An Unusual Massive Swelling of Neck Mimicking Parotid Tumor - A Case Report

Akhilanand Chaurasia
Department of Oral Medicine and Radiology, Faculty of Dental Sciences,
King George Medical University Lucknow, UP, India.
Email: chaurasiaakhilanand49@gmail.com

ABSTRACT

Lymphomas are the second most common malignancies affecting the head and neck region. Hodgkin lymphoma is one of the most frequent lymphomas in the Western world. The Hodgkin's lymphoma is primarily malignancy of peripheral lymph node however it can involve liver, lung and bone marrow also. The lymphadenoppathy is preceded by constitutional symptoms also known as "B-symptoms" like fever, night sweats, unexplained pain and weight loss in 40% cases. Detection of Hodgkin's lymphoma is usually incidental in nature. They can be found while evaluation of patient by imaging procedures performed for assessment of other conditions and sometimes by patient itself. However they can be also be detected while evaluating patients for nonspecific symptoms like fever, fatigue, unexplained pain and weight loss. Here we are reporting a case of massive Hodgkin's lymphoma without any paraneoplastic symptoms.

Keywords: *Lymphoma, Lymphadenopathy, Paraneoplastic syndrome*

INTRODUCTION

Hodgkin's disease also known as Hodgkin's lymphoma is a malignant solid tumor that arises from B lymphocytes and less commonly from T lymphocyte which was first described in 1832[1]. Broadly hodgkin's lymphoma is classified into classic Hodgkin lymphoma and lymphocyte predominant variant. The classic hodgkin lymphoma is divided into four subtypes -nodular sclerosis type, mixed cellularity type, lymphocyte rich type and lymphocyte depleted[2]. The onset of Hodgkin's lymphoma follows a bimodal peak of incidence with a first peak in 3rd decade and second peak in 5th decade of life. Men are more predilected for Hodgkin's lymphoma than women among all subtypes[3]. The nodular-sclerosis subtype is most common subtype of Hodgkin's lymphoma affecting young adults however the mixed cellularity subtype is more common in older age group[3]. Clinically Hodgkin's Lymphoma is characterized by asymptomatic painless, rubbery, matted or discrete superficial lymphadenopathy in the neck and supraclavicular areas followed by constitutional symptoms or "B-symptoms" like fever, night sweats and weight loss [4].In more than 50% cases, Hodgkin's Lymphoma most commonly involves cervical, supraclavicular and mediastinal lymph nodes followed by sub-diaphragmatic nodes however the involvement of epitrochlear nodes, Waldeyer's ring, testicular and gastrointestinal lymph nodes are rare and uncommon[4]. The individuals affected by Hodgkin's Lymphoma have three-to nine-fold increased risk of development of Hodgkin's Lymphoma in family members[5]. The clinical evaluation of patients of Hodgkin's lymphoma include thorough medical history, complete physical exam, blood tests, lymph node biopsy, computed tomography and positron emission tomography scans. Staging is carried out according to the Ann Arbor system[6].Clinically Hodgkin's lymphoma have many secondary features like cerebellar degeneration, acute inflammatory demyelinating polyneuropathy, chorea and ataxia, subacute sensory neuropathy, motor neuron disease, myasthenia gravis and brachial neuropathy[7]. The characteristic Reed-Stenberg cells, a type of multinucleated giant cells are found in Hodgkin's lymphoma histologically[6]. The recommended treatment consists of chemotherapy in combination with doxorubicin, bleomycin, vinblastine and dacarbazine. It is the standard regimen for individuals suffering with Hodgkin lymphoma[8, 9].

CASE REPORT

A 48-year-old male **(fig.1)** patient reported with chief complaint of a swelling on the left side of the face and neck since the past 3 years and it was causing severe facial asymmetry and disfigurement. The patient has noticed this swelling 3 years back in left parotid region when it was pea size. Since then the swelling was progressively increasing in size up to present form. The swelling was approximately 20x20 cm in size and have irregular shape. There was no history bleeding or pain associated with swelling. There was no associated difficulty except facial asymmetry. There was no significant past medical and dental history. The patient was well built and had normal gait and posture. He was well oriented to surroundings and had normal intelligence. There was no sign of pallor, cyanosis and generalized lymphadenoppathy. His vitals were within normal limit. On extra-oral examination an irregular massive swelling of approx. 20x20 cm was noted in left side of face and neck extending from the left parotid region to the left supraclavicular region anteriorly **(Fig. 1 & Fig. 2).** Posteriorly the swelling was

