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Case Report

An unusual case of nasopharyngeal carcinoma

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Abstract:

Though rare in other parts of the world, nasopharyngeal carcinoma (NPC) is relatively rare in Eastern and Southeast Asia (18% of all malignant tumours). Nonetheless spindle cell type of NPC is extremely rare all over the world. Ophthalmo-neurologic manifestations of nasopharyngeal carcinoma are also very rare. Here we present a case of spindle cell type of nasopharyngeal carcinoma with ophthalmo-neurological and cutaneous manifestations.

Key words: *Nasopharyngeal cancer; spindle cell type; ophthalmoplegia; vocal cord paralysis; skin manifestation*

Introduction:

Nasopharyngeal carcinoma (NPC) is a rare tumour in most of the world. But in Eastern and Southeast Asia it comprises 18% of all malignant tumours[1]. However, spindle cell type of NPC is extremely rare all over the world and described in very few papers[2, 3]. Usual manifestations of NPC include neck lump due to metastasis to cervical lymph nodes, ear fullness, hearing loss due to blockage of Eustachian tubes along with nasal bleeding and obstruction[1,4,5]. Ophthalmo-neurological manifestations due to cranial nerve paralysis and cutaneous involvement are very rare[5, 6]. In this case report, a case of NPC is presented for its very rare histopathological and clinical presentations.

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Case report:

A 12-year-old boy was admitted to Islamia Eye Hospital, Dhaka, Bangladesh with the complaints of redness and squint of the right eye with decreased vision for one year and hoarseness of voice for one month. On examination, there was paralysis of right lateral rectus muscle and multiple, firm, mobile, enlarged lymph nodes, measuring in between 1cm and 0.5cm in the right cervical region (figure 1).

The boy was HBsAg positive. His routine haematological examination revealed neutrophilic leukocytosis and high ESR (60mm after 1st hour). MRI of brain showed a slightly lobulated left cerebellopontine (CP) angle mass compressing the adjacent part of brachium pontis, brain stem and 4th ventricle and extending anteriorly to the parasellar region and also posterolatrally and

