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Case Report

Skull base non-Hodgkin's lymphoma

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ABSTRACT

Primary skull base lymphoma accounts for 1% - 2% of all skull base tumours. It is a very rare condition, which poses a diagnostic challenge in clinical practice but needs early diagnosis and treatment. We present a case of primary lymphoma of the skull base in a 53-year-old woman, who was admitted with complaints of bilateral temporal pain, facial numbness, slurred speech, difficulty in swallowing and deafness. Computed tomography (CT) and Magnetic Resonance Imaging (MRI) showed ill-defined destruction of the petrous temporal bone with a high signal area noted on T2 weighted images in the right temporal lobe which initially was thought to be skull base osteomyelitis. However, a finding of a thin subperiosteal dense soft tissue in the left parieto-occipital region with intact adjacent bone cortex similar in appearance to the of the primary skull base pathology was seen which alerted the team to possible diagnosis of skull base lymphoma.

Keywords: Central skull base lymphoma, Primary malignant non-Hodgkin's lymphomas of skull base, Primary non-Hodgkin's lymphoma of the skull base

INTRODUCTION

Non-Hodgkin's lymphoma can involve any structure in the head and neck region; however primary malignant non-Hodgkin's lymphoma of the bone is rare, accounting for 1-2% of all malignant lymphomas. It is extremely rare to be found in the skull base.[1,2]

CASE REPORT

A 53-year old female was initially referred by her General Practioner with complaints of bilateral temporal pain, facial numbness, slurred speech, difficulty swallowing and deafness 2 weeks after dental extraction and crown repair secondary to a broken tooth. The reported symptoms started after the dental procedure and were worsening over days before admission. The patient was well in herself prior to the dental procedure and had no symptoms. Her history included asthma, cervical cancer, hypertension, COPD and endometrial adenocarcinoma that was treated with surgery.

On examination she had reduced sensation over the left side of face, but no other cranial nerve deficits noted. CT head was unremarkable. As her symptoms persisted, an MRI head was organized. This showed no evidence of acute intracranial pathology, however significant inflammatory changes were noted in the mastoid air cells, maxillary antra bilaterally with near total opacification of the left maxillary antrum and ethmoid air cells. The inflammatory changes in paranasal sinuses and mastoid air cells could explain the deafness and facial pain experienced by the patient however the rest of her symptoms were unexplained. Review by neurology and otorhinolaryngology teams revealed no acute issues. Impression after neurology team review was that

symptoms were due to nerve damage post tooth extraction and she was subsequently discharged home. She then presented 2 weeks later with haematemesis and melena, diagnosed and managed as Upper Gastro-Intestinal bleed. While in hospital she reported symptoms from her previous admission to be worsening. Cranial nerve Examination revealed palsy of the right 5th, right 7th and 10th cranial nerves.

Given her worsening symptoms and cranial nerve palsies, another MRI head was organized. The MRI head (Figure 1) was limited as it was not completed due to patient discomfort, but it still showed diffuse mucosal thickening in the paranasal and mastoid sinuses with a high signal area noted on T2 weighted images in the right temporal lobe. This area was suspicious for underlying mass lesion, inflammatory change such as encephalitis or early abscess formation.

As the MRI picked up new findings not seen on the previous scan, a discussion with a consultant radiologist was had to compare the images and see if the findings were indeed new. On review of the previous images, some changes were seen in her skull base that raised the suspicion of skull base osteomyelitis. On further review of all the MRI images, a finding of an ill-defined destruction of the anterior middle skull base extending into the parapharyngeal spaces and pterygoid muscles was seen. Also seen was a suspected bony breach in the right temporal region which was leading to right sided temporal lobe oedema. As the patient's symptoms developed after a dental procedure, initial impression was made of features consistent with that of skull base osteomyelitis.

