Advanced nasopharyngeal carcinoma in a teenager presenting with neck abscess

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ABSTRACT

Nasopharyngeal carcinoma (NPC) is the second most common cancer in Malaysian man, after lung cancer. It commonly manifests with cervical lymphadenopathy, nasal symptoms, aural dysfunction, headache, diplopia, facial numbness, weight loss and trismus. We reported a rare case of a teenage male who presented with right neck abscess as the sole manifestation of advanced NPC. Failure to improve along with development of other symptoms after adequate treatment for neck abscess led to a diagnosis of advanced NPC. This case illustrates a rare presentation of NPC.

Keywords: Nasopharyngeal carcinoma, abscess, tumour, head and neck

INTRODUCTION

Nasopharyngeal carcinoma (NPC) is common in Southeast Asian countries. In Malaysia, the incidence is 25 cases per 100,000 population. NPC is the most common head and neck cancer after lung cancer in men. [1] Malaysian Chinese men has the second highest incidence in comparison to the rest of the world. [1] The incidence rises with age, particularly in those between the age of 50 and 60.

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CASE REPORT

An 18-year-old Malay man presented with right lateral neck swelling for two weeks. It had increased in size and was associated with intermittent throbbing pain and low grade fever. He also experienced odynophagia and trismus. He denied any epistaxis, nasal blockage and ear symptoms. There was no previous history of any dental problem and trauma. There was no history of contact with pulmonary tuberculosis patient.

Examination revealed a right neck swelling that extended from the mastoid region up to the mid sub-occipital region, measuring about 8 x 8cm. It was warm, tender and

firm. Multiple neck cervical lymphadenopathies in the right side at levels I, II and V were also palpable. Nasoendoscopic examination revealed a normal Fossa of Rosenmuller. Oropharynx examination and laryngoscopy were normal.

A computed tomography (CT) scan of neck obtained showed a diffuse swelling in the right side of the neck which extended from the level of the right ear pinna until C6 with associated with ill-defined hypodense lesion seen medial to the right sternocleidomastoid. However, there was no peripheral enhancement seen. Presence of multiple rounded minimally enhancing lesions with adjacent hypodense centre were consistent with enlarged node with necrotic centres. The nasopharynx including FOR were normal. Part of the brain that was visualised brain was normal. The features were suggestive of inflammatory changes with early abscess formation and multiple lymphadenitis.

The patient was empirically started on intravenous Cefuroxime and Metronidazole for suspected neck abscess. After one week on treatment, he developed a right diplopia. A repeat CT scan of the neck revealed previously seen ill defined lesion in the right anterior cervical and right submandibular region showed as irregular multi-septated peripherally enhancing lesion with central hypodensity, in keeping with abscess formation. Medially the lesion extended into the right parapharyngeal space and showed loss of fat plane in the right oropharyngeal wall. Superiorly heterogenous enhancement and bulky appearance of the right prevertebral muscle in the nasopharynx with obliteration of the right Fossa of Rosenmuller were suggestive of extension with loss of fat plane with the right medial pterygoid muscle. Thinning and erosion of apex of the right petrous temporal bone causing intracranial extension forming an extradural lesion in the posterior right temporal region was noted. There was also suspicion of involvement of the right cavernous sinus. The visualised orbits were normal. Inferiorly the lesion extended to the level of the C6 vertebrae. The impression was a right anterior neck abscess with local and intracranial extension (Figure 1).

Referral to ophthalmologist confirmed the right third and sixth cranial nerve palsy most likely secondary to extension of anterior neck and submandibular abscess. Incision and drainage was performed. Intra-operatively 25 cc pus was drained. Pus was sent for acid fast

Fig. 1: The axial computed tomography of the neck showed an irregular multiseptated peripherally enhancing lesion with central hypodensity, consistent with abscess formation in right anterior and right submandibular region.

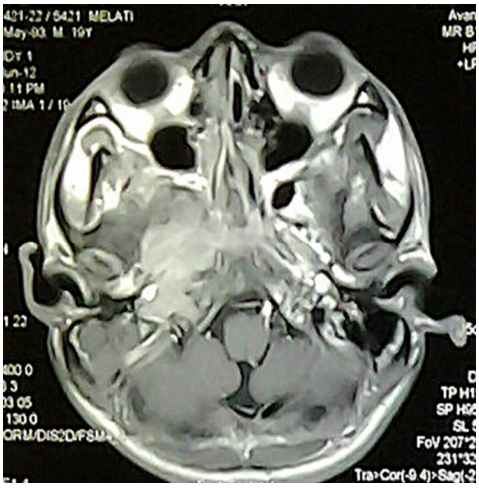

Fig. 2: Magnetic resonance imaging showing an enhancing extradural lesion in the right temporal region. The lesion is seen to encase the petrous portion of right internal carotid artery without involvement of the cavernous sinus and cavernous portion of right ICA are preserved.

bacilli (AFB) and culture and sensitivity. However no abscess wall sent for HPE.

Post-operatively despite two weeks of antibiotic with the neck mass regressing, the patient had persistent diplopia. Magnetic resonance imaging (MRI) showed the previously enhancing extradural lesion in the right temporal region to be unchanged (Figure 2). The lesion is seen encasing the petrous portion of the right internal carotid artery (ICA) with close proximity to the inferolateral wall of the right cavernous sinus. However, both the cavernous sinus and the cavernous portion of the right ICA were preserved. The impression was features of intracranial extension with encasement of the right petrous portion of ICA. No evidence of cavernous sinus thrombosis. Expert opinion was sought from neurosurgical department regarding patient's further management.

