

Extranodal nasal natural killer/T-cell lymphoma (ENKTL)

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Section: Head & neck imaging

Area of Interest: Head and neck Oncology

Procedure: Contrast agent-intravenous

Procedure: Diagnostic procedure

Imaging Technique: CT

Imaging Technique: MR

Special Focus: Neoplasia Case Type: Clinical Cases

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Patient: 53 years, male

Clinical History:

Patient with severe left facial oedema of two months of progress, treated with several cycles of antibiotics without any improvement.

Imaging Findings:

Facial CT showed a voluminous solid lesion centred in the left nasal cavity, which extended from the middle meatus to the nasopharynx and the ipsilateral pterygopalatine fossa, reaching the left infratemporal fossa and with left orbital invasion (Fig. 1). Mild lysis of the left middle turbinate and ipsilateral papyraceous lamina was appreciated (Fig. 2).

MRI confirmed the presence of an infiltrating growth pattern mass centred in the left nasal cavity which extended anteriorly to the nasolabial soft tissues, posteriorly to the left nasopharynx, reaching the left pterygopalatine fossa and the left masticatory space. The lesion was isointense to the muscle in T1WI and showed mild hyperintensity in T2, with restricted diffusion (ADC 0.4×10^{-3} mm²/s) and avid heterogeneous contrast enhancement, due to necrotic areas (Fig. 3, 4). Thus, perineural spread through left V3 and denervation changes were depicted. No intracranial extension was found (Fig. 4).

Whole body CT, PET-CT and gammagraphy showed no systemic findings.

Discussion:

This entity has been classified as extranodal NK/T-cell lymphoma (ENKTL), nasal type, by the WHO since 1994 [1]. It is almost always associated with Epstein-Barr virus. It has dramatic ethnic and geographic predilection, accounting for 7% to 10% of non-Hodkin's lymphoma (NHL) cases in Asia and Latin American countries, but only 1% of Caucasian cases [2, 3]. It has higher prevalence in men, especially in the sixth decade.

75% of ENKTL cases occur in the upper aerodigestive tract. Most frequent locations are nasal cavities and paranasal sinuses, especially the maxillary sinus [2].

Most common symptoms are nasal obstruction and facial swelling.

CT is the first imaging procedure. Findings include a solid mass arising from the nasal cavity which tends to infiltrate surrounding structures. Mild bone destruction is frequently seen [2, 3, 4, 5].

Typical MR findings are ill-defined invasive heterogeneous soft tissue mass, which has heterogeneous signal on both T1 and T2WI. Necrotic components are often present [3, 6, 7].

Final diagnosis is histopathological. Sample shows commonly an inflammatory invasion of the vessel wall with final occlusion of the lumen and necrotic changes. The immunophenotypic profile of these tumour cells includes: CD2 +, CD56 +, CD3e cytoplasmic +, CD3 surface - and TCR- [8, 9].

Even though ENKTL is a rather aggressive lymphoma, it remains localised in the nasal primary site in many cases, with only 15-30% regional nodal spread. Systemic dissemination is uncommon, under 20%. It metastasises to the skin, gut, ovaries, testis and muscle. Staging is based upon local invasion, extranasal spread, regional nodal spread and metastases [4, 5].

Stages I & II include T1-T2-T3N0M0 and T1-2N1M0. Median survival period is 2 years. Radiotherapy is mandatory. PET/CT aids the staging of and radiotherapy planning.

Stage III includes T3N1M0, T1-3N2M0 and T4N0M0. Recommended treatment is a combination of chemotherapy and radiotherapy.

Stage IV includes TanyNanyM1. Treatment includes high doses of chemotherapy. 5-year survival rate is below 10%. CHOP is the commonly used regimen [4, 9].

The tumour shows high chemosensitivity and radiosensitivity but it presents a high rate of recurrence. Allogenic haematopoietic stem-cell transplantation represents another optional treatment.

