Reminder of important clinical lesson

A young boy with a maxillary swelling and closed rhinolalia

Shaji Thomas,¹ Preeti Nair,² Anil Kumar,² Karthik Hegde,² Vikram Singh²¹ Department of Oral & Maxillofacial Surgery, People's College of Dental Sciences and Research Centre, Bhopal, India² Department of Oral Medicine and Radiology, People's College of Dental Sciences and Research Centre, Bhopal, India

Correspondence to Preeti Nair, shajihoss@gmail.com

Summary

Non-Hodgkin's lymphoma belongs to a group of lymphoid neoplasia that is diverse in the manner of presentation, response to therapy and progress. The most common region for extranodal lymphoma is the gastrointestinal tract while for oral cavity buccal vestibule, posterior hard palate or gingiva is the common site. The maxilla is affected more commonly than the mandible. The oral picture being the sole manifestation highlights the importance of bearing this entity in the differential diagnosis of swelling in the jaws.

BACKGROUND

A rapidly enlarging lesion in a patient in the second decade must arouse a suspicion of non-Hodgkin's lymphoma (NHL).

CASE PRESENTATION

A 10-year-boy complained of pain and swelling on the right side of his face since 3 months and difficulty in breathing from the right nostril since 1 month. He presented with swelling on the right side of the maxilla, which rapidly increased in size in 3 months. Swelling was associated with pain radiating to the temporal and frontal regions. Sleep was disturbed and nasal tone of voice was noticed. He had difficulty in breathing, which increased in intensity on lying down.

Extraorally, a diffuse swelling was present on the right side of the maxilla. It measured approximately 6×7 cm in size, extending from the infraorbital ridge up to 1 cm above the lower border of the mandible. The surface was smooth; it was non-tender and firm in consistency (figure 1). Intraorally, a tender, soft swelling was present on the buccal vestibule in relation to the upper right first molars and also on the soft palate crossing the midline in relation to 16, 17. Grade III mobility was noted in 16, 17 (figure 2).

INVESTIGATIONS

Intraoral periapical view and occlusal radiographs revealed bone destruction in relation to 16, 17 (figures 3 and 4). Orthopantomograph revealed displacement of 18 apically and PNS view revealed a radiopaque mass obliterating the right maxillary sinus (figures 5 and 6).

Contrast-enhanced computed tomography (CECT) showed a moderately enhancing soft tissue density involving the maxillary sinus, nasal cavity and right ethmoidal air cells. Lesion causing erosion of medial and lateral wall of right maxillary sinus, lateral nasal wall and nasal septum was seen filling the nasal cavity causing compression over the nasopharynx. CECT was suggestive of malignant mass in the right maxillary sinus region (figure 7).

Fine needle aspiration cytology reports suggested round cell malignant tumour while the biopsy report confirmed NHL of diffused large B cell type (figure 8).

DIFFERENTIAL DIAGNOSIS

Differential diagnosis of rhabdomyosarcoma, juvenile ossifying fibroma, adenoid cystic carcinoma and NHL were arrived at.

TREATMENT

The patient was administered the CHOP regimen (cyclophosphamide, adriamycin, vincristine and prednisone) and showed considerable remission.

OUTCOME AND FOLLOW-UP

Patient is currently under regular follow-up and remains disease-free (figure 9). There was significant improvement in his voice following treatment.

DISCUSSION

Early diagnosis and staging are essential for effective treatment, and lymphomas must always be included in the

Figure 1 Swelling on the right side of the face.

Figure 2 Swelling on the buccal vestibule in relation to the upper right first molars and also on the soft palate crossing the midline.

Figure 3 Intraoral periapical radiograph showing loss of alveolar bone and root resorption of 16, 17.

Figure 4 Occlusal view radiograph showing bone loss and resorption of roots in relation to 16, 17.

differential diagnosis of lesions of the oral cavity associated with the maxillary sinus.

