

REVIEW ARTICLE

NECROTISING OTITIS EXTERNA: AN OVERVIEW OF IMAGING MODALITIES

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Necrotising Otitis Externa (NOE) has often posed some challenges in view of diagnosis and management by clinicians. One such challenge is the appropriate and timely use of imaging techniques, since its use is critical not only in diagnosis but also determining the extent and resolution of the disease. Hence, doctors in both primary and secondary health care need to be familiar with presenting symptoms, while specialists need to be appraised of advances in imaging techniques in management of NOE. Whilst there is a general consensus amongst clinicians on some aspects of management of NOE, there is very limited consensus on the use of imaging modalities. There is no single modality of imaging that can provide a complete picture of diagnosis, disease progression and resolution. There are some advantages and limitation of each methodology, which indicate that a multi-modal imaging technique at particular stages of the disease may provide better management outcomes. However, further research in this area is required, as there is not yet an established 'gold standard' for imaging in NOE.

Keywords: Necrotising Otitis Externa; Malignant Otitis Externa; Skull base Osteomyelitis; Tc-99m bone scan; Ga-67 bone scan; Indium 111 labelled leukocyte scanning; SPECT

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INTRODUCTION

Necrotising Otitis Externa (NOE), also referred to as Malignant External Otitis, is a rare but severe infection of the external auditory canal and surrounding structures. This disease commonly affects immuno-compromised and elderly persons with Diabetes Mellitus. Given the increasing longevity of patients suffering from diabetes, the incidence of NOE is also on the rise. NOE has often posed some challenges in view of its diagnosis and management by clinicians. One such challenge is the appropriate and timely use of imaging techniques, which is critical not only in diagnosis but also determining the extent and resolution of the disease. Early diagnosis and prompt management by relevant medical teams is key in NOE, which has a high morbidity-mortality rate.¹ Hence, doctors in both primary and secondary health care need to be familiar with presenting symptoms while specialists need to be appraised of advances in imaging techniques in management of NOE.

Background and scope:

The imaging trends with regards to NOE have largely remained unchanged over the years. Whilst there is a general consensus amongst clinicians on some aspects of management of NOE, there is very limited consensus on the best use of imaging modalities.^{1,2} There is no single modality of imaging that can provide a complete picture of diagnosis, disease progression and resolution of NOE.³ Therefore, a

multi-modal approach involving various imaging modalities at particular stages of the disease can provide better management outcomes. However, it is also important to be cognisant of the fact that the use of these imaging modalities is also dependant on availability of resources and other factors around the world.

This article broadly discusses multimodal imaging methodologies, which in turn can help clinicians in making appropriate decisions with regards to timely referrals and management.

What is Necrotising Otitis Externa?

NOE is a rare but severe and aggressive infection of the external auditory canal and surrounding structures. This commonly spreads to involve the periosteum and reaches the bone of the skull base. Once bone involvement is confirmed on radiological findings, it is also referred to as Skull Base Osteomyelitis (SBO). However, SBO is often referred to as a complication of NOE. As mentioned earlier NOE, is also widely known as Malignant Otitis Externa as it mimics a neoplastic condition where the disease spreads and deteriorates rapidly like malignancy.⁴ This is actually a misnomer as the condition is not in reality a neoplastic condition. It is known as NOE due to extensive soft tissue involvement. If untreated, cranial neuropathies of which facial nerve is the most common can develop due to sub-temporal extension of the infection. Early diagnosis and prompt management by relevant medical teams is thus key in NOE, which has a high

morbidity-mortality rate.¹ Hence delays in the diagnosis of NOE can affect disease outcomes significantly.⁵

On clinical examination, granulation tissue is present on the floor of the osseo-cartilaginous junction and otoscopic examination can reveal exposed bone.⁶ Granulation is virtually pathognomonic of NOE except in those with HIV/AIDS where granulation tissue may be absent in the external auditory canal.