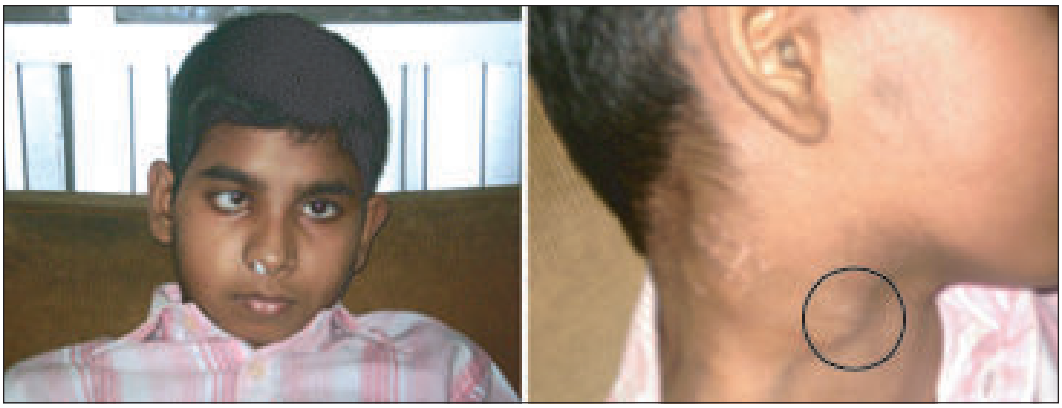

Figure 1: *Squint of right eye (inset - right cervical lymphadenopathy).*

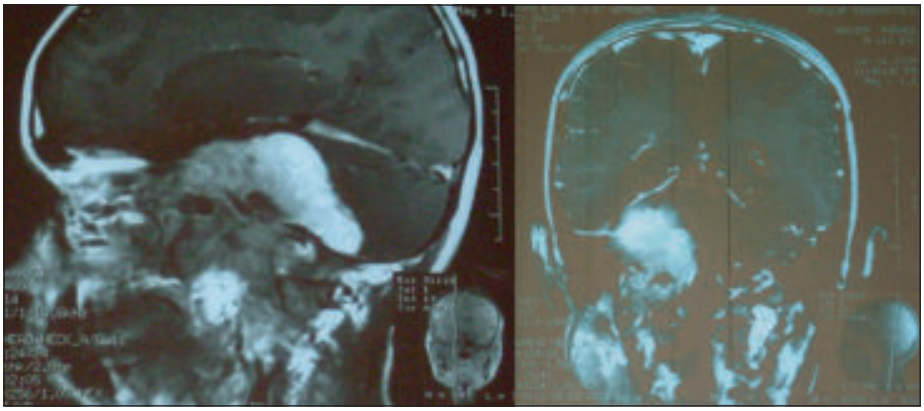

Figure 2: *MRI of brain showing the tumour, sagittal and coronal view).*

inferiorly. The lesion also extended into the right acoustic meatus. The radiological impression was acoustic neuroma. Differential diagnosis was meningioma (figure 2).

The boy was referred to the Neurosurgery Department of Bangabandhu Sheikh Mujib Medical University (BSMMU), Dhaka, Bangladesh. Fine needle aspiration (FNAC) of right cervical lymph nodes revealed non specific changes. Subsequent biopsy of lymph nodes showed a malignant tumour suggestive of metastatic undifferentiated carcinoma possibly nasopharyngeal carcinoma. Differential diagnosis was metastatic sarcoma.

Otolaryngology consultation with fibreoptic nasopharyngoscopy revealed a swelling in the right fossa of Rosenmullar in the nasopharynx. Right vocal cord palsy was also observed, may be due to vagus nerve involvement in posterior cranial fossa. Histopathology of tissue from right nose revealed a malignant epithelial tumour made of spindle shaped cells (figure 3).

The tumour cells were strongly positive for pancytokeratin and were negative for vimentin (figure 4). The final report was nasopharyngeal carcinoma of spindle cell type.

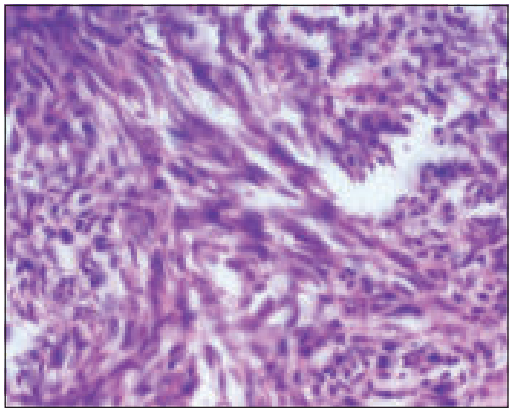

Figure 3: *Photomicrograph of tumour tissue from nose showing spindle tumour cells arranged in fascicles, H & E, x 400.*

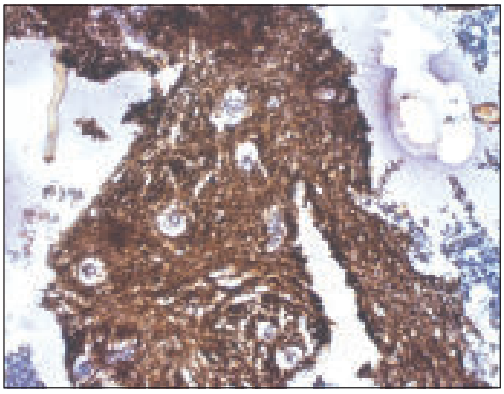

Figure 4: *Photomicrograph of immuno-histochemistry of tissue from nose showing that the tumour cells are strongly positive for pancytokeratin.*

The patient was referred to National Institute of Cancer Research and Hospital (NICRH), Dhaka for further management. But his guardians refused to continue further treatment and took him back to their village home. Follow up of the patient was continued with his father through telephone. The patient thereafter developed cutaneous rashes and nodules all over the body presumably due to metastasis of the tumour. He died nine months after diagnosis.

Discussion:

WHO has classified NPC into three groups: nonkeratinizing carcinoma (differentiated or undifferentiated), keratinizing squamous cell carcinoma and basaloid squamous cell carcinoma. Non keratinizing undifferentiated NPC, the commonest form, consists of large cells with round to oval vesicular nuclei, large central nucleoli and scant amphophilic or eosinophilic cytoplasm. In a few cases, the tumour may have plump or slender spindle shaped cells (spindle cell type) focally or extensively arranged in fascicles[7].

A study in 1987 reported that spindle cell type of NPC have a better prognosis (41% five year survival rate) than other types of NPC except keratinizing squamous cell carcinoma[8]. A study observed that spindle cell type of NPC has a low apoptotic index (AI) which correlates with its better prognosis[9]. However, Saw et al. in 1985 found no correlation between histological types and prognosis of stage I nasopharyngeal carcinoma[10].