Further investigation was done simultaneously; FNAC of the lymph node at level V right side was taken. Patient condition deteriorated. He had projectile vomiting and headache of which necessitated an urgent CT brain. It showed worsening intracranial extension with evidence of right sigmoid sinus thrombosis. Heparin infusion was started. A fine needle aspiration cytology (FNAC) of the neck lymph node revealed metastatic poorly differentiated carcinoma. Deep biopsy of the tissue from right FOR was taken even though no obvious lesion seen. It revealed an undifferentiated, non keratinising squamous cell carcinoma (WHO type III). CT scan abdomen pelvis showed multiple lesions in the liver. The final diagnosis was NPC stage IVC ($T_4N_2M_1$). The oncology team planned the patient for chemoradiotherapy. However, the patient and family refused further intervention and treatment and he was managed palliatively.

DISCUSSION

The provisional diagnosis of a sudden onset in the neck in a young healthy patient is a neck abscess. NPC presenting with acute features of neck abscess is very uncommon, especially when the nasoendoscopy and imaging fail to reveal any lesion in the Fossa of Rosenmuller. Multiple cervical lymphadenopathies is seen in abscesses. In NPC, lymphadenopathy is generally a common complaint that brings a patient to the medical attention. Most of these patients will have a benign, self-limiting process. However, some patient with serious systemic disease or malignancy may present with cervical lymphadenopathy, though the number is by far is small. Out of 2,556 patients in a Dutch study, who presented with unexplained

lymphadenopathy to their family physicians, 256 (10%) were referred to a subspecialist and 82 (3.2%) required a biopsy, but only 29 (1.1%) had a malignancy. [2]

Patients with NPC usually present either with mass in the nasopharynx, Eustachian tube dysfunction, fifth and sixth cranial nerve palsy and neck masses.[3] A retrospective analysis of 4768 patients showed most common symptoms of NPC as neck masses (76%), followed by nasal symptoms (73%), aural dysfunction (62%), headache (35%), diplopia (11%), facial numbness (8%), weight loss (7%) and trismus (3%). [3]

Neck abscess as the only clinical manifestation of NPC is uncommon and rarely reported in literature. To our knowledge, only six cases of NPC have been reported presented as neck abcess. [4] Neck abscess can occur as a result of infection to the cervical lympadenopathy. The pathophysiology of head and neck cancers presenting as neck abscess is unclear. It has been postulated abscess-forming bacteria in lymph nodes are probably derived from an infected ulcer of the primary tumor. [5] The organism usually found in such tumour abscess is *Staphylococcus aureus.*[5] Thus, a biopsy specimen from the wall of abscess during drainage was diagnostic for cancer. [6] Unfortunately, we did not manage to send the abscess wall for HPE in this patient.

Rare and nonspecific presentation of NPC causes diagnostic and therapeutic challenges, causing significant spread of the disease prior to diagnosis. [7] This patient had developed intracranial extension complicated with sigmoid sinus thrombosis. There are no specific clinical features for sigmoid sinus thrombosis. [8] The presenting symptoms varies from headache to mental status changes. [9] The infection spread to the dura causing damage to the intima of blood vessels. These factors, hypercoagulation and decreased blood flow further leading to formation of thrombus within the vessels. [10] Mural thrombus may enlarge to occlude the sinus lumen completely. Organism may invade the thrombus causing intrasinus abscess that may release infected emboli into blood stream causing septicemia. The thrombus may also spread to confluence of sinuses and to superior sagittal sinus or cavernous sinus. [11]

Radiotherapy alone is well accepted for the management for early stage (I and II) NPC. Several studies have reported combination of chemotherapy with radiotherapy for the management of locoregional advanced NPC. [3] The first study, the Intergroup 1997 study showed that chemotherapy with radiotherapy improved overall survival compared with radiotherapy alone. [11] The conclusion from the three approaches tested (neoadjuvant, concurrent and adjuvant chemotherapy), showed that concurrent chemoradiotherapy is the most efficacious. However, the administration of chemotherapy adjunctive to radiotherapy for stage III and IV remained a controversial issue due to conflicting reports in the literature. [11]

In term of treatment outcome, WHO type 1 NPC is considered to be less radiosensitive than types 2 and 3. Therefore, WHO type 1 NPC is associated with the worst prognosis. Despite the use of concurrent chemoradiotherapy, presence of distant metastases remains a major cause of treatment failure, and the prognosis for stage IV patients remain poor. [12]

The ten-year relapse-free survivals following radiotherapy range from 70% to 80% for those with well-localised T_1 nasopharyngeal cancer to 0% to 10% for those with T_4 carcinomas and cranial nerve involvement. [13]

In conclusion, a thorough history and complete physical examination of the head and neck region, with addition of a biopsy specimen from the wall of abscess during drainage was diagnostic for cancer must be performed for patient with neck infection to enable early detection and immediate treatment of any underlying malignancy. We should have high index of suspicion in managing the patient especially in endemic region as NPC is endemic in Southeast Asia and can have various clinical presentation.

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