Infection begins in the skin and cartilage of the external auditory meatus and spreads rapidly and causes necrosis of the surrounding soft tissues, cartilage and bones by invading them and even spreads to the cranial nerves. NOE is referred to as 'necrotising and invasive' due to the fact the infection invades the adjacent peri-audicular tissues into the cartilage and bones resulting in the necrosis and osteomyelitis of the temporal bone as well as base of the skull.^{7,8}

Spread of the disease outside the external auditory canal occurs rapidly and progressively through the fissures of Santorini and the tympano-mastoid suture to the skull base. Periostitis spreads along the under surface of the skull base to involve the stylo-mastoid foramen and then the jugular and hypoglossal foramina and hence the facial nerve which lies in close proximity here can be affected easily.⁹ The disease can be fatal if treatment is not aggressive and timely, especially if it spreads outside the auditory canal with involvement of the cranial nerves.⁷

The symptoms of NOE are easily recognisable. These include persistent and foul-smelling yellow or green drainage from the ear (otorrhea) accompanied with otalgia that gets worse when moving the head and at night. Other symptoms include hearing loss, persistent itching in the ear canal, fever, difficulty in swallowing and weakness in the facial muscles. Laryngitis may be experienced by those where the infection travels to the larynx. In addition, skin around the ear appears swollen and red.¹⁰

Diagnosis:

Definitive diagnosis of NOE is frequently vague and has a high index of suspicion and involves various laboratory and imaging modalities, and histologic exclusion of malignancy.¹¹

The diagnosis of NOE is mainly based on number of clinical findings, markers such as an elevated erythrocyte sedimentation rate, presence of certain bacteriological pathogens found on laboratory analysis, radiographic evidence of soft tissue with or without bone erosion in the external canal and infra-temporal fossa.^{12,13}

A physical exam including a complete medical history to identify underlying conditions that may compromise the immune system is important. In addition, an ear examination may reveal granulation tissue or drainage which may indicate an infection. A sample is then sent for analysis and the identification of particular pathogens such as *Pseudomonas aeruginosa*, Methicillin resistant *Staphylococcus Aureus* (MRSA), *Staphylococcus epidermidis*, *Proteus*, *Klebsiella*, *Aspergillus fumigatus* and *Candida* species can indicate NOE.¹² Additional tests such as a neurological examination is also carried out. Since NOE has potentially life-threatening complications, imaging studies are strongly recommended.¹⁴ Imaging can play synergistic roles in the management of NOE and it is important that clinicians assess clinical signs and symptoms, radiological imaging and inflammatory markers for desirable disease outcomes.

Imaging modalities:

The modalities related to imaging in NOE have not changed drastically over the years. However, it is important to note that there has also been increased understanding amongst the otolaryngologists or ear, nose and throat (ENT) specialists on the appropriate and timely use of these techniques. Thus, the timely and suitable use of radiologic modalities and management protocol can lead to improved outcomes which in turn can reduce the need for surgical intervention.¹⁵ In addition, it is worth noting, that whilst clinical presentation, diagnosis, and pathophysiology of NOE is well understood, there is still no consensus on the best use of imaging techniques.¹⁶

The availability of latest imaging technology means that clinicians are now able to use more advanced imaging techniques in many parts of the world. Despite the availability of such techniques, NOE continues to pose some challenges such as appropriate imaging modalities in diagnosis and throughout disease progression and follow-up due to its complex nature. Various imaging modalities have their own advantages and limitations which need to be fully understood in order to achieve optimal management outcomes.

As mentioned earlier, NOE has a high suspicion index. Thus, lack of clarity on initial examination and disproportionate symptoms to clinical findings can further delay diagnosis.¹⁷ Often disease progression remains unclear until radiological findings and radio-nucleotide results are obtained. It is therefore imperative that a sound radiological assessment should be supplemented by clinical and serological analysis to establish the disease.¹⁸ Let us

look at the advantages and limitations of imaging methodologies further.

Whilst conventional radiology does not have any use in the diagnosis of NOE, the radiological diagnosis of the disease tends to remain limited largely to computed tomography (CT) scans and magnetic resonance imaging (MRI). In addition, depending on the availability of resources, Technetium-99 methylene diphosphate (Tc-99m) bone scanning, Gallium-67-citrate (Ga-67) bone scanning and Indium 111 labelled leukocyte scanning are also widely used.¹² Whilst CT and MRI are used for anatomical imaging, nuclear techniques are useful in understanding the functional process. In addition, there is a growing trend in the use of hybrid techniques such as positron emission tomography (PET) -CT and more recently PET-MRI which combine anatomical and functional biomarkers and provide a higher level of imaging.¹⁷