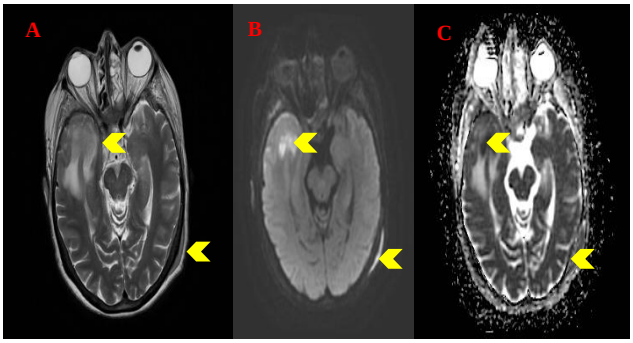

Figure 1: A) Axial T2WI, B) Axial diffusion C) ADC Map. Initial MRI-Right temporal brain lesion and Left parietal bone lesion also showing restricted diffusion.

MRI head was repeated (Figure 2) that showed increased bulk involving bilateral pterygoid muscles, masticator space and parapharyngeal region, which was likely an abscess. It also showed mottled persistence of the previously seen right temporal lobe oedema and destruction of central skull base, basi-sphenoid, both sphenoid wings which were in keeping with osteomyelitis. Also seen was an extension of inflammatory phlegmon on the right side into the deep lobe of parotid gland abutting the retromandibular vein which would explain the patient's right 7th nerve palsy. She received antibiotics while being further investigated with no improvement of symptoms.

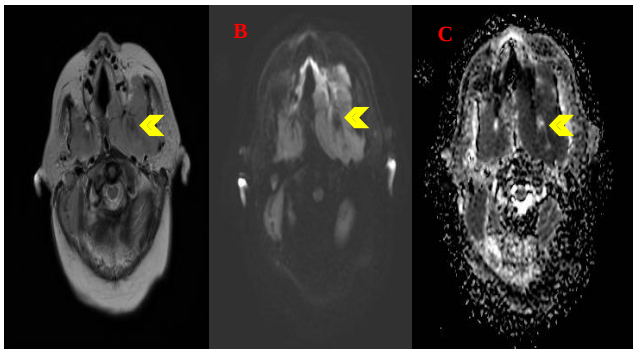

Figure 2: A) Axial T2WI, B) Axial diffusion, C) ADC Map Bilateral bulky pterygoid muscles showing restricted diffusion.

A follow up CT head and neck was done as she did not tolerate MRI scan. Images (Figures 3,4,5,6) showed extensive skull base sclerosis with cortical thickening involving sphenoid wing, basi-sphenoid and the pterygoid plates extending into the left maxillary sinus walls. bulky inflammatory phlegmon with abscess formation in both Parapharyngeal regions and the masticator space was seen that was significantly narrowing the nasopharyngeal airway.

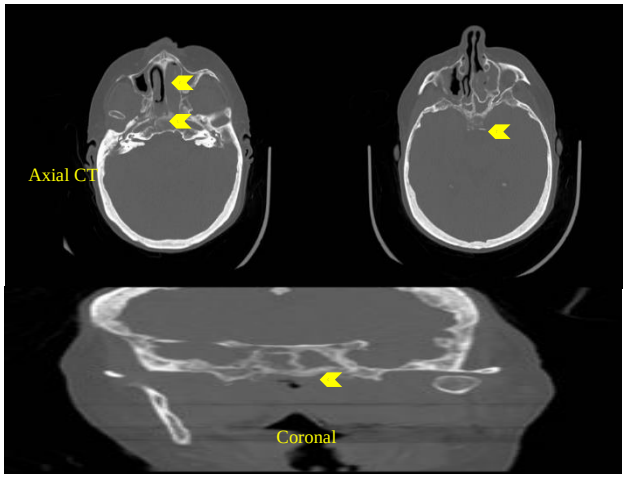

Figure 3: Left nasal cavity mass with skull base sclerosis.

Retained secretions were also seen in bilateral mastoids due to Eustachian tube dysfunction caused by naos-pharyngeal oedema. this explained her deafness. New findings of enlarged bilateral level II lymph nodes were noted along with persistence of the right temporal lobe oedema with extension of inflammatory changes into both the cavernous sinuses with arteries still patent.