involving left mastoid region and occipital region (nape of neck) and extending till left supraclavicular region involving whole length of left side of neck **(Fig. 3 & Fig. 4).** Superiorly swelling was involving left mastoid and parotid region and extending till whole length of body of the mandible till submental region **(Fig. 3).** Inferiorly swelling was involving left supraclavicular region in its full length. On palpation swelling was found to be non tender, firm to hard in consistency, non fluctuant, non-compressible and non reducible. There was no local rise in temperature. The overlying skin over swelling was smooth and erythematous at few places. Slight tenderness was noted at apex of swelling. The left ear lobe was elevated **(Fig.2).** On intraoral examination no abnormality was detected related to swelling. There was black staining on teeth **(Fig.5 and Fig.6).** On the basis of clinical finding a provisional diagnosis of parotid gland tumor has been reached. The patient cannot be advised for routine conventional radiography as swelling was so massive that patient cannot be positioned properly. So patient was advised for CT with contrast and referred to radiodiagnosis department. On the CT examination an ill-defined, heterogeneous enhancing soft tissue lesion (measuring 11.1x18.6x16.7 cm) with necrotic areas is seen in the left cervical region. Superiorly the lesion extends up to the level of the mastoid process and external auditory canal with upward displacement of the parotid gland. Inferiorly the lesion extends to the supraclavicular region with compression of the left subclavian artery and vein. Medially the lesion abutts and displaces the left lobe of the thyroid. The lesion extends into the submandibular region with compression and infiltration of the submandibular gland. The interface with the masseter and medial pterygoid muscles is ill-defined. The left common carotid artery is medially displaced by the lesion with encasement of internal and external branches and the left internal jugular vein is compressed. The left sternocleidomastoid muscle is infiltrated by the lesion and the interface with trapezius and splenius capitis muscles is ill-defined. Posteriorly the lesion is extending into paravertebral soft tissues **(Fig.7,8,9,10,11)**. The patient is further gone to routine blood investigation and incisional biopsy was done in department of surgical oncology. Microscopically **(Fig.12, Fig.13)** scattered large malignant cells known as Reed-Sternberg cells admixed with a reactive cell infiltrate composed of lymphocytes, histiocytes, and plasma cells(40X) (haematoxyline and eosin staining). On the basis of radiopathological correlation a final diagnosis of Hodgkin's Lymphoma has reached.

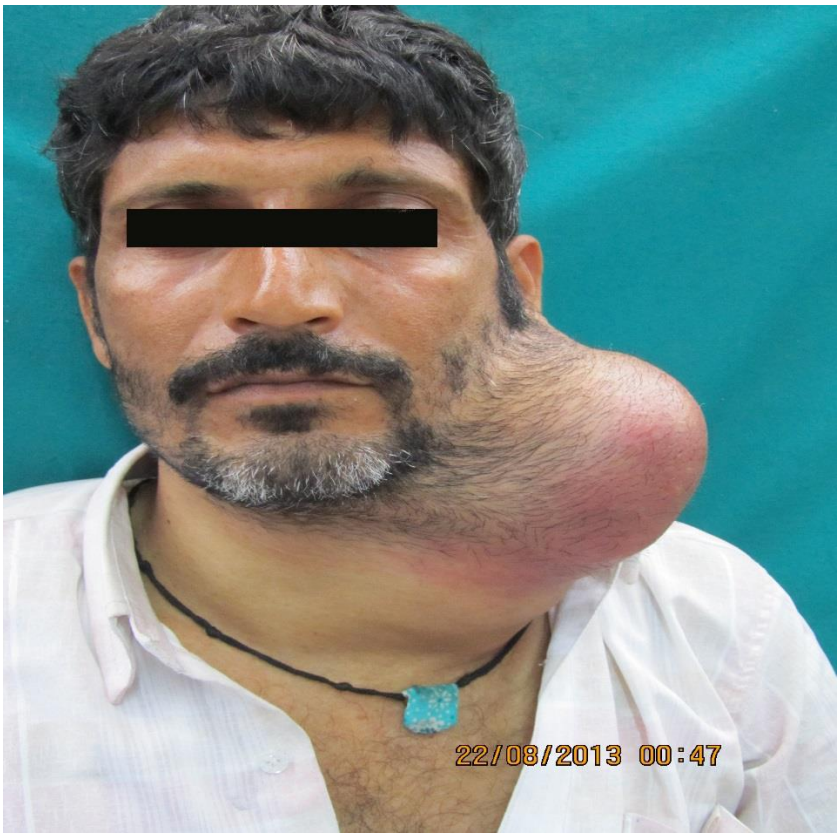


Fig. 1: Photograph of patient showing a swelling on the left side of the face and neck. Extending from the left parotid region to the left supraclavicular region anteriorly.

