

CASE REPORT

Undifferentiated Nasopharyngeal Carcinoma – A Rare Case report

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ABSTRACT:

Undifferentiated Nasopharyngeal carcinoma (NPC) is a rare malignancy in most parts of the world and is one of the most confusing, commonly misdiagnosed, and poorly understood diseases. It is unusual among all the head and neck cancers due to its marked geographical predilection, highly malignant tumor growth characteristics, special difficulties in detection and staging and a high rate of treatment failure despite of its radiosensitivity. This carcinoma is rare in most parts of the world with reported incidence of around 1 case per 100,000 population per year. Nasopharyngeal carcinoma is common in Southern China and North Africa, where the etiology is related to dietary habits but it is rare in other parts of the world. Therefore, for better understanding of etiopathogenesis, clinico-pathological features, early diagnosis and prognosis of UNPC, we report a rare case of a 37 year old Male, presenting with Headache and Blurring of vision since last 1 month.

Key words: Neuroblastoma, Undifferentiated Nasopharyngeal carcinoma.

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A 37 year old Male presented with Headache and Blurring of vision since last 1 month. On examination he had Papilloedema and loss of Olfaction. He underwent CT and MRI scan which showed an enhancing lesion extending from right frontal and ethmoidal sinuses to anterior skull base to right frontal area with compression of Right frontal horn. It was excised by a combination of right frontal Craniotomy and Functional Endoscopic Sinus Surgery. Then the first postop CT scan showed near total excision of mass lesion. The Histopathology report showed Neuroblastoma. 15 Days later patient presented with severe headache and vomiting. Repeat MRI scan showed huge recurrence. So PET scan, Bone marrow aspiration, CT chest was done to look for Metastatic lesions. But there was no evidence of metastasis, so it was decided to reoperate. This time Transcranial and transfacial approach was taken and lesion

was excised. The final Immunohistochemistry report came as Undifferentiated Nasopharyngeal Carcinoma. The second post op MRI shows total excision and this time he was subjected to Radiotherapy.

INTRA OPERATIVE

1. It was excised by a combination of Right Frontal Craniotomy and Functional Endoscopic Sinus Surgery.
2. After 15 days, MRI scan showing huge recurrence.
3. PET scan was done to rule out Metastasis.
4. Reoperated through transcranial and transfacial approach.
5. Second post op MRI scan shows total excision of tumor.

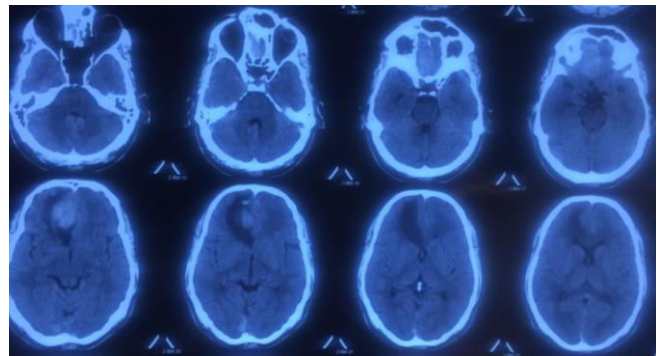
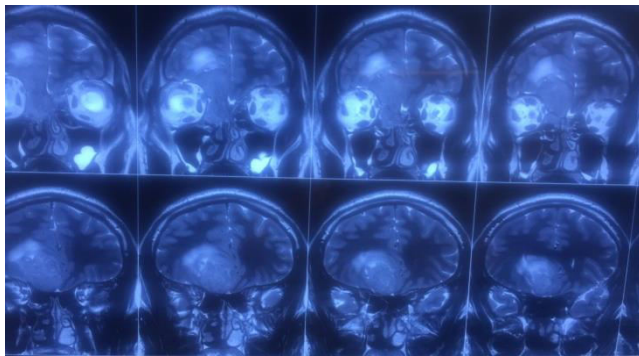


Fig 1: Pre operative CT & MRI Scan showing mass lesion.