Children present with a higher proportion of extranodal NHL of the head and neck, and an early diagnosis is necessary for adequate and successful management.¹

The sites of NHL involvement in the head and neck area can be subdivided into three distinct categories: (1) nodal; (2) extranodal, lymphatic; and (3) extranodal, extralymphatic. Extranodal, lymphatic NHL refers to tumour found in the Waldeyer's lymphatic ring. The extranodal, extralymphatic category includes all NHL not in nodes or Waldeyer's ring with the major areas of involvement including the orbit, sinus, nose, mandible, deep facial spaces, parotid gland and dermis. In the head and neck region, nodal NHL is most common, followed by extranodal, extralymphatic and extranodal, lymphatic.²

The WHO in 2001 gave a compilation of lymphoma statistics. According to the classification, there are four major types of childhood NHL:

- ▶ B cell NHL (Burkitt and Burkitt-like lymphoma) and Burkitt leukaemia
- ▶ Diffuse large B cell lymphoma
- ▶ Lymphoblastic lymphoma
- ▶ Anaplastic large cell lymphoma

There are also other types of NHL occurring in children, which include:

- ▶ Lymphoproliferative disease associated with a weakened immune system
- ▶ Rare NHL that is more common in adults than children.³

The most common presenting symptoms of sinonasal lymphomas are nasal obstruction, epistaxis, headache and unilateral facial, cheek, or nasal swelling. Other infrequent symptoms are diplopia, blurred vision and nasal or cheek pain.¹

NHLs of the sinonasal tract are uncommon malignancies representing 3–5% of all malignancies, with NHL accounting for 60% of all lymphomas. Sinuses are the primary site of NHL in 90% of cases, most commonly invaded by diffuse large B cell lymphoma while primary nasal tumours occur in just 10% of cases.^{3–5} In a study by Chalastris *et al*, the most frequent histological type was B cell lymphoma.^{1 6}

NHL classifications fail to effectively predict biologic behaviour and treatment plans for this disease. Alternative methods like AgNOR staining could be used to determine cell proliferation rate and differentiation of benign from malignant neoplasms like NHL, bone tumour, skin tumour and liver tumour.⁷

Figure 5 Paranasal sinus radiograph showing radiopaque mass obliterating right maxillary sinus and displacement of 18.

Eisenbud and Slootweg found 70% of lesions in the maxilla, the most common site being the palate and gingiva. Freeman estimates that intraoral presentations represent 2.6% of all extranodal NHL and Fukuda quotes a figure of 5%.⁸

There are no characteristic clinical features of lymphoma of the oral region. The presenting signs and symptoms are secondary to the lesion. They occur as local bone swelling, tooth mobility, painless inflammation of the mucosa with or without ulcerations, and rarely facial or dental pain. Additional observations include trismus, otalgia, gingival

ulceration, sinusitis or cervical lymphadenopathy.⁸ The oral lesions are characterised by swelling, which may grow rapidly and then ulcerate. In some cases, these become large, fungating, necrotic, foul-smelling masses. Pain is a variable feature.⁹

Sinonasal NHL can be expansile with permeative bony wall involvement, as suggested by Kondo *et al.*²

In Western populations, lymphomas of the maxillary sinus are more common than in the nasal cavity. On the contrary, in Asian patients, the nasal cavity is more common as a primary site than the maxillary sinus.⁸

Figure 6 Orthopantomogram showing apically displaced 18, root resorption in 17 and haziness of right maxillary sinus.

Figure 7 Soft tissue density involving maxillary sinus, nasal cavity and right ethmoidal air cells. Lesion causing erosion of medial and lateral wall of right maxillary sinus, lateral nasal wall and nasal septum was seen filling the nasal cavity causing compression over the nasopharynx.

Figure 8 Fine-needle aspiration cytology report.

Figure 9 Postoperative after 26 days, no extraoral swelling.

Bone marrow morphology continues to be the gold standard in evaluating marrow pathology in lymphoma, immunohistochemistry and molecular studies being supplementary techniques.¹² High-grade lymphomas are associated with a 60% mortality at 5 years after diagnosis and treatment.¹⁰

A review of several reports suggests that the best treatment outcomes are obtained with the CHOP regimen,

given at three-week intervals. Rituximab is a therapeutic antibody directed against the CD20 surface antigen, which is frequently present in lymphoma cells. Its use in conjunction with CHOP augments a treatment response in lymphomas expressing the CD20 antigen. Chemotherapy is frequently followed by loco-regional radiotherapy at a dose of 30 to 40 Gy.^{6 11–15}

Learning points

- ▶ NHL should be considered in the differential diagnosis of fast growing swellings with closed rhinolalia.
- ▶ Radiological and histopathological investigations should be used for the diagnosis of NHL.
- ▶ Role of oral physicians (specialists in Oral Medicine & Radiology) is important as they may be the first to encounter such patients in institutional practice.
- ▶ Correct diagnosis and treatment can decrease the morbidity usually seen in such diseases.

Competing interests None.

Patient consent Obtained.

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