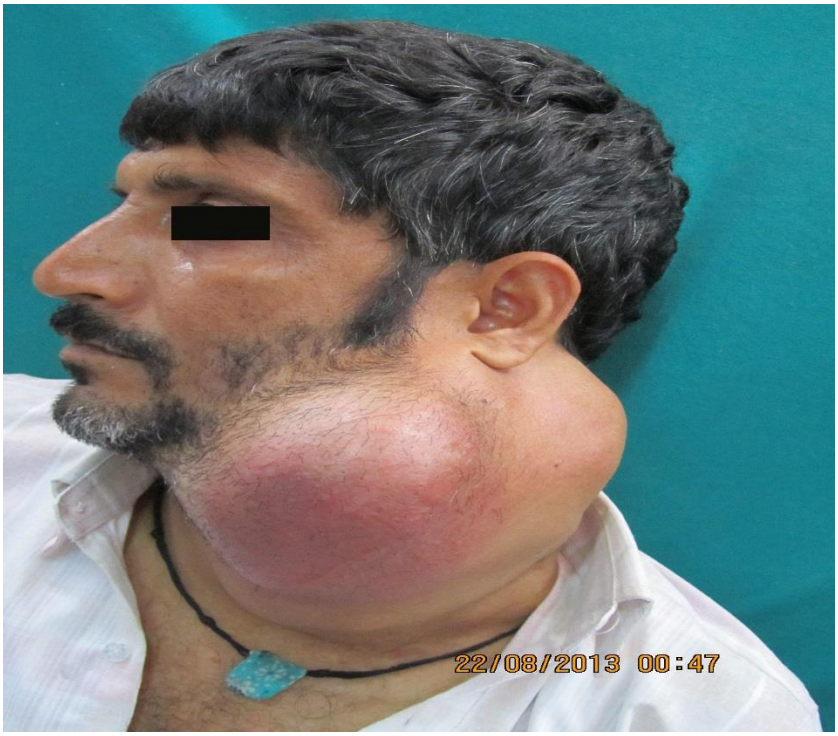
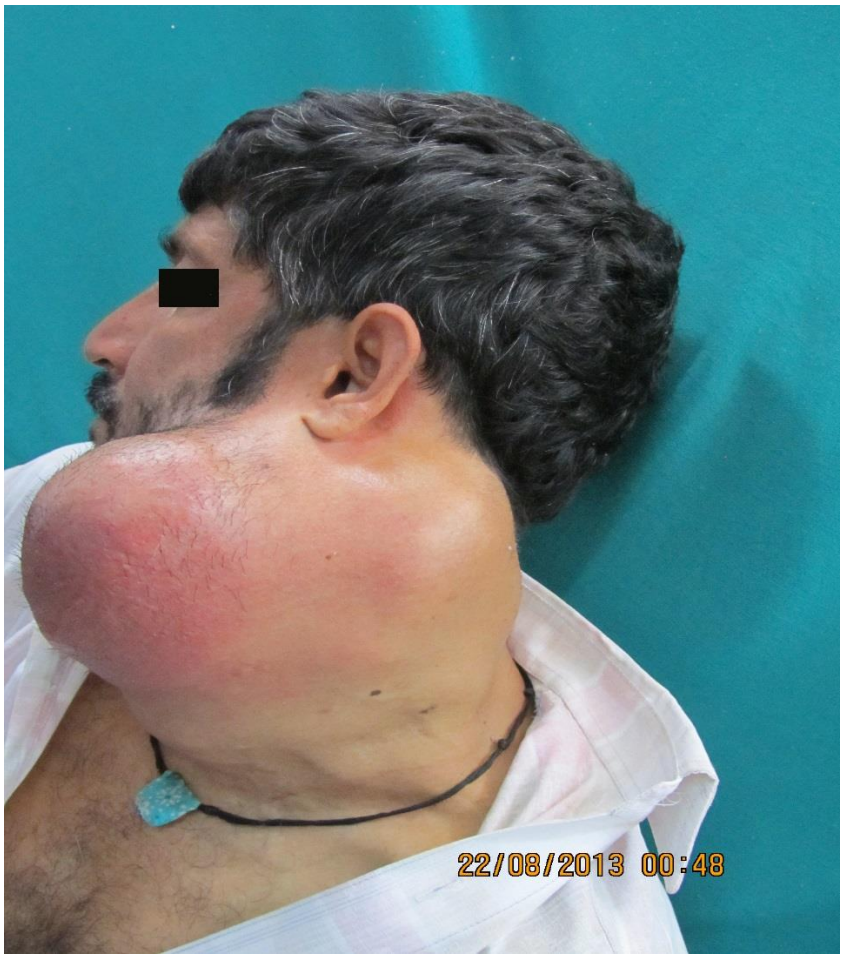


Fig.2: Lateral view of patient showing a massive swelling of approx. 20x20 cm involving left mastoid and parotid region and extending till whole length of body of the mandible till sub mental region.



