN for an indolent, calcified chondroid mass in the distal extremities or TMJ of a middle-aged adult
- Confirm with histology + FN1 fusion testing, and separate it from chondrosarcoma
- Treat with conservative, complete excision