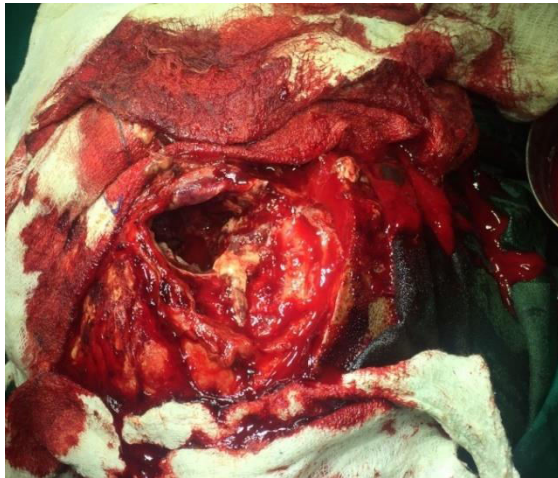


Fig 2: Excision of tumour mass by a combination of right frontal craniotomy and functional endoscopic sinus surgery

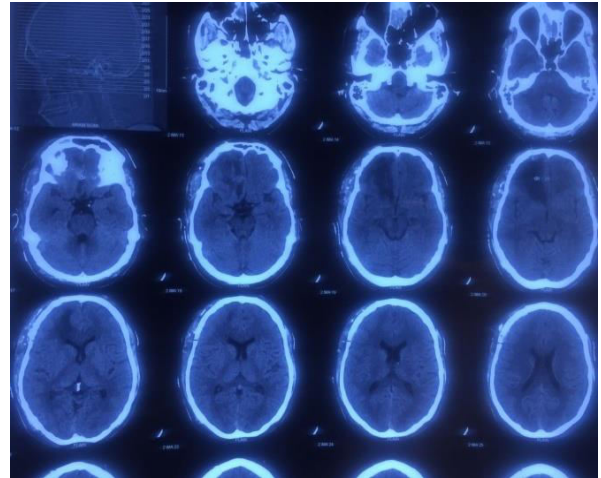


Fig 3: Post OP CT scan showing near Total excision.

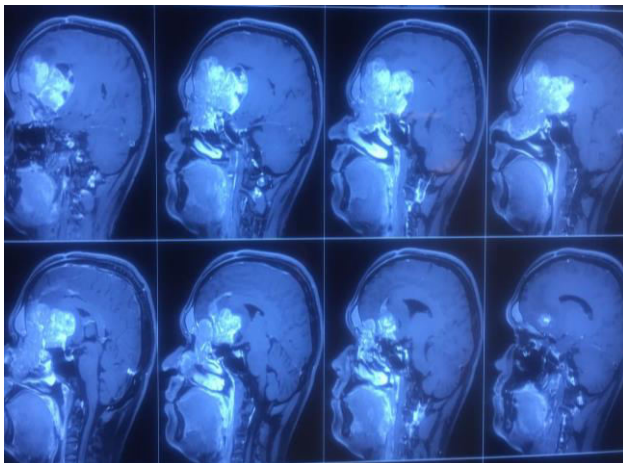


Fig 4: MRI scan showing huge recurrence

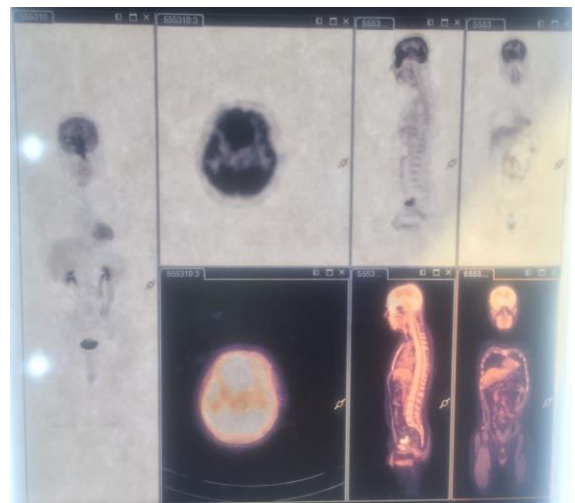


Fig 5: PET scan done to rule out Metastasis.