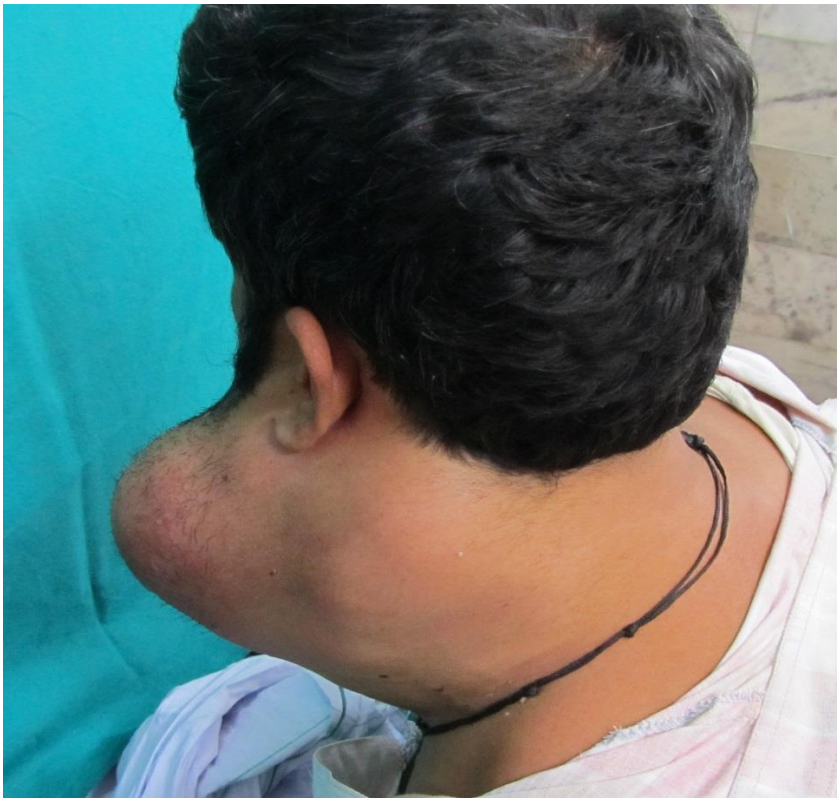


Fig. 3 & Fig. 4: Lateral view of patient showing of approx.20x20 cm involving left mastoid region and occipital region (nape of neck) and extending till left supraclavicular region involving whole length of left side of neck.

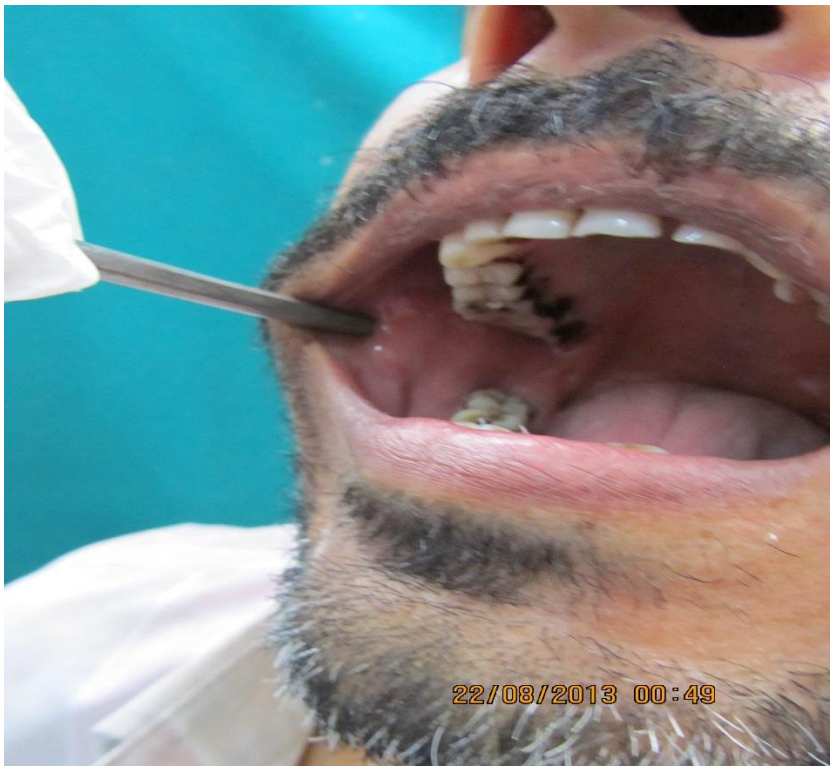


Fig. 5 and Fig. 6: Intra-oral view showing no significant finding related to swelling.