Teaching points:

1. ENKTL nasal type is rare, has an ethnic predilection and it is associated with Epstein-Barr virus.
2. MRI/CT findings include a mass with diffuse invasion of the nasal cavity, necrosis, bony destruction and extension into the nasopharynx.
3. Tumour shows a high rate of chemosensitivity and radiosensitivity.
4. For disseminated and refractory cases the 5-year survival rate is below 10%.

Differential Diagnosis List: Extranodal nasal natural killer/T-cell lymphoma, Squamous cell carcinoma, Other NHL, Wegener's granulomatosis, Granulomatous infections, Adenoid cystic carcinoma, Olfactory neuroblastoma, Malignant melanoma

Final Diagnosis: Extranodal nasal natural killer/T-cell lymphoma

References:

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Figure 1

a



Description: Heterogeneous mass centred in the left nasal fossa invading ipsilateral orbital cavity and facial soft tissue. **Origin:** Department of Radiology, Complejo Hospitalario de Navarra, Spain.

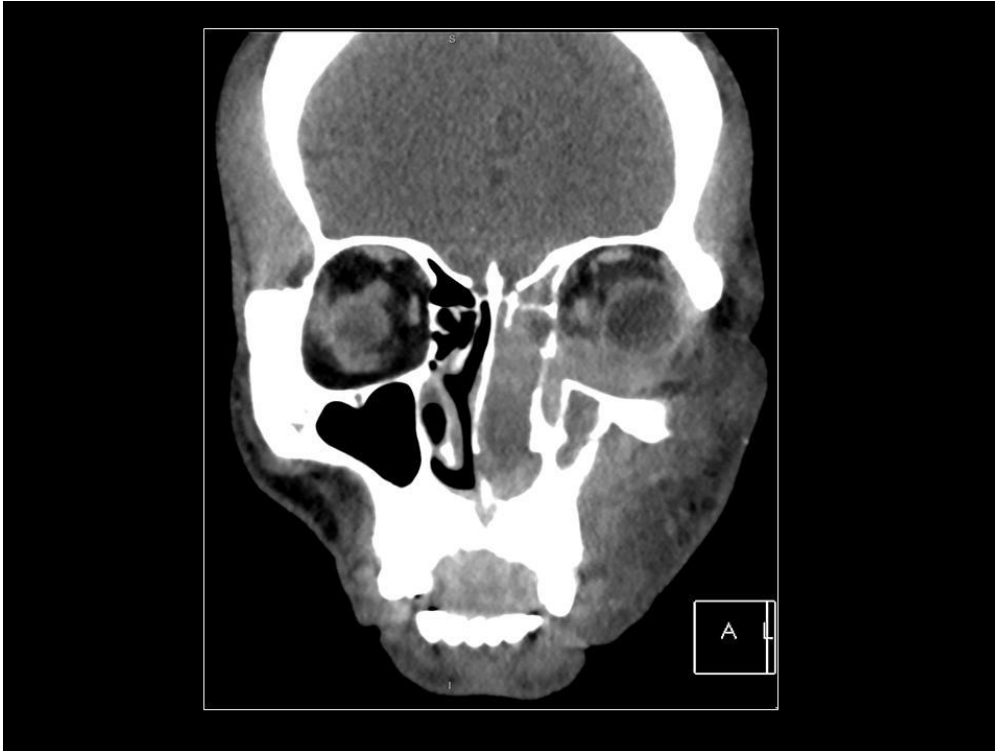
b



Description: Heterogeneous mass centred in the left nasal fossa invading ipsilateral orbital cavity and facial soft tissue. **Origin:** Department of Radiology, Complejo Hospitalario de Navarra, Spain.