Features were all still in keeping with a diagnosis of skull base osteomyelitis.

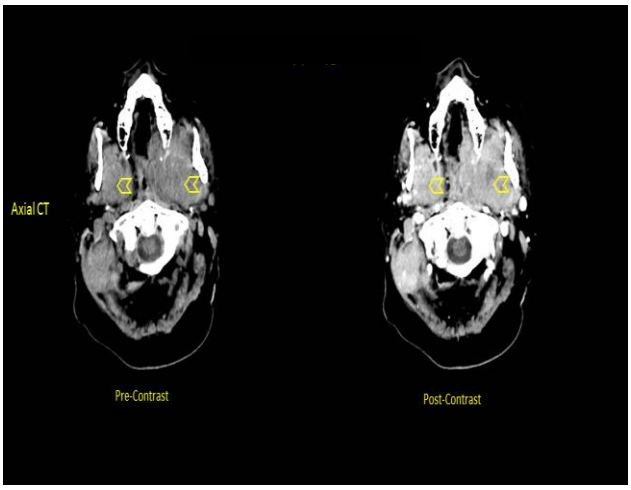

Figure 4: Bilateral bulky bilateral bulky pterygoid muscles.

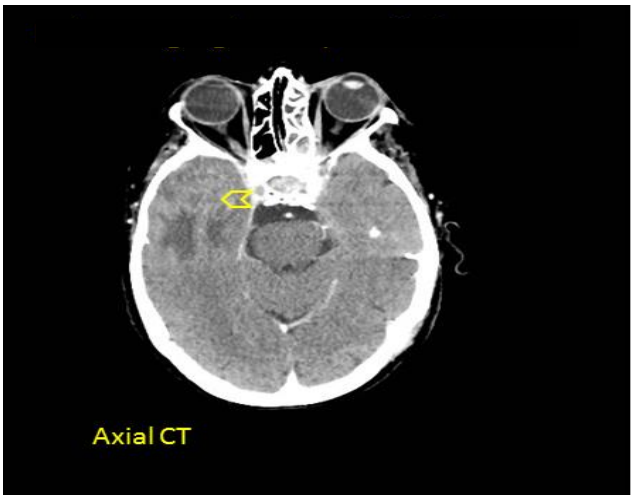

Figure 5: Axial CT Enhancing right temporal lobe mass.

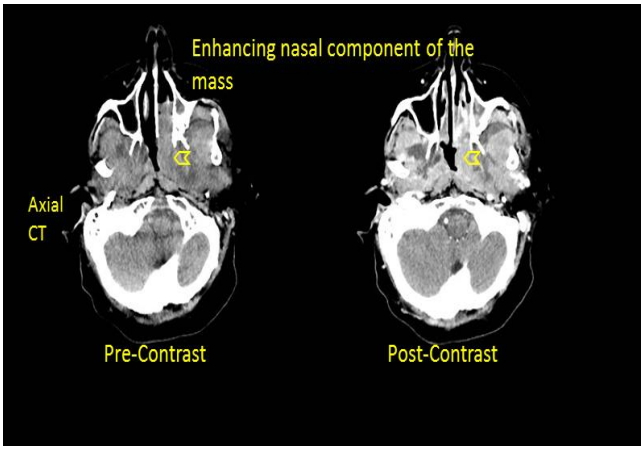

Figure 6: Enhancing nasal component of the mass.

What was peculiar however was a thin subperiosteal dense soft tissue in the left parieto-occipital region seen with intact adjacent bone cortex. Appearance of this lesion was similar to that of the primary skull base pathology on all MRI sequences. This finding highly raised the possibility of main diagnosis of a primary skull base lymphoma. This lesion was actually present in the previous MRI after review of all images (Figure 4).