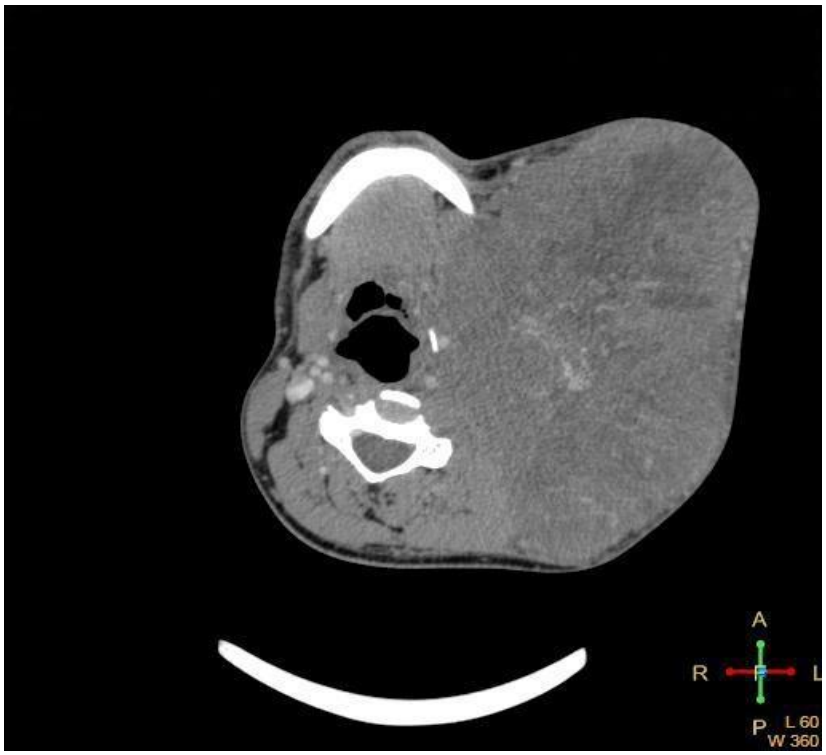


Fig. 7: Axial contrast enhanced CT showing Ill-defined heterogeneous enhancing lesion extending into the left submandibular region with compression and infiltration of the left submandibular gland while the right submandibular gland was normal.

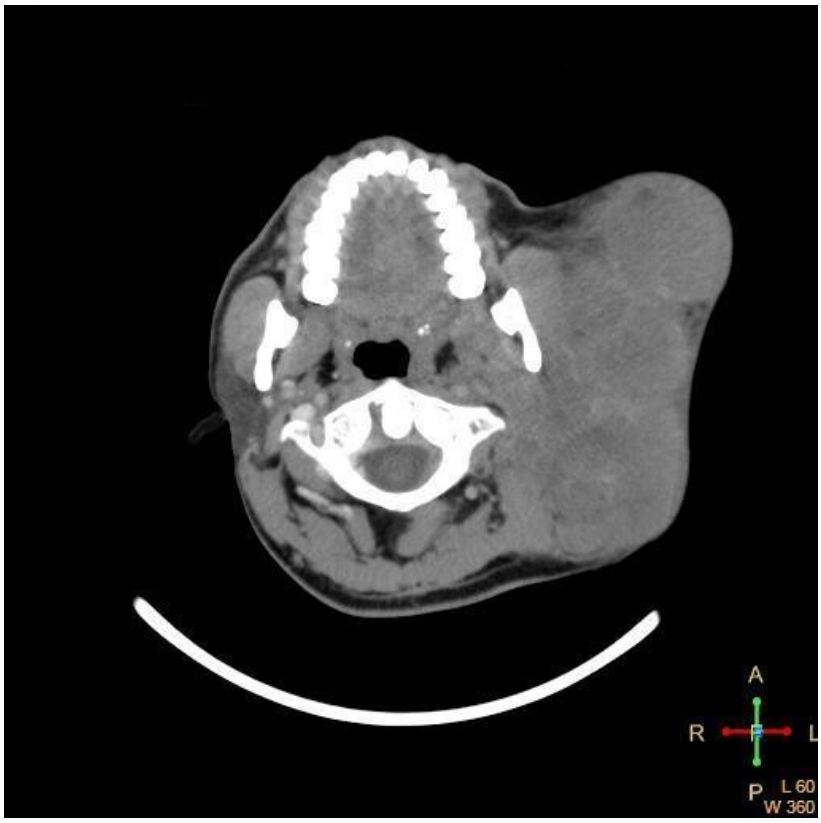


Fig. 8: Contrast-enhanced axial CT at level of C1 vertebra shows showing a lobulated heterogeneous mass lesion at the left parotid and left submandibular region. The margin with masseter and pterygoid muscles is ill-defined.