Another study in 1993 showed that Epstain Barr virus (EBV) DNA is present in 100% cases of NPC. But viral genome is more abundant in nonkeratinizing, spindle cell and undifferentiated carcinoma[11].

In broad sense, spindle cell carcinoma of head neck is thought to be a biphasic tumour composed of squamous cell carcinoma and a malignant spindle cell component. It was first described by Virchow in 1865. Its etiology and clinical features are same as squamous cell carcinoma. But it is more aggressive and has worse response to radiotherapy in contrast with squamous cell carcinoma[12].

In a study in 1986 described 25 spindle cell tumour of upper aerodigestive tract. They found that, spindle cells in twelve out of twenty five cases were keratin positive and in almost all cases, were vimentin positive. However,

when combining ultrastructural characters with immunohistochemistry, fifteen cases revealed epithelial differentiation[13].

The first case of spindle cell type NPC was described by Nofal from Bristol, United Kingdom in 1983[2, 3]. In a series in 1983 reported 140 cases of NPC. Among these, 12 were fibrosarcoma like (spindle cell type)[14]. The next two cases were reported from India in 1986[3].

In most of the reported cases, tumour cells have squamous differentiation. Spindle shaped cells are usually present at only some foci and merge with squamous cells[3]. In the current case, the tumour cells are mostly spindle shaped and unlike other reports, these cells are positive for pancytokeratin only and negative for vimentin.

Common manifestations of NPC includes neck lump (60%), ear fullness (41%), hearing loss (37%), nasal bleeding (30%), nasal obstruction (29%), head pain (16%), ear pain (14%), neck pain (13%), weight loss (10%), diplopia (8%)[4].

Cranial nerve paralysis is an important but less common presentation in patients with NPC. In most reports, the frequency of cranial nerve paralysis ranged from 13.3-29.9%. Patients with erosion of the base of the skull and/or cranial nerve paralysis are considered to be the most advanced stage of NPC[5]. Single or multiple cranial nerve palsies in NPC depend on the extent and direction of tumor invasion. Invasion of the cavernous sinus and orbit may affect cranial nerves II to VI, while erosion of the bone in the posterior cranial fossa may induce palsies of IX to XII[15]. The most frequently involved cranial nerves are V, VI, IX, X accounting for 50% of all palsies. IX and X are invariably involved together and are most common group to be affected. Isolated single cranial nerve palsy is common with nerve V and VI[4].

Ogunleye in 1999 carried out a retrospective study with analysis of 79 patients with nasopharyngeal carcinoma seen in 12 years between 1986 and 1997. Among the 79 cases 20 (25%) had ophthalmo-neurologic manifestations. The ages ranged between 11 years and 70 years with a male: female ratio 2.4:1. In this study, visual impairment and corresponding optic nerve lesions constituted 20% of the manifestations. Diplopia and ophthalmoplegia constituted 17% and 18% respectively[16].

The current case presented with a huge tumour which invaded into the parasellar region and extended posteriorly upto cerebellopontine angle and also posterolaterally and inferiorly. The patient had squint of right eye due to paralysis of right lateral rectus muscle which is supplied by abducens (6th cranial) nerve. The patient also had hoarseness of voice due to right vocal cord palsy probably due to vagus (10th cranial) nerve involvement.

Distant metastasis of nasopharyngeal carcinoma (NPC) usually occurs to skeleton, lung, liver and extra regional nodes. Skin metastases from NPC are extremely rare[6]. A report in 2004 presented four cases with skin metastasis of NPC and identified 12 previous cases in a literature search[17]. A paper in 2006 reported another case of NPC with extensive nodular skin metastasis[6].

Distant skin metastases are thought to develop through hematogenous spread whereas local metastases occur by spread via the dermal lymphatics. Common sites of cutaneous metastasis are the scalp, neck, chest, trunk, forearm, thigh, and penis. In the cases reported in the literature, skin metastases were observed five to 35 months following the treatment of the primary tumor. The survival rate of patients with skin metastases is generally poor. It varied

between one and ten months in the reported cases[6].

In the current case, patient's father described the cutaneous lesions as nodules and rashes as seen on tiger's skin. These were distributed diffusely all over the body specially over trunk. The patient died nine months after diagnosis and the rashes appeared a few days before his death.

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