While CT scanning allows determining the location and extent of disease, MRI is the method of choice in determining intracranial extension. A limitation for both CT scan and MRI is that active inflammation and resolving infection is hard to differentiate and hence CT and MRI imaging does not necessarily correlate to the prognosis and outcome of the disease. However, bone scintigraphy can detect NOE earlier than any CT or MRI and hence these should be accompanied for the initial diagnosis.¹

The advantage of CT scanning over other modalities is detecting bony erosions and demineralization. In addition, CT findings usually include obliteration of the fat planes in the sub-temporal area and destruction of the bony cortex of the mastoid.¹⁹ CT scan may reveal clinically relevant imaging findings such as a thickened mucosa of the external auditory canal and auricle. In addition, a destructive appearance indicating of the tympanic and mastoid bone strongly suggest NOE.²⁰ This early sign of evolving osteomyelitis, as well as infiltration of the temporo-mandibular fat pad are usually noted as subtle cortical erosions visible on CT scan and indicative of the disease.¹³

MRI is superior to CT scan in detecting anatomical locations and since it has a superior contrast resolution, it can assess soft-tissue involvement in NOE. It is thus regarded the imaging technique of choice.¹⁴ MRI can demonstrate meningeal enhancement better and can reveal intracranial extension of the disease as well and reveal complications such as thrombosis and intracranial spread.²⁰ Adjacent soft tissues demonstrating focal areas of rim enhancing fluid are consistent with abscesses. Posteromedial extension to the jugular foramen or carotid space is absolutely

critical. Retro-condylar fat infiltration is one the earliest changes seen on MRI in a patient suspected of NOE. A pre-treatment MRI can also help identify the initial compartments (anterior, medial, midline and intracranial and extra-cranial) affected by the disease. The areas on high signal intensity of the MRI, will show thickening and infiltration of soft tissue which indicate infection. Follow-up MRIs at intervals of 2–3 months can then help evaluate the response to treatment by comparison with the initial scans. The subsequent MRIs can reveal direction of the spread of disease from each compartment as well as the overall progression.²¹

Ga-67 scan and Tc-99m bone scanning are both important imaging tools in NOE. Ga-67 is a useful tool to monitor the resolution of the disease which can be seen as increased uptake in the affected area. However, in order to get a more accurate picture, the lesion to non-lesion ratio needs be determined.²² Tc-99m can be useful for the initial evaluation of the disease but is not useful in assessing the prognosis of the disease. Tc-99m stays positive for a long period; even after the resolution of the infection.²³ Indium In 111-labelled leukocyte scans on the other hand show similar findings as a Ga-67 scan, it is more specific to an inflammatory process. It is also reliable and a timely indicator of resolution of infection.²⁴

CT and/or MRI can be supplemented by Single-photon emission tomography (SPECT) bone imaging for initial diagnosis of NOE routinely as well as for follow-up of NOE cases. Whilst SPECT can also be useful in assessing the response to disease and recurrence, Ga-67 should be the investigation of choice to assess disease progression.³

CONCLUSION

Imaging techniques with regards to NOE have evolved in recent years, although there are several geographical and resource limitations. It is well established that CT scan, MRI, bone scintigraphy and leukocyte imaging provide an adequate picture of diagnosis, spread of disease, impact of treatment and disease regression. However as highlighted earlier, each modality has its advantages and limitations.

For example, as various studies establish, CT scan is useful in early stages and also shows the extent of bone erosion and demonstrates the progression of bony disease. However, it cannot be used to follow resolution in cases where there is a possible central skull base osteomyelitis or analyse the impact of treatment. Similarly, MRI is superior to CT scan in detecting anatomical locations and since it has a superior contrast resolution, whilst radionuclide scans can provide better information on the overall spread of inflammation and soft tissue involvement.

CT and MRI are both used for anatomical imaging, and nuclear techniques aid in functional process imaging. Hybrid techniques PET-CT and PET-MRI are the newest modalities which combine imaging strengths.¹ No single modality is able to address the scope of NOE and whilst clinicians have increased reliance on CT scans and MR-I in the initial diagnosis and some aspects of follow-up of NOE, a combination of modalities and their stage-specific use remains critical for positive disease outcomes there continues to be no "gold standard" for establishing disease resolution.

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