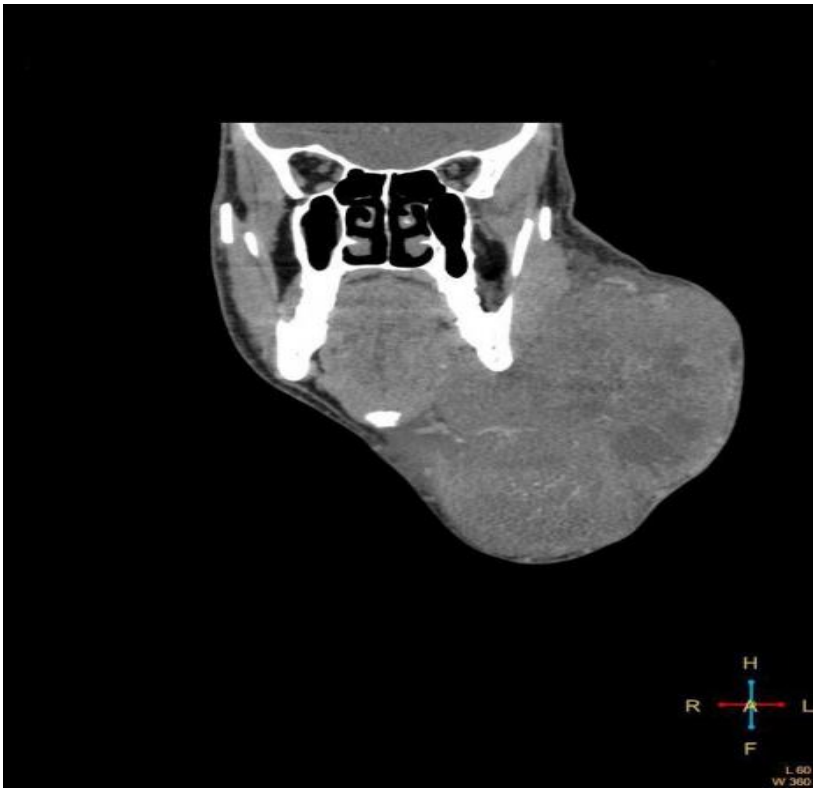


Fig.9: Coronal CECT showing large heterogeneous mass on the left extending into the submandibular region.

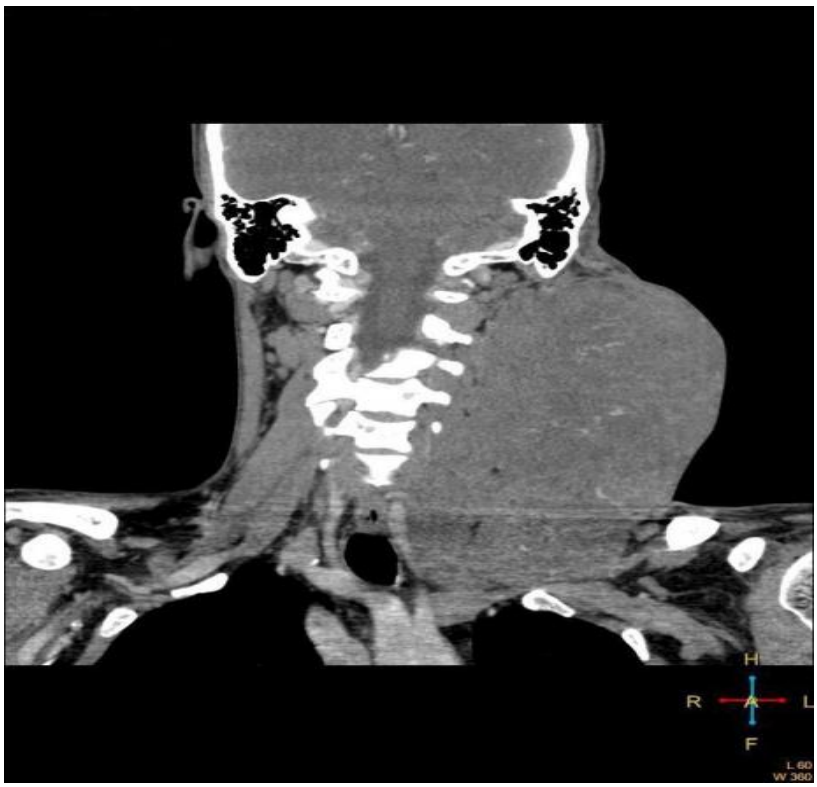


Fig. 10: Coronal CECT at vertebral level showing Ill-defined heterogeneous enhancing lesion extending up to the level of the mastoid process and external auditory canal. The sternocleidomastoid muscle is infiltrated by the lesion.

