CASE REPORT

Nasopharyngeal Carcinoma in Children

A report of three cases with review of literature

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Abstract

Nasopharyngeal carcinoma is rare in children and adolescents. Nevertheless, it is considered to be the only tumour of surface epithelium afflicting children and young adults. Three such cases seen over a period of eight years (1990–1997) are reported with a review of relevant literature.

Introduction

Nasopharyngeal carcinoma is not an uncommon tumour of head and neck region in adults, more so in the high risk areas of Southern China, Hong Kong etc. Though a second peak in age incidence in the second decade has been observed in non-endemic areas (4), the occurrence of the tumour in childhood is very rare (6). It has been estimated that 5 % of primary malignant tumours in children originate in the area of head and neck (9), while nasopharyngeal carcinoma constitutes about 2% of head and neck malignant tumours in children (5). Relevance of these statistics is evident not only because the nasopharyngeal cancer has been considered as the only tumour of surface epithelium afflicting children and young adults (14) but also because the diagnosis of this rare tumour may be delayed being masked by more frequent problems of childhood, such as infections of the upper respiratory tract or other tumours (6).

We here report three cases of nasopharyngeal carcinoma in paediatric age group seen over a period of eight years (1990–1997) in E.N.T. department of S.M.H.S. Hospital, Srinagar with a review of relevant literature.

Case I

M.A, 8 year old boy presented with history of nasal obstruction, snoring and change in voice (Rhinolalia clausa) for 2 months. There was no history of epistaxis, headache, sore throat or fever. On examination, patient was a mouth breather (Fig. 1), nasal cavities were normal, except for a little excoriation of nasal vestibular skin on both sides due to long standing nasal discharge. Examination of oral cavity and pharynx was unremarkable except for mildly enlarged tonsils without hyperaemia. Neck was free of lymphnode enlargement. Posterior rhinoscopy could not be done. X-Ray soft tissue

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of nasopharynx revealed a soft tissue shadow in nasopharynx with no bone erosion.

Patient was scheduled for surgery with a provisional diagnosis of hypertrophied adenoids. Surgery and immediate post operative period was uneventful. Patient had bled, slightly more than usual, during surgery but did not require any additional measures or blood transfusion during or after surgery. Patient reported in the second week after discharge from the hospital with recurrence of nasal obstruction and epistaxis. On examination, a proliferative mass involving the nasopharynx and pushing the soft palate inferiorly, was seen. Cervical lymphnodes were enlarged in anterior and posterior triangles of neck. This time a biopsy from the mass in nasopharynx was taken, which was reported as poorly differentiated squamous cell carcinoma.

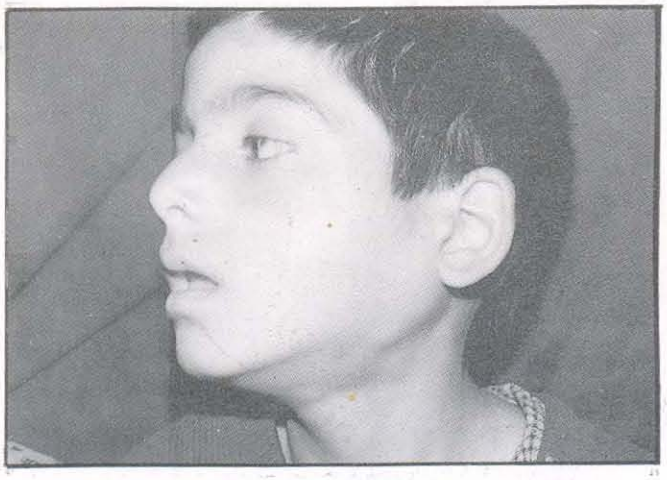

Fig. 1. Note the open mouth posture and multiple cervical lymphnode enlargment.

Case II

ZA, 9 year old boy presented with progressively increasing right sided swelling in the neck for 3 months (Fig. 2). There was no history of sore throat, nasal obstruction, epistaxis etc. On examination, a single firm lymphnode mass with smooth surface and distinct margins and somewhat restricted mobility was n under the angle of mandible on right side. No o lymphnodes were enlarged. Rest of E.N.T. examin was unremarkable. Posterior rhinoscopy could n completed. Fine needle aspiration of the swelling inconclusive and an incisional biopsy under gen anaesthesia was planned. Under anaesthe nasopharyngoscopy was first performed wh revealed an ulcero-proliferative growth on r lateral nasopharyngeal wall Biopsy was taken fr the growth.

Biopsy was reported as undifferentiated squamous carcinoma. Repeat FNAC of neck swelling revea secondary deposits in the lymphnodes of neck.

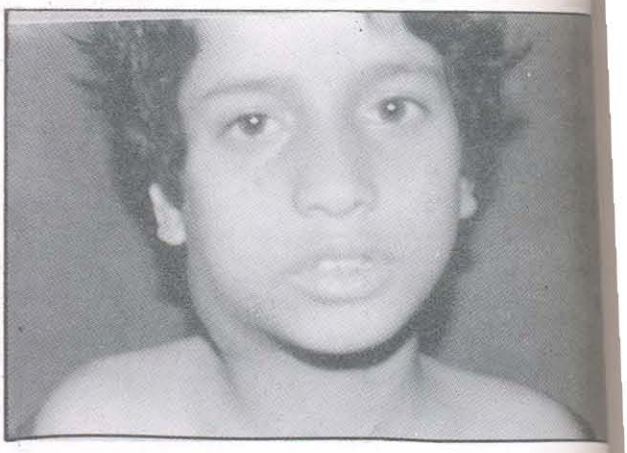

Fig. 2. Patient presented with unilateral upper deep cervic lymphnode enlargment only.