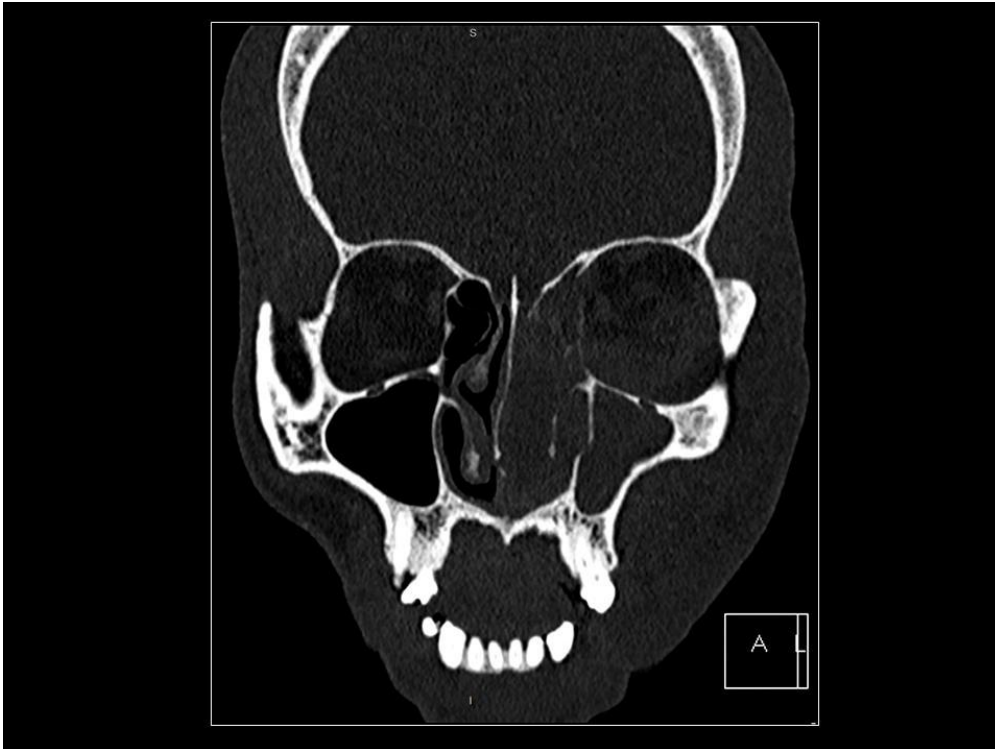
Figure 2

a



Description: Pay attention to the mild bone erosion and orbital invasion associated. **Origin:** Department of Radiology, Complejo Hospitalario de Navarra, Spain.

b



Description: Pay attention to the mild bone erosion and orbital invasion associated. **Origin:** Department of Radiology, Complejo Hospitalario de Navarra, Spain.

Figure 3

a



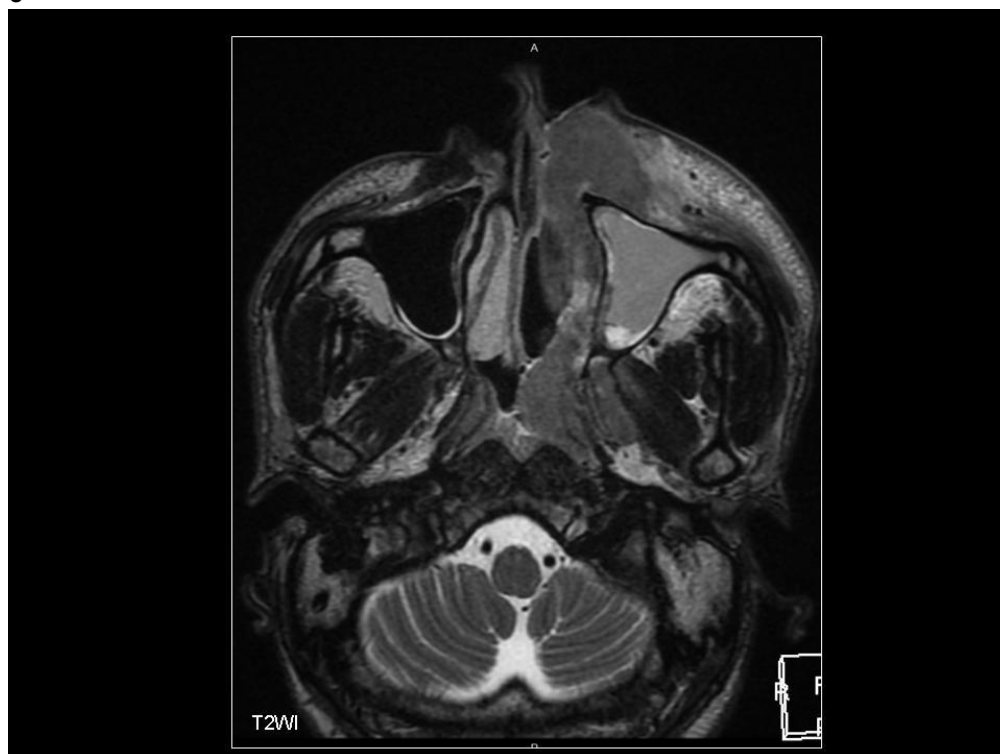
Description: Lesion shows heterogeneous signal in T1 & T2 sequences, restricted diffusion and moderate enhancement with intravenous contrast. **Origin:** Department of Radiology, Complejo Hospitalario de Navarra, Spain.