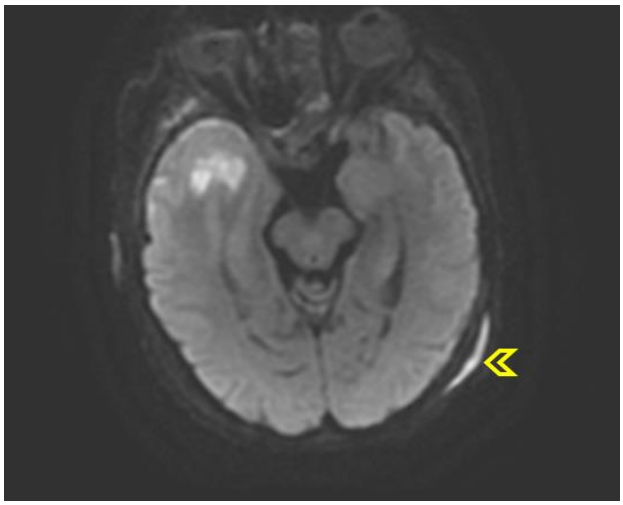

Figure 7: Sub periosteal soft tissue in the left parieto-occipital region seen with intact adjacent bone cortex.

No neurosurgical intervention was advised by the neurosurgery team after discussion. Unfortunately, she deteriorated despite all medical care and was subsequently taken to theatre by the ENT team to have functional endoscopic sinus surgery mainly for tissue biopsies and culture. Intraoperatively a small left middle meatal polyp was seen, removed and sent for histology. Histology confirmed diagnosis of Diffuse Large B-cell Lymphoma (DLBCL). As the histology confirmed diagnosis of lymphoma, staging CT chest, abdomen and pelvis was organized (Figures 8 and 9).

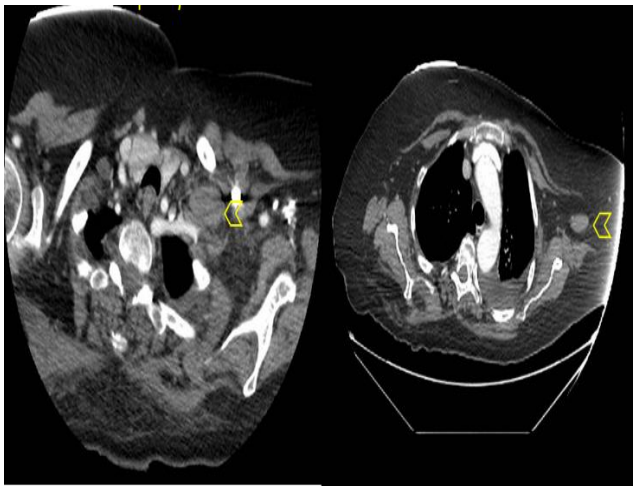

Figure 8: Axial CT Thorax showing left supraclavicular and axillary adenopathy.

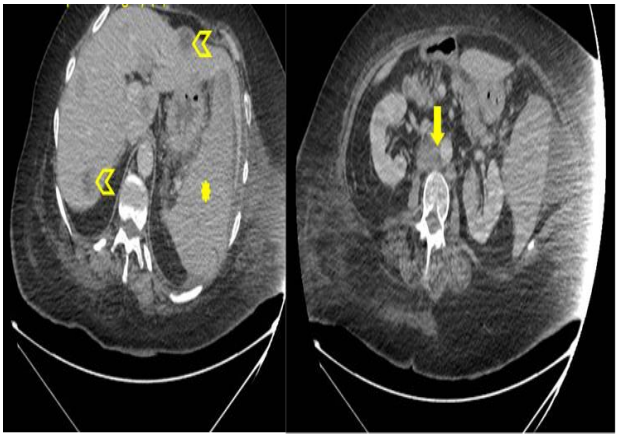

Figure 9: Axial CT Abdomen showing Liver lesions, aortocaval nodes (solid arrow) and splenomegaly (*).