Case III

F.A., 13 year old boy presented with a classical histor of nasal obstruction, epistaxis and enlarged cervic lymphnodes on both sides of neck (Fig. 3). O examination, a proliferative growth arising from rig side of nasopharynx extending into posterior part of nas cavity was seen. Otoscopy on right side showed evidenc of fluid in middle ear. Biopsy was taken under loca

anesthesia which was reported as poorly differentiated squamous cell carcinoma.

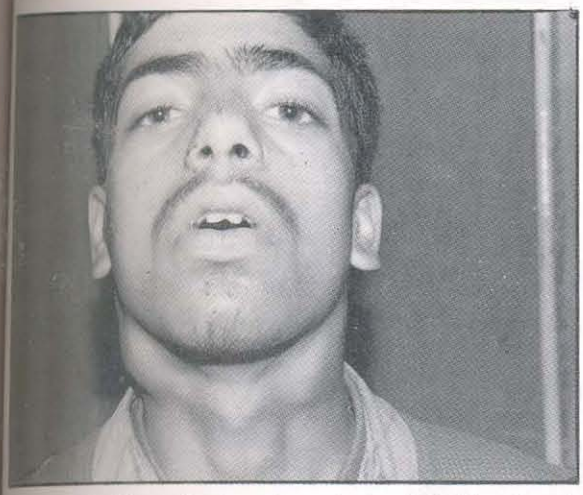

Fig. 3. Note the bilateral enlarged cervical nodes with epistaxis and open mouth posture.

Discussion

Nasopharyngeal carcinoma in children is rare. Cunningham in a 20 year review of head and neck malignancies in children, reported an incidence of 5%–12 cases (5). Hodgkins and other malignant lymphomas and soft tissue sarcomas were more common. The tumour registry in Manchester, U.K., listed 12 cases of carcinoma of the nasopharynx in children upto 15 years of age, out of a total of 1482 cases of malignant diseases of childhood from 1954–1980 (7). Fernandez identified 10 patients under 15 years of age with a diagnosis of carcinoma nasopharynx (lymphoepithelioma) in their study spanning 17 years (6). Carcinoma of the nasopharynx in adults is endemic in Chinese and other South-East Asians, where the age incidence rate begins to rise at the end of 2nd decade of life, reaches a peak in the fourth decade and then stays at a plateau (4). In certain low risk populations, however, a bimodal age distribution has been described i. e., there is also a high proportion of nasopharyngeal carcinoma in patients below 20 years of age (1, 2, 8, 10). This is thought to be the influence of different aetiological factors or variations in host response (6).

Nasopharyngeal carcinoma in children presents mainly with cervical lymphadenopathy (5, 6). This common (60–90%) presentation of nasopharyngeal carcinoma has been related to the rich network of lymphatics in the nasopharynx (6, 7, 14). It has rightly been observed that despite the comparatively high frequency of reactive cervical lymphadenopathy, congenital lesions and benign neoplasms in the paediatric population, a firm, non-tender neck mass in a child should be considered a malignancy until proven otherwise (5). Nasal obstruction, hearing disturbances, nasal discharge and epistaxis are other frequent complaints in this tumour (7). These symptoms are not very different from adult population. Since such complaints are common in children due to frequent upper respiratory tract infections, this rare tumour can, therefore, be masked, resulting in considerable delay in establishing the correct diagnosis. Our first reported case is an example in this context. Here the patient was first diagnosed as having adenoid hypertrophy.

The sites of distant metastasis, which may manifest even after complete regional control of the disease, are no different in children and adolescents when compared with adults. Metastatic disease has been reported in thoraco lumbar spine, scapula, sacroiliac joint, ilium and lungs (6).

Terminology and histological classification of nasopharyneal carcinoma has been a matter of considerable debate among pathologists. The WHO classification recognises three histological types of this tumor on the basis of their light microscopic

appearances (7) :

1. Squamous cell carcinoma :
   (a) Well differentiated
   (b) Moderately differentiated
   (c) Poorly differentiated
2. Non keratinising carcinoma
3. Undifferentiated carcinoma

It has been observed that nasopharyngeal carcinoma that occurs in children and young adults is not a differentiated squamous carcinoma as keratinization in them is absent (14). This tumour in this age group occurs more often as undifferentiated carcinoma or poorly differentiated epidermoid carcinoma (5, 12, 14). Histological type "lymphoepithelioma" reported as most frequent histological variant in children and young adults in older literature, is now regarded as a undifferentiated nasopharyngeal carcinoma since the lymphocytic element in this tumour is not neoplastic (4, 6, 12, 13, 14, 18). These facts are substantiated in the three cases being reported here. The histopathological picture in case I and III was poorly differentiated squamous cell carcinoma and that of case II was undifferentiated carcinoma.

Radiation therapy is an established mode of treatment for nasopharyngeal carcinoma in adults. (3, 4) Same treatment when applied to children and young adults produces prompt and complete tumour regression in almost all patients and results in cure in 30 to 50% (6, 10, 11, 14, 15). The better 5 year survival rate of 50 to 62.5% in younger patients, despite the aggressive disease in them, has been related to (a) higher degree of radiosensitivity of undifferentiated tumour and (b) better tolerance of radiation by the young, since their tissues and vessels have not been altered by ageing a atheromatosis (6, 12, 14). Radiation therapy, tho effective in locoregional control, can induce long t morbidity in the form of hypopituitarism hypothyroidism. (16).

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