b



Description: Lesion shows heterogeneous signal in T1 & T2 sequences, restricted diffusion and moderate enhancement with intravenous contrast. **Origin:** Department of Radiology, Complejo Hospitalario de Navarra, Spain.

c



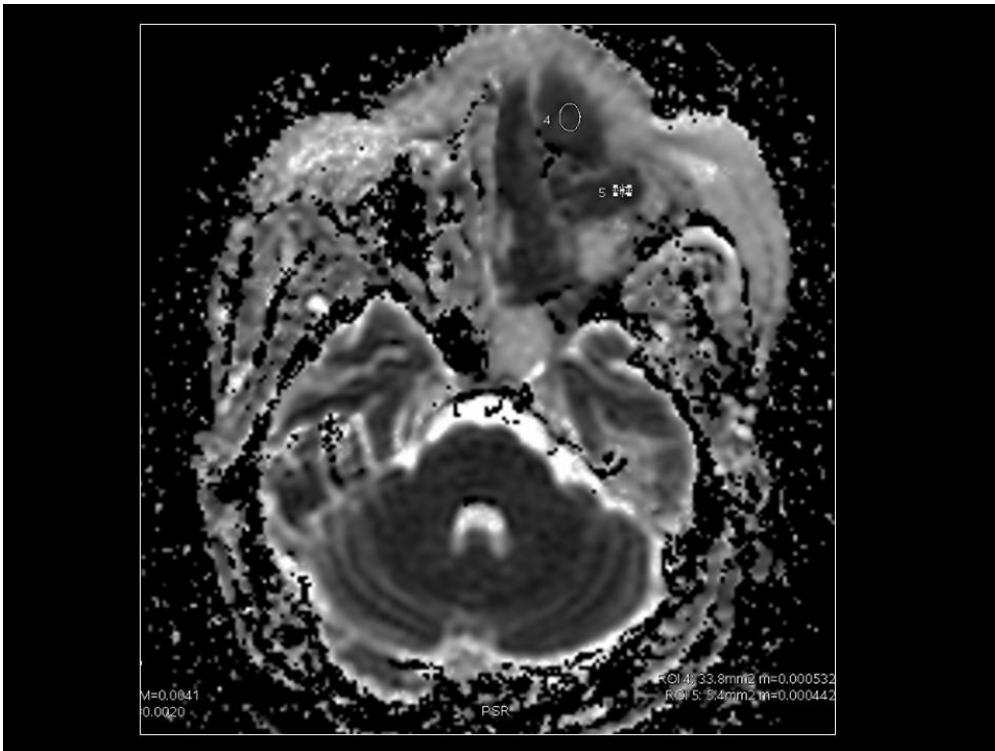
Description: Lesion shows heterogeneous signal in T1 & T2 sequences, restricted diffusion and moderate enhancement with intravenous contrast. **Origin:** Department of Radiology, Complejo Hospitalario de Navarra, Spain.