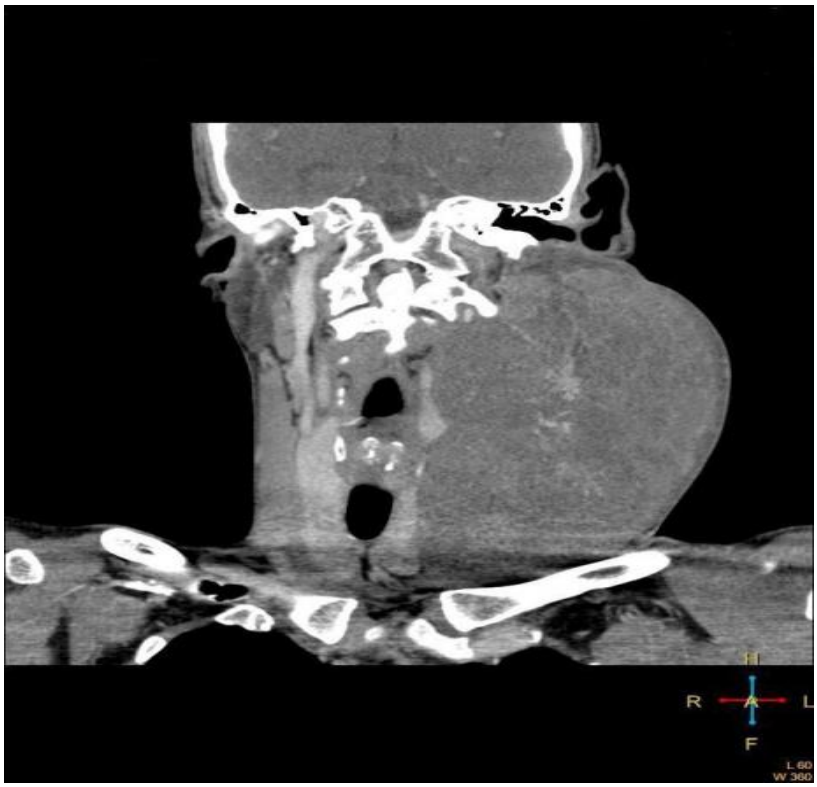


Fig. 11: Coronal CECT at vertebral level Ill-defined heterogeneous enhancing lesion displacing the left common carotid artery medially with encasement of internal and external branches. Left internal jugular vein is compressed.

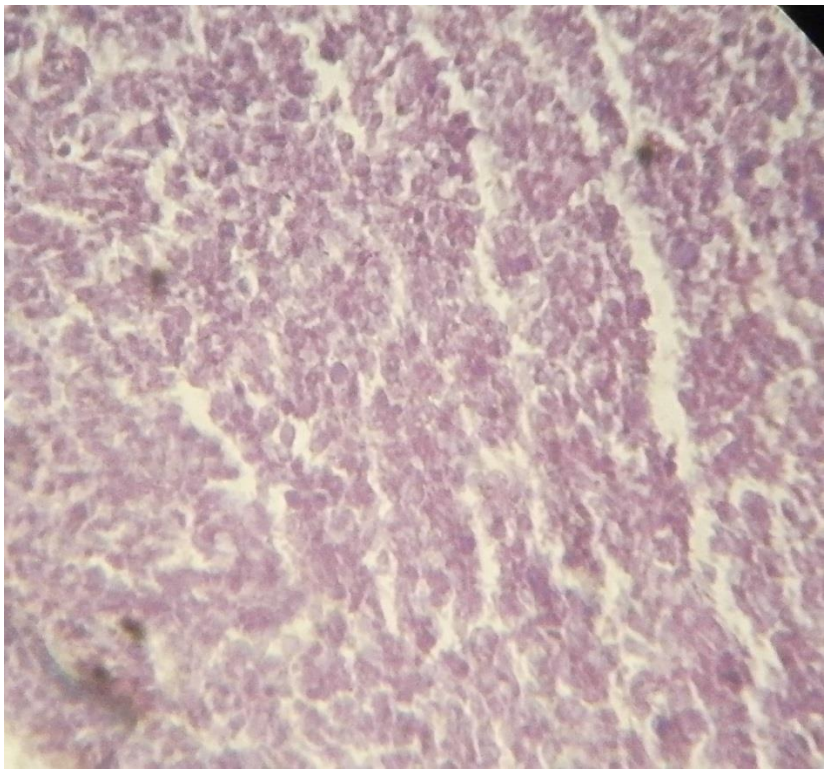


Fig. 12: Photomicrograph showing on higher magnification(100X) lymphocytes reveals features of dysplasia, cells shows severe pleomorphism and nuclear pleomorphism. Some cells are hyperchromatic but others showing vesicular nuclei.