This showed lymphadenopathy in left level II, left supraclavicular, left axillary, retroperitoneal, para-aortic, bilateral iliac and right groin regions. Enlargement of the liver and spleen with numerous focal liver lesions and few poorly enhancing lesions in kidneys and a left adrenal nodule were also seen. She was discussed in the haematology Multi-Disciplinary Team Meeting (MDT) and it was felt that given the advanced stage of DLBCL, CNS involvement and poor performance status she would not be fit for chemotherapy. Decision was made to treat her palliatively and be kept comfortable. End of life care pathway was discussed with her family who were in agreement of the decision. She was transferred to a hospice with end of life package where she passed away 2 days later.

DISCUSSION

Primary malignant non-Hodgkin's lymphoma of the bone is a rare entity and accounts for 1-2% of all malignant lymphomas with occurrence in the skull base being extremely rare. [2] Though this is a rare diagnosis, there has been increasing number of cases reported suggesting improved diagnostic abilities and practice.[3] Literature has shown that the most common subtype of primary malignant non-Hodgkin's lymphoma that primarily involves the central nervous system is Large B-cell lymphomas. This subtype is also shown as most commonly involved in studies that looked.[4]

Clinical picture of patients with primary skull base lymphoma has been shown to be similar to intracranial infection with multiple cranial nerve palsies, extradural abscesses with osteomyelitis.[5,6] This stays true to our case as she had multiple cranial nerve palsies and presented with features of intracranial infection with radiological images suggesting skull base osteomyelitis.

Modern imaging techniques play a vital role in the diagnosis of skull base tumours. Both CT and MRI are essential for the complete assessment of skull base lesions. MRI with gadolinium has been stated to be considered the imaging method of choice for the diagnosis of intracranial lymphoma.[5] Han et al. demonstrated the MRI characteristics of non-Hodgkin's lymphoma of skull base and noted MRI findings of an erosive lesion of the skull base, invasion of the cavernous sinus without arterial narrowing, infiltration along the Dural surface and iso or hypo intensity within the brain on T2 weighted imaging should suggest lymphoma.[7] Similar features were seen in another study by Choi et al.[1] These features are also consistent with what was seen on the MRI images of our patient.

Another study by Ogden et al, looking at MRI diffusion features of skull base osteomyelitis compared with skull base malignancy, showed the apparent diffusion coefficient values in patients with skull base osteomyelitis were higher than the apparent diffusion coefficient values of patients with lymphoma which can be a key tool to differentiate the two conditions as they appear similar on conventional sequences.[8] This can help prevent delay in the diagnosis and treatment of both conditions. Our patient did have MRI with diffusion weighted imaging that showed restricted diffusion on sequences, which are typical of lymphomas and in keeping with findings detailed in the previously mentioned study.

Radiological imaging is therefore invaluable in assessing and diagnosing skull-based lymphomas. However, the definite diagnosis is histological after tissue sample is obtained surgically. Differential diagnosis of skull base lesions includes meningioma, metastasis, skull base osteomyelitis, nasopharyngeal carcinoma, granulomatous diseases, tuberculosis and fungal infections.

Surgical options to facilitate diagnosis include open surgery, fine needle aspiration, ultrasound guided needle biopsy and endoscopic skull base surgery, which are to be considered in selected cases. If lymphoma is confirmed, the recommended treatment option is systemic chemotherapy.[9] Chemotherapy would have been the primary treatment in our patient if she was fit and able to tolerate it. Diffuse B-cell lymphoma is rapidly fatal because of its aggressive progression if left untreated.[10]

CONCLUSION

Diffuse large B-cell lymphoma (DLBCL) of the skull base is thankfully rare but is associated with significant morbidity and also mortality as in our case. It should be considered along with skull base osteomyelitis and invasive fungal infections in patients being investigated for skull base pathologies with all suspected cases undergoing MRI head with diffusion.

Early intervention mostly for tissue biopsies and cultures is of foremost importance to establish the diagnosis which facilitates commencement of prompt treatment that can be lifesaving.

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