d



Description: Lesion shows heterogeneous signal in T1 & T2 sequences, restricted diffusion and moderate enhancement with intravenous contrast. **Origin:** Department of Radiology, Complejo Hospitalario de Navarra, Spain.

e



Description: Lesion shows heterogeneous signal in T1 & T2 sequences, restricted diffusion and moderate enhancement with intravenous contrast. **Origin:** Department of Radiology, Complejo Hospitalario de Navarra, Spain.

f



Description: Axial T1 VIBE + IVC. Infiltrating mass centred in the left nasal cavity extending into the left pterygopalatine and temporal fossa. **Origin:** Department of Radiology, Complejo Hospitalario de Navarra, Spain.

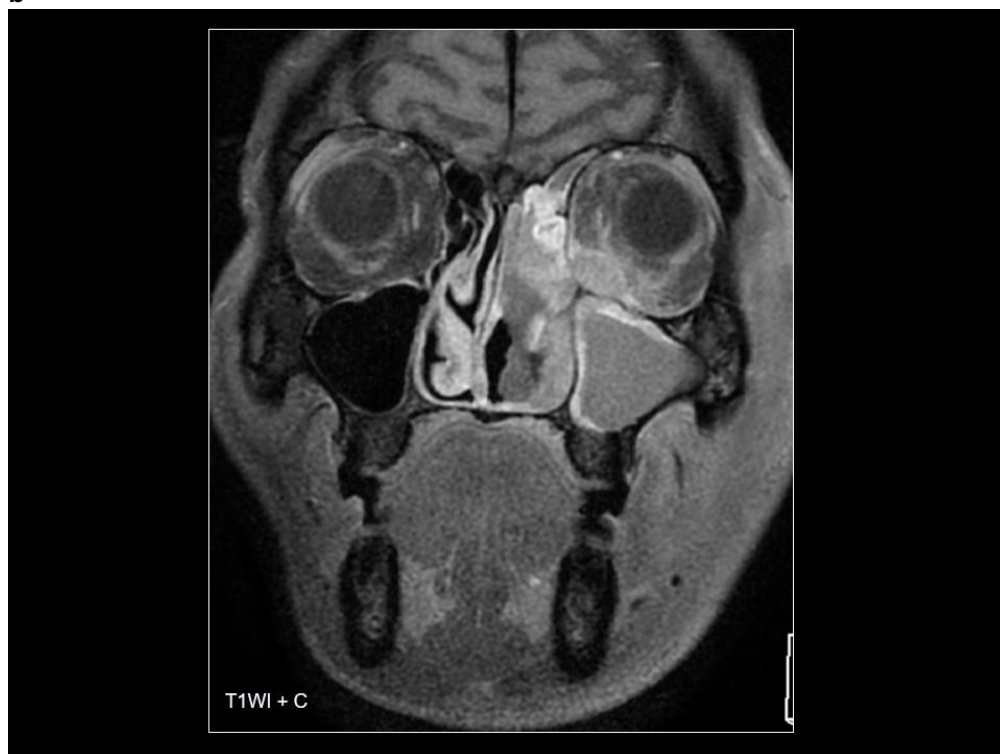
Figure 4

a



Description: Lesion invades left orbital cavity and shows moderate enhancement with intravenous contrast. **Origin:** Department of Radiology, Complejo Hospitalario de Navarra, Spain.

b



Description: Lesion invades left orbital cavity and shows moderate enhancement with intravenous contrast. **Origin:** Department of Radiology, Complejo Hospitalario de Navarra, Spain.

c



Description: Coronal T2WI. See the infiltrating mass arising from the left nasal cavity without intracranial extension. **Origin:** Department of Radiology, Complejo Hospitalario de Navarra, Spain.