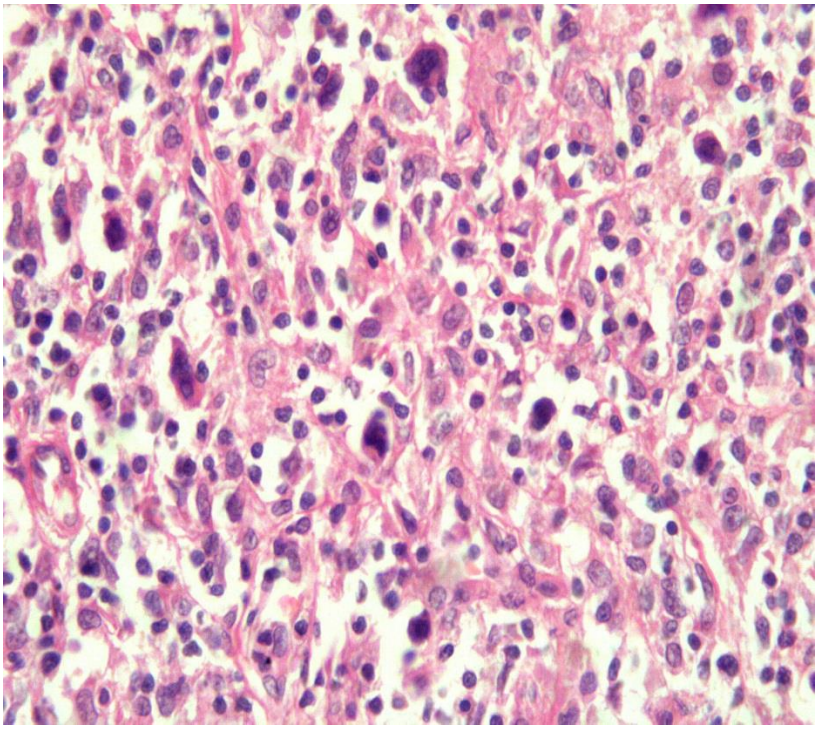


Fig. 13: Photomicrograph showing scattered large malignant cells known as Reed-Sternberg cells admixed with a reactive cell infiltrate composed of lymphocytes, histiocytes, and plasma cells.(400X) (haematoxyline and eosin staining).

DISCUSSION

Lymphomas are the second most common malignancies affecting the head and neck region. They are malignant neoplasm's of the lymphocyte cell lines[10]. Lymphomas are basically classified in two types as Hodgkin's or non-Hodgkin's lymphoma. Hodgkin's lymphoma is a disease which primarily involves the lymph nodes with secondary extranodal spread[10].They can originate either from B-lymphocyte or T-lymphocyte however Hodgkin's lymphoma originates from neoplastic proliferation of B- lymphocyte[10]. Hodgkin's lymphoma is further classified by WHO as a classical variant and a nodular lymphocyte predominant variant[11]. Hodgkin's lymphoma constitutes about 10%–12% of all lymphomas and 1-5% of head and neck tumors [10,12]. It shows bimodal pattern of incidence with major peak in 20s and broader peak over age 50 years of age. The male are more predilected than females. The incidence ratio between male to female is 1.4:1[13].

The exact etiology of Hodgkin's lymphoma is not certain. However hereditary traits, chronic infections, immunodeficiency diseases may play a role in its pathogenesis. The Epstein-Barr virus infections has been mainly implicated in pathogenesis of Hodgkin's lymphoma however exact role of Epstein–Barr virus still unknown[14]. The clinical presentations of Hodgkin's lymphoma are determined by age, geographic area and immunity of patients. In adolescents and young adults with competent immunity, it is characterized by persistent, painless and firm lymphadenopathy of cervical nodes, supraclavicular nodes and less commonly in axillary node[15].It may also present as paraneoplastic syndromes involving central nervous , renal and cardiac systems but it is quite rare[15]. In immunocompromised individuals Hodgkin's lymphoma is characterized as extra nodal disease associated with night sweats, fever, and weight loss and bone marrow involvement[16]**.** However primary extranodal Hodgkin's lymphoma is very rare[16]. Approximately ¾ cases of Hodgkin's lymphoma of head and neck involve cervical lymph nodes however waldeyer's ring involvement as primary site is unusual[17,18,19]. Hodgkin's lymphoma is primarily diagnosed by pathologic, morphologic, immunophenotypic and cytogenetic evaluation of a biopsied tissue specimen. A baseline evaluation of patients with Hodgkin's lymphoma is done by a thorough and complete physical examination,complete blood count, metabolic profile, serum lactate dehydrogenase level. Staging of Hodgkin's lymphoma is determined according to the Ann Arbor staging system which was further modified at the Cotswold's meeting in 1989[20]. Computed tomography of the head and neck, chest, abdomen and pelvis is the mainstay of staging for lymphomas. Bone marrow biopsy is mandatory for staging. The positron emission tomography (PET) with 18F-fluorodeoxyglucose (FDG) and PET computed tomography (PET/CT) is also useful for staging and evaluation of the therapeutic potential of drugs used in treatment of Hodgkin's lymphoma[21].In 1944 Jackson and Parker[22] were the first to classify Hodgkin's lymphoma histologically into paragranuloma, granuloma, and sarcoma which are equivalent to nodular lymphocyte predominant, nodular sclerosis, and lymphocyte depleted types respectively as recognized in the current Cotswold's classification system[23]**.** Due to evolutionary development in radiotherapy, chemotherapy and other salvation techniques now a day's Hodgkin's lymphoma is highly curable with long term survival. Combination chemotherapy followed by radiotherapy is now standard of care[24].

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