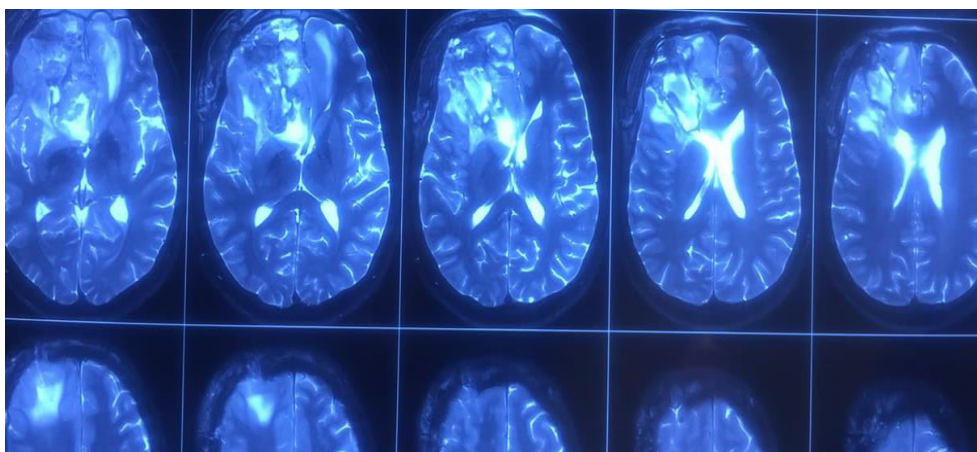


Fig 6: Second post op MRI scan shows total excision of tumor.

DISCUSSION

Nasopharyngeal carcinoma (NPC) has a high endemic incidence more common in eastern Asia, northern Africa and Alaska and is pathogenetically correlated with Epstein-Barr Virus (EBV) and is usually appreciated as a separate clinical entity. The histological classification of nasopharyngeal carcinoma proposed by WHO in 1978, categorized tumours into three groups: type I included typical keratinising squamous-cell carcinomas, similar to those found in the rest of the upper aerodigestive tract; type II included non-keratinising squamous carcinomas; and type III included undifferentiated carcinomas.

An alternative classification has divided tumours into two histological types, namely squamous-cell carcinomas and undifferentiated carcinomas of the nasopharyngeal type. The second classification is correlated with EBV serology patients with squamous-cell carcinomas have a reduced EBV titre, whereas those with undifferentiated carcinomas of the nasopharyngeal type have raised titres. The detection of the Epstein-Barr virus (EBV) nuclear antigen and viral DNA in nasopharyngeal carcinoma has revealed that EBV can infect epithelial cells and is associated with their malignant transformation. Copies of the EBV genome have been found in cells of preinvasive lesions, suggesting that it is directly related to the process of transformation. Although the incidence varies according to geographic location, approximately 1 in every 100,000 children are diagnosed annually in North America. A male preponderance is observed. The male-to-female ratio is approximately 2:1. Nasopharyngeal carcinoma has a bimodal age distribution. A small peak is observed in late childhood, and a second peak occurs in people aged 50-60 years. Childhood nasopharyngeal carcinoma is usually a disease of adolescence. Nasopharyngeal carcinoma rarely comes to medical attention before it has spread to regional lymph nodes. Enlargement and extension of the tumor in the nasopharynx may result in symptoms of nasal

obstruction (eg, congestion, nasal discharge, bleeding), changes in hearing (usually associated with blockage of the eustachian tube, but direct extension into the ear is possible), and cranial nerve palsies (usually associated with extension of the tumor into the base of the skull).

- Nasal symptoms: including bleeding, obstruction, and discharge (78%)
- Ear symptoms: including infection, deafness, and tinnitus (73%)
- Headaches (61%)
- Neck swelling (63%)

The most common physical finding is a neck mass consisting of painless firm lymph node enlargement (80%). Neck involvement is often bilateral; the most common nodes involved are the jugulodigastric, and upper and middle jugular nodes in the anterior cervical chain. Cranial nerve palsy at initial presentation is observed in 25% of patients. On nasopharyngoscopy, a mass arising in the nasopharynx is often visible. The most frequent site is the fossa of Rosenmüller. A paraneoplastic osteoarthropathy has been described in patients with widespread metastatic or recurrent disease.

Radiotherapy (RT) is the standard treatment for NPC, although it can produce undesirable complications after treatment because of the location of the tumour at the base of skull, closely surrounded by and in close proximity to radiation dose-limiting organs. In RT a dose of 65–75 Gy is normally given to the primary tumour and 65–70 Gy to the involved neck nodes, whereas the dose for prophylactic treatment for a node-negative neck is 50–60 Gy. Because of the high incidence of occult neck node involvement, prophylactic neck RT is usually recommended. The results of clinical trials that include both radiation therapy and chemotherapy generally report long-term survival rates of 50-80% overall. In a study by

Serin et al, the 5-year overall survival rate was 42% with radiotherapy alone and 58% with